Upper Face and Orbit "Degloving" Dog Bite Injury

Joseph Bergmann, M.D., Ken Lee, M.D.†, Richard Klein, M.D.†, and Charles B. Slonim, M.D., F.A.C.S.**

Abstract: **A 70-year-old woman who attempted suicide lay unconscious on her floor for an unknown time period while her 2 pet dachshunds chewed her upper face and bilateral periorbital areas including all 4 eyelids, both lacrimal glands, all of the conjunctiva from both eyes, the extraocular muscles on the left side only, and anterior orbital fat from both sides. A subtotal exenteration was performed on her left orbit and a temporoparietal fasciocutaneous flap was used to reconstruct her right orbit with buccal mucosa replacing both the bulbar and palpebral conjunctiva. To the authors' knowledge, this is the first report of such extensive orbital injuries from dog bites.**

CASE REPORT

A 70-year-old woman who attempted suicide was found unconscious on the floor of her home. Her 2 pet dachshunds chewed her upper face and periorbital areas, devouring all 4 eyelids, both lacrimal glands, all of the conjunctiva from both sides, the 4 rectus muscles on the left side only, and anterior orbital fat from both sides. The orbital rims were completely denuded (Fig. A).

Upon arrival to the emergency department, all of the skin and subcutaneous tissues between her zygomatic arches and her hairline including all 4 eyelids had been consumed. There was no evidence of the supraorbital neurovascular bundle on either side. Her left eye was completely disinserted from all 4 rectus muscles and was displaced inferonasally. The anterior chamber of this eye had a hyphema with no evidence of corneal or scleral rupture. The conjunctiva, lacrimal gland, and anterior orbital fat were absent. The right eye had a remarkably clear cornea with no damage to the extraocular muscles. Similar to the left eye, the entire conjunctiva, lacrimal gland, and anterior orbital fat were absent, without damage to the globe.

Enucleation of the left eye was performed immediately; its rehabilitation would have been impossible because the conjunctiva and lacrimal gland were absent. All 4 of its rectus muscles had been mangled. Because mucous membrane grafting was planned for reconstruction of the more intact right eye, it was felt that the limited quantity should not be shared with the left eye. Amniotic membrane grafting was not considered because of a lack of conjunctiva progenitor cells.

To repair the right eye, 2 large mucous membrane grafts were harvested from both lateral buccal mucosal areas. These were sutured to the limbus with 8-0 Vicryl sutures. Each of the 2 grafts were oriented in a vertical direction and folded in half to create superior and inferior fornices.

The approximate location of the superficial temporal artery and the frontal and parietal arterial branches on the scalp were marked (Fig. B). A large, elliptical temporoparietal, fasciocutaneous flap on the right side was created and placed over the right orbit. The flap was placed through a subcutaneous tunnel created over the lateral orbital rim (Fig. C). Direct closure of the recipient site was accomplished with staples after undermining and mobilization of the surrounding scalp.

There are disadvantages to using a hair-bearing flap, such as the temporoparietal flap used in this case, to reconstruct the eyelids. Because the hair follicles are not disrupted in these full-thickness flaps, hair growth in front of the visual axis or toward the ocular surface is possible.

A full-thickness opening in the center of the elliptical flap was made to accommodate the cornea. Three preplaced, double-armed 5-0 chromic sutures were placed in the fold of each mucous membrane (the proposed fornix) that was already attached to their respective superior and inferior limbal areas. Each of the double-armed chromic sutures was passed full-thickness through the temporoparietal flap ~1 cm above the "corneal" opening in the flap superiorly and 1 cm below the "corneal" opening inferiorly. A single 6-0 silk traction suture was placed at the free edge of each mucous membrane graft and brought through the "corneal" opening so that these edges could be easily retrieved to later form a new mucocutaneous junction with the skin of the temporoparietal flap.

Once the six 5-0 chromic "fornix" sutures were secured, the free edges of the mucous membrane grafts were sutured to the skin edges of the corneal opening in the flap. Split-thickness skin grafts from the thigh were used to cover the remaining exposed areas of denuded facial bones and the enucleated socket. The right cornea remained clear. The corneal opening was filled with Bacitracin ophthalmic ointment to further protect the cornea (Fig. D).

During the first 3 to 4 months after surgery, the patient maintained hand motion vision OD. The hair-bearing, temporoparietal flap required shaving to keep the corneal opening patent. A small 3- to 4-mm area of clear cornea could be seen after the ointment that filled it was removed (Fig. E). The area around this clear zone appeared to be keratinizing. Examination at the slit lamp was not possible at that time.

Five months after reconstruction, the skin of the temporoparietal flap was closed over the cornea. The patient was seen by a dentist in maxillofacial surgery to fabricate an external prosthesis for her left side. She elected to have osseointegrated prostheses, which will require placing 2 implants in the right side of the nose and 3 implants in the left orbit.

DISCUSSION

It is estimated that more than 1 million Americans per year suffer bite injuries inflicted by cats or dogs.[1] Dog attacks, particularly those on children, account for 10 to 20 deaths in the United States each year.[2] Dog bites to the head, face, and neck are far more common in young children and account for ~70% of these injuries.[1] Facial injuries from dog attacks most commonly involve soft-tissue structures. Bony injuries primarily occur in younger children, with 87% of these cases being reported in children younger than 16 years of age.[1] Common injuries resulting from dog bites include laceration, facial nerve damage, lacrimal duct injury, levator resection with resulting ptosis, and major blood loss. As with all bite injuries, infection is a cause of concern. The flora of a canine mouth is diverse but the most common bacterial isolates from dog bite injuries include *Pasteurella* species and anaerobic bacterium. *Staphylococcus aureus* and *Streptococcus pyogenes*, which are commonly in-

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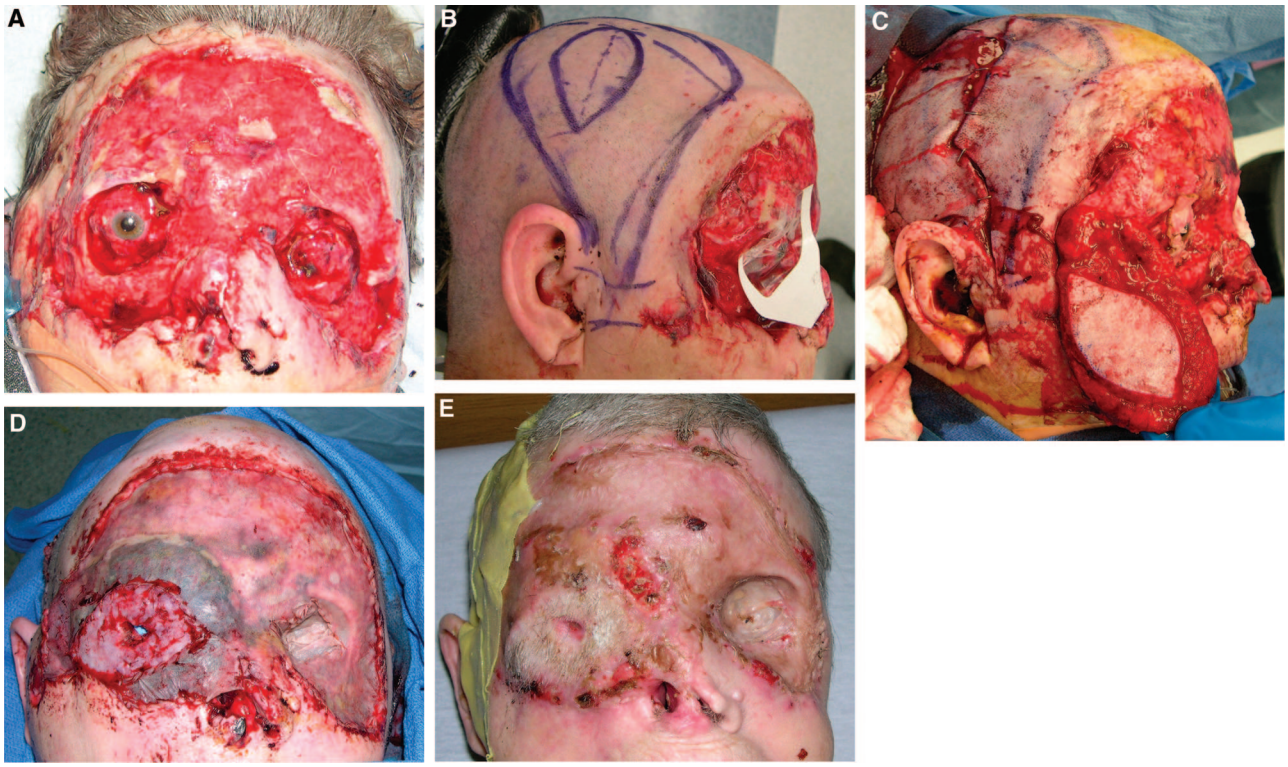

A, Preoperative injury. **B,** Outline of temporoparietal fasciocutaneous flap. **C,** Harvested temporoparietal flap. **D,** Immediate postoperative result with split-thickness skin graft over remainder of defect. **E,** Two months postoperatively.

volved in cutaneous infection, were relatively uncommon in dog bite injuries.[3] Tetanus and rabies must also be considered complicating factors of a bite injury.

Culturing the wounds at the time of the injury prior to treatment is of little value in predicting potential wound infections because most wounds contain multiple organisms. Wound culture results can later direct the appropriate antibiotic treatment when initial empirical therapies are found to be ineffective. It is important to prevent secondary wound infections, which can cause systemic complications with occasionally fatal results.[4]

The temporoparietal fascial flap used in this case was originally described by Monks in 1898[5] but rarely appeared in the medical literature until the early 1980s. In our case, in which there was a total loss of neurovascular supply to the traumatized area, a pedicle flap with its own blood supply was used. The temporoparietal fascial flap is an ideal option in cases requiring extensive reconstruction because of its dependable blood supply, minimal impact on donor site, large area available for harvest, and flexibility of use.[6] Furthermore, the low incidence of necrosis and infection make such flaps well suited for extensive reconstruction in injuries that result in poor host sites for other tissues.

Microvascular free flaps have the advantage of providing healthy, vascularized tissue for recipient sites that may have been compromised by large tissue defects. Large fasciocutaneous free flaps such as the anterolateral thigh flap described by Song et al.[7] in 1984, lateral thigh flap described by Baek[8] in 1983, radial forearm flap,[9] and ulnar forearm flap[10] have also been used for large head and neck tissue defects. The advantage of using radial forearm, ulnar forearm, or lateral arm flaps is the availability of thinner and more pliable skin for grafting.

Face transplantation, still considered experimental, has been performed in a case of severe dog bite injury.[11]

In conclusion, this case technically represents a degloving-like injury of the upper face and orbital areas. Reconstruction was aimed at preserving vision first and covering the facial defects second. Both were accomplished initially, however, preserving useful vision has been difficult. The total lack of ocular adnexa and the support they typically offer an intact globe is paramount to the survival of the eye. The use of a temporoparietal flap assisted in covering the orbitofacial defect on the right side but is severely limited in serving as a substitute for the natural orbital adnexa.

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Browlifting as an Alternative Procedure for Apraxia of Eyelid Opening

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Abstract: **Essential blepharospasm is an idiopathic disorder that consists of spontaneous, spasmodic, and involuntary eyelid closure in the absence of ocular disease. Some patients develop an inability to open their eyelids in the absence of orbicularis spasms. These patients have essential blepharospasm combined with apraxia of eyelid opening. Botulinum toxin injections are the treatment of choice for blepharospasm but results may be insufficient, especially in cases associated with apraxia. Apraxia can be treated surgically by levator aponeurosis advancement, frontalis suspension, and upper myectomy. The authors report the first browlift using polypropylene suture to manage eyelid apraxia associated with blepharospasm as an alternative and minimally invasive procedure.**

Essential blepharospasm is an idiopathic disorder that consists of spontaneous, spasmodic, and involuntary eyelid closure in the absence of an ocular disease. In blepharospasm, eyebrows are lowered beneath the superior orbital margin and eyelids are closed by contraction of the pretarsal, preseptal, and orbital fibers of the orbicularis oculi muscle.[1] When the spasms subside, the eyelids can open. However, some patients develop an inability to open their eyelids in the absence of orbicularis spasms. These patients have essential blepharospasm combined with apraxia of eyelid opening. Botulinum toxin injections are the treatment of choice for blepharospasm but results may be insufficient, especially in cases associated with apraxia.[1] Advancing the levator aponeurosis, frontalis suspension, or upper myectomy are the only useful surgical treatments for eyelid apraxia.[2,3] We describe the first browlift using polypropylene suture (Contour Threads, Surgical Specialties Corp., PA, U.S.A.) as an alternative and minimally invasive procedure to manage eyelid apraxia associated with blepharospasm.

CASE REPORT

A 71-year-old Asian woman had a 15-year history of blepharospasm. Thirteen years ago, she underwent upper blepharoplasty performed by a plastic surgeon, with no improvement of spasms.

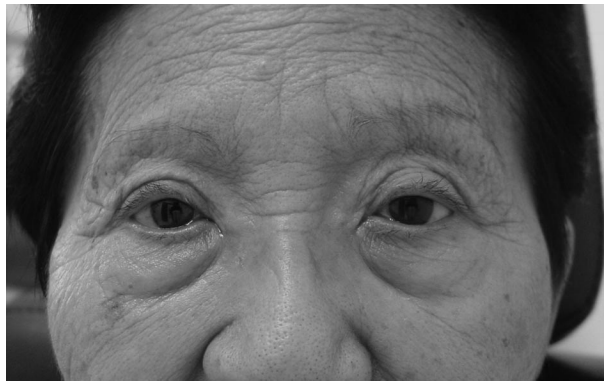

FIG. 1. A 71-year-old Asian woman with blepharospasm and secondary eyelid apraxia.

At our first evaluation, she had involuntary bilateral blepharospasms, bilateral ptosis with elevated eyelid crease, and good levator muscle function (12 mm) OU. After successful botulinum A toxin treatment, we performed levator aponeurotic reattachment and excised some preseptal orbicularis muscle. Thereafter, she had 6 botulinum toxin injections at intervals of 4 to 6 months, with good results.

Eventually, however, she reported that she was not able to open her eyelids anymore, even when the orbicularis spasms ceased. Despite elevation of her eyebrows, her eyes remained closed.

To offer a conservative treatment, we performed a browlift using a modified polypropylene suture through minimal incisions in the scalp and in the superior margin of the eyebrow. The suture was fixed to the temporalis fascia. After the procedure, the patient was able to open her eyelids again and she reported an improvement in her quality of life (Figs. 1 and 2). She was followed for 8 months and remained free of symptoms in this period with no additional botulinun toxin injections.

DISCUSSION

Although the etiology of eyelid apraxia is not well understood, it has been seen in some extrapyramidal disorders such as Parkinson disease, Huntington chorea, progressive supranuclear palsy, and Shy-Drager syndrome,[4] which suggests basal ganglia involvement.[1,2] Jordan et al.[2] found 7 cases (7%)

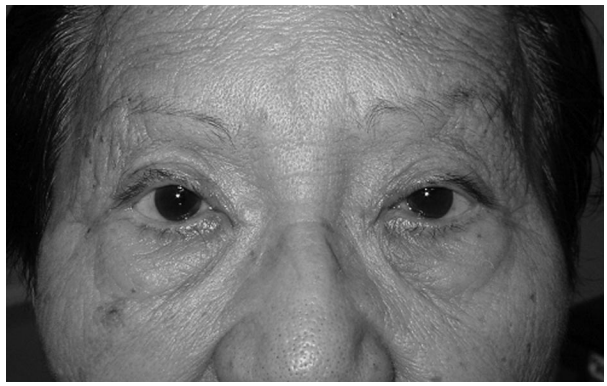

FIG. 2. Six months after browlifting, the patient reported no recurrence of symptoms and improvement in her quality of life.

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of functionally disabling apraxia of eyelid opening in 100 consecutive patients with blepharospasm studied.

The diagnosis of apraxia of eyelid opening is not easy to make in patients with blepharospasm. Ptosis from levator disinsertion or induced by botulinum toxin injections, and difficulty in eyelid opening because of ocular irritation must be ruled out. In a recent study, Georgescu et al.[3] stated the incidence of apraxia of eyelid opening in the general population of patients with blepharospasm is much higher than their previously described rate of 7% as a result of a better understanding and recognition of this disorder.

Adenis et al.[4] advocated that in blepharospasm with apraxia, orbicularis resection alone is not sufficient and should be combined with frontalis suspension. However, our patient underwent aponeurotic ptosis repair and limited myectomy, and developed apraxia about 2 years later.

Frontalis suspension is a proven and widely used method for treatment of ptosis with poor or absent levator muscle function, and it seems to be a valuable procedure for blepharospasm and apraxia of eyelid opening.[5] However, the frontalis suspension technique is generally performed under general anesthesia and requires a strip of autogenous or banked fascia lata or synthetic material.

Recently, Georgescu et al.[3] assessed the results of upper eyelid myectomy surgery on 45 patients with blepharospasm with associated apraxia. Fifteen patients (33%) reported that their apraxia was cured by myectomy. Thirty patients (66%) had residual apraxia after myectomy but with improved symptoms. Among these, 13 patients (43%) had more than 75% improvement in apraxia after the first procedure. The authors concluded that upper eyelid myectomy appears effective in treating apraxia associated with blepharospasm. However, there are several negative side effects of myectomy that are predictable and occur to some degree in all patients, and the healing process may take several months.[1]

Based on the need to offer a more conservative procedure for these patients, we attempted an essentially simple surgical technique for browlifting using a polypropylene suture. The suture has many dents with sharp edges along its length that hold tissue firmly.[6] Sulamanidze et al.[6] used these threads to lift facial tissue in 186 patients with failure in just 4 cases (2.5%). The patient we described had a successful browlift with improvement of symptoms during follow-up.

In our patient, the use of dented polypropylene suture was a very simple, conservative, time-saving, and effective procedure for browlifting. We theorize that browlift surgery alleviates apraxia of eyelid opening because it has the effect of holding the weight of the eyelid, which could make the effort to open the eyelid easier. More studies are needed to confirm the success of this procedure in managing apraxia of eyelid opening.

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Bulbar Conjunctival Autograft With Rectus Muscle Transeyelid Traction Suture for Diplopia Due to Bulbar Conjunctival Scarring

Robert H. Hill III, M.D., and Thomas A. Bersani, M.D.

Abstract: **Cases of bulbar conjunctival fibrosis from pterygium surgery, strabismus surgery, trauma, chemical exposure, and inflammatory conditions can be difficult to manage. Despite surgical approaches entailing autografts, amniotic membranes, and antimetabolites, preventing postoperative scarring and contracture can be difficult, particularly in patients with susceptibility to scarring. The authors present a case of a 39-year-old man with diplopia secondary to subconjunctival scarring and fibrosis after multiple pterygium surgeries. The authors describe a unique surgical approach using a medial rectus transeyelid traction suture as a means of preventing postoperative conjunctival graft contracture and functional disability.**

Bulbar conjunctival scarring and fibrosis is not an uncommon sequela of pterygium surgery, strabismus surgery, trauma, chemical exposure, and inflammatory conditions of the eye. With severe contracture, bulbar conjunctival scarring can cause diplopia and can also be cosmetically displeasing.[1] Surgical methods for prevention including conjunctival autograft, amniotic membrane graft, and antimetabolites have been well documented.[2] We present a case of bulbar conjunctival scarring after multiple pterygium procedures causing diplopia that was surgically corrected with a bulbar conjunctival autograft and rectus muscle transeyelid traction suture.

CASE REPORT

A 36-year-old man had a nasal pterygium removed from his right eye 2 years prior to presentation. He subsequently had the pterygium re-excised with a conjunctival autograft 8 months later. The patient later developed diplopia in lateral gaze and had a second recurrence of the pterygium. He underwent a third excision with the intraoperative use of the antimetabolite mitomycin-C, again with recurrence of diplopia.

The patient was referred to our care in November 2006, with persistent diplopia in primary and lateral gaze. On examination, the patient had limitation of abduction OD to only 20° and also experienced pain with attempted abduction. The patient had binocular fusion with left, up, and downgaze, but developed binocular, horizontal diplopia in primary and right gaze. There was a dense band of subconjunctival scarring from the medial limbus extending to the caruncle with the caruncular

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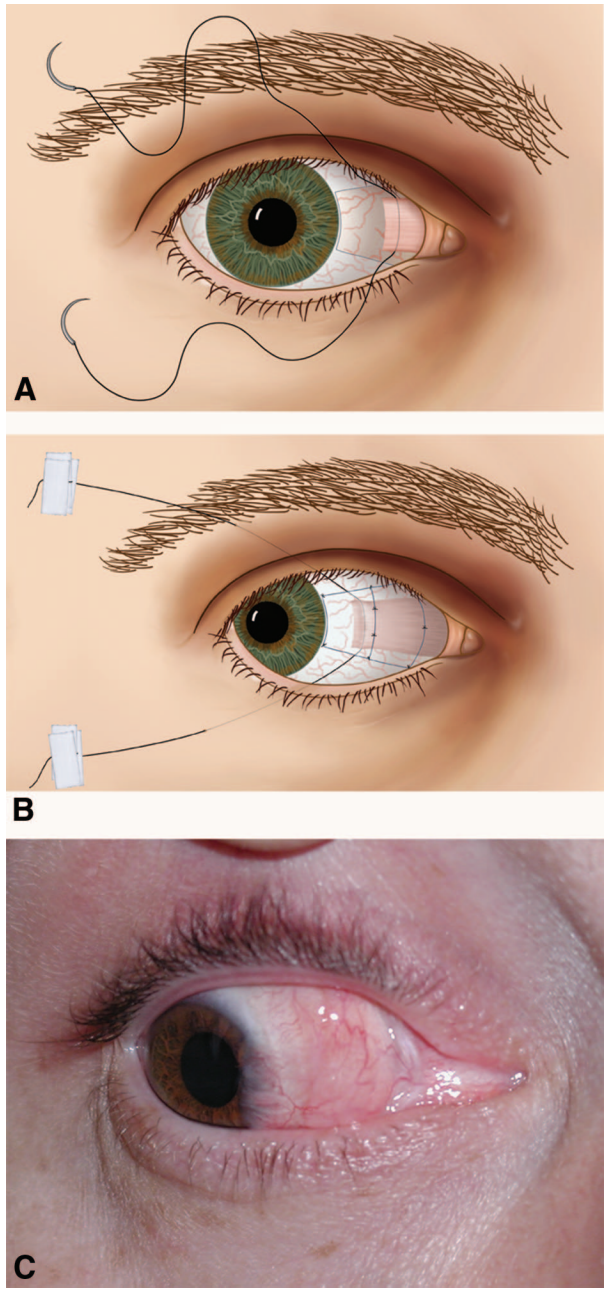
epithelium dragged laterally. There was also scarring extending supramedially in the supramedial conjunctival fornix with some symblepharon and restriction of the upper eyelid.

After waiting ~6 months from his past surgery for the conjunctival tissue to sufficiently heal, the patient was brought to the operating room. Forced ductions under anesthesia demonstrated severe restriction with attempted abduction of the affected eye. The medial bulbar conjunctiva was undermined medially toward the caruncle at the medial canthal angle, releasing scarring in the subconjunctival plane. The scar tissue and conjunctiva were dissected off the globe extending medially. Subconjunctival scar tissue was encountered on the surface of the medial rectus as far medially as the caruncle and the superficial medial orbit. As the scar tissue was undermined, the globe was noted to become more mobile, until forced ductions were freely mobile without restriction or adhesion, and the globe could be placed in an abducted position with minimal tension. Deep orbital dissection was not necessary. A bridle suture of 6-0 nylon was passed underneath the medial rectus muscle insertion (Fig. A) and then the inferior arm was passed through the inferolateral conjunctival fornix and brought out through the lateral preseptal lower eyelid skin. The superior arm of the suture was passed through the superior lateral conjunctival fornix and the lateral preseptal upper eyelid skin. With traction of the sutures, the globe was brought in an abducted position. Mitomycin-C 0.4 mg/ml was applied topically to the medial bulbar surface and medial rectus muscle for 1 minute with a moistened cotton tip applicator and then vigorously irrigated away. The conjunctival autograft was harvested from both the ipsilateral and contralateral superior bulbar surface with no closure of the donor sites required. The conjunctival grafts were placed side by side, one along the corneoscleral limbus and one adjacent to the caruncle and were sutured to one another and the surrounding conjunctiva using multiple 7-0 chromic sutures. Next, the 2 ends of the 6-0 nylon traction suture were taped to the temporal skin superiorly and inferiorly with tincture of benzoin and steri-strips, placing the globe in extreme abduction (Fig. B).

Two weeks postoperatively the medial rectus bridle suture was removed. The patient had mild limitation (~5°) of horizontal eye movements OD. Range of fusion was estimated clinically as the number of degrees from primary that the patient was able to hold fusion before developing diplopia. He had full binocular fusion in upgaze (50°) and downgaze (60°), but did develop diplopia with more than 55° of right gaze and more than 45° of left gaze. The patient returned 4 months postoperatively with diplopia only in extreme abduction (>55°) and minimal bulbar conjunctival scarring (Fig. C). One year later the patient has diplopia with ~30° of abduction, has no pain, and is very pleased with the results.



A, Sketch demonstrating area of excised conjunctiva and scar tissue. A bridle suture of 6-0 nylon has been passed underneath the medial rectus muscle insertion. **B,** Sketch demonstrating the bridle suture passed through the inferolateral conjunctival fornix and brought out through the lateral preseptal lower eyelid skin. The superior arm of the suture is passed through the superior lateral conjunctival fornix and the lateral preseptal upper eyelid skin. The conjunctival autograph is in place with 7-0 chromic sutures and the bridle suture has been taped to the temporal skin with a tincture of benzoin and steri-strips. **C,** Postoperatively, the bulbar conjunctival autograft has healed with minimal globe restriction in abduction.

DISCUSSION

A series of reactions encompasses wound healing including an acute inflammatory reaction, regeneration, and finally contraction.[3] The contraction phase of wound healing in the eye can be disabling if it impairs vision or motility. Conjunctival wounds or autografts with significant scarring and contracture can be cosmetically displeasing and can cause functional impairment. Conjunctival tissue in particular may be particularly prone to scarring and contracture due to a constant change in tissue size and conformation secondary to ocular motility. We performed a surgical technique placing the eye in traction to prevent postoperative scarring and fibrosis and to treat diplopia. We believe our results were favorable because of both decreased eye motility and prevention of autograft malleability. While having the eye in traction was somewhat uncomfortable postoperatively for this patient, it was tolerable, and after removal of the suture all of his discomfort was immediately resolved. We believe this technique is a practical solution to avoid postoperative scarring and fibrosis and to restore eye motility.

The use of a traction suture and mitomycin-C for restrictive strabismus is well documented.[4,5] Preoperatively, it was not clear whether the patient's diplopia was secondary to restriction from scar contracture or from a restrictive strabismus, but given his surgical history, we believed it was the former. During surgery, we were able to demonstrate that his diplopia was caused by scar contracture and fibrosis of the overlying bulbar conjunctiva rather that from restriction of the muscle when the eye became freely mobile after releasing the scar tissue. If we had found that it was in fact the muscle that was scarred, this particular procedure would probably not have been beneficial and strabismus surgery should be a consideration.

During the surgery, when attention was turned to harvesting the conjunctival graft, the amount of tissue harvested from the ipsilateral eye was limited by the small area of scarring superiorly from the previous surgeries. We acknowledge that the use of an amniotic membrane graft is a useful alternative when there is limited tissue available. However, we decided to harvest tissue from both the ipsilateral and contralateral bulbar surfaces because we believed that having an epithelium-lined autograft would be more likely to heal by primary intent. We also acknowledge that fibrin glue may have been an alternative to suturing the conjunctival autograft, theoretically causing less conjunctival reaction. We have limited experience using fibrin glue in this particular situation, although we believe that we have demonstrated that suturing the graft is an effective alternative in treating this condition.

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Steatocystoma Simplex of the Eyelid

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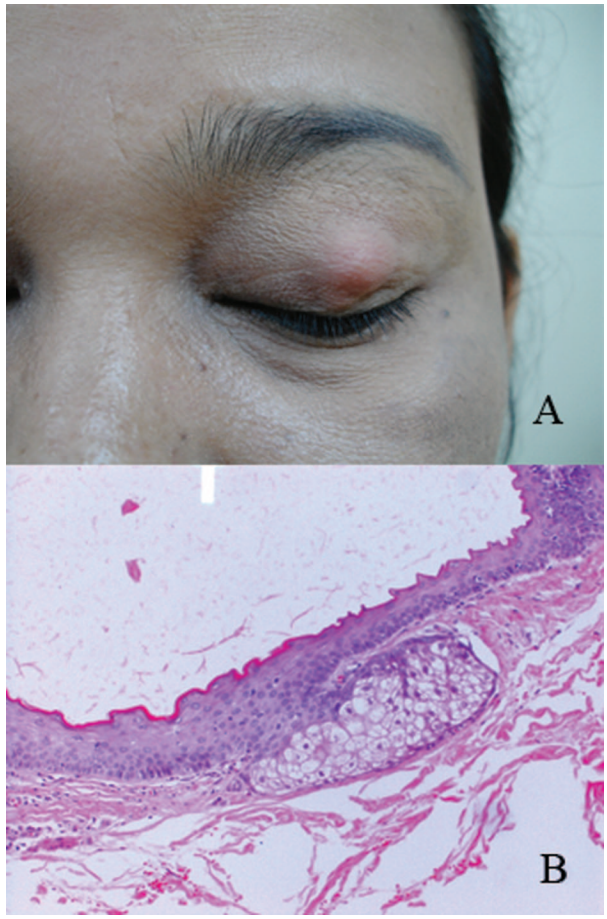
Abstract: **A 47-year-old woman presented with a recurrent nodular lesion on the left upper eyelid that had been resected twice in the previous 5 years. Intraoperative findings showed a thin-walled cystic nodule containing a creamy substance. Histopathology identified steatocystoma; the diagnosis of steatocystoma simplex was further characterized by the absence of a family history and involvement of only one lesion. Steatocystoma is a cystic lesion that is often seen in dermatology practices but that has been reported rarely on the eyelid. Its clinical appearance is indistinguishable from other cystic lesions of the periorbital area and it should be considered in the differential diagnosis of eyelid lesions.**

A variety of etiologies cause nodular lesions of the eyelids. The lesions we frequently encounter are eyelid infections and inflammations, namely, hordeolum, chalazion, and papilloma, nevus, and inclusion cyst. Nonetheless, some atypical causes can mimic those etiologies, such as liquid silicone migration from aesthetic injections.[1] We report a patient with a nodular lesion of the upper eyelid that was diagnosed as steatocystoma simplex.

CASE REPORT

A 47-year-old woman presented with a recurrent nodular lesion of 1-year duration with mild ptosis of the left upper eyelid (Fig. A). She had undergone resection twice in the previous 5 years without supporting pathologic reports. The lesion enlarged insidiously and was cystic in consistency.



A, Macroscopic photograph shows a nodular lesion on the left upper eyelid. **B,** Microphotograph of the cyst shows an eosinophilic corrugated surface and sebaceous gland attached to the wall of the cyst (hematoxylin-eosin, ×200).

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Examination showed a painless, slightly mobile lump measuring 1.9 × 1.4 cm on the left upper eyelid, with mild erythema. The margin reflex distance was 3 mm OD and 2.5 mm OS. Other findings were unremarkable.

The provisional diagnosis could have been either a recurrent chalazion or a sebaceous cyst. The patient underwent surgical excision under local anesthesia. Intraoperatively, a cystic lesion that adhered firmly to the tarsus and levator aponeurosis was observed. After excision of the entire cyst, there appeared to be a buttonhole on the tarsus. A segment of the levator aponeurosis was also disconnected and reattached over the tarsus. The cyst contained a creamy yellowish substance.

The histopathologic report described a cyst lined with stratified squamous epithelium containing amorphous material. The epithelial lining of the cyst had a corrugated luminal surface with flattened sebaceous lobules attached to the cystic wall. The pathologic diagnosis was steatocystoma (Fig. B).

DISCUSSION

Steatocystoma originates from the nevoid malformations of the pilosebaceous duct junction. Two entities have been documented: steatocystoma simplex and multiplex. Steatocystoma simplex, which is usually sporadic and was first described by Brownstein[2] in 1982 in a series of 30 patients, manifests as one lesion on the face, scalp, neck, chest, axilla, upper or lower limbs, and back, and has also been reported at the caruncle.[3] Steatocystoma multiplex, which is autosomal dominant and presents with multiple lesions, is identical in appearance and histology to steatocystoma simplex.[2] Steatocystomas have a unique histology and express keratin 10 and 17, as found in the cysts. Steatocystoma is rare in ophthalmologic practice. There has been only one report of a follicular hybrid cyst on the tarsus that described a lesion composed of pilomatricoma and steatocystoma that perforated the palpebral conjunctiva.[4]

Steatocystoma cannot be distinguished from other cystic lesions in the clinical setting, so histopathologic study plays an important role in differentiation and diagnosis. In steatocystoma, the cyst is lined by a thin layer of stratified squamous epithelium, there is no granular layer, and a characteristic dense hyaline cuticle is present on the epithelium. The luminal surfaces are undulating and the pathognomonic sign is a sebaceous gland attached to the cystic wall. The lesion resembles an epithelial inclusion cyst that is lined with stratified squamous epithelial cells with epidermal keratinization, and keratin 10 was identified by immunohistology. Other cysts, such as vellus hair cyst, are lined with stratified squamous epithelium, their lumen contains small hairs and keratin 17, while the tricholemmal cyst is lined with stratified squamous epithelium and exhibits tricholemmal keratinization; the cell wall grows continuously and narrows the lumen. All these cysts derive from the infundibular part of the hair.[5] Other developmental cysts, such as dermoid cyst, should be considered because they have a sebaceous gland in the cystic wall that is lined with keratinized epidermis and dermal appendage; the lumen contains keratin, oil, and hair shafts. In the current case, the histopathologic findings showed the unique feature of steatocystoma, although the keratin was not studied. The patient had only one lesion and no family history of such lesions, both of which are compatible with steatocystoma simplex.

In conclusion, rare reports of steatocystoma simplex on the eyelid have been published. Ophthalmologists should be aware of this unusual case and consider steatocystoma in the differential diagnoses of any such cystic mass in this area.

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Schwannoma of the Lower Eyelid in a 13-Year-Old Girl

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Abstract: **A 13-year-old girl presented with a history of a slowly enlarging, mobile, right lower eyelid mass of 2 years' duration. Excisional biopsy revealed the characteristic histopathologic features of a schwannoma. After complete excision, there was no recurrence at 1-year follow-up. Schwannoma is a rare eyelid tumor, appearing sporadically and in a solitary form, which should be considered in the differential diagnosis of eyelid tumors in children.**

Schwannomas are benign neurogenic tumors arising from Schwann cells of the peripheral nerve sheath. Schwannomas are located in the soft tissues throughout the body but also may be seen in the ocular tissues. The orbit is the most commonly affected area,[1] but schwannomas of the uveal tract and the conjunctiva also have been reported. Eyelid schwannomas are rare. Only 9 cases have been reported: 8 in adults and 1 in a child.[2–9] We report the first case of lower eyelid schwannoma in an adolescent.

CASE REPORT

A 13-year-old girl presented with a painless right lower-eyelid mass that had been slowly enlarging for the previous 2 years. Aside from swelling, there had been occasional redness of the skin over the lesion. The patient was diagnosed as having a chalazion and offered an intralesional steroid injection by another physician; this option was rejected by the patient's family.

On physical examination, a 2-cm, round, nontender, mobile lesion was felt between the skin and inferior orbital rim (Fig. A). The results of an ophthalmic examination were unremarkable. Under local anesthesia, the encapsulated mass lesion lying anterior to the orbital septum just beneath the skin and orbicularis muscle was completely excised via anterior dissection through the skin (Fig. B, C). Histopathologic examination of the lesion revealed bundles of spindle cells with no mitotic activity (Fig. D). Immunohistochemical analysis dem-

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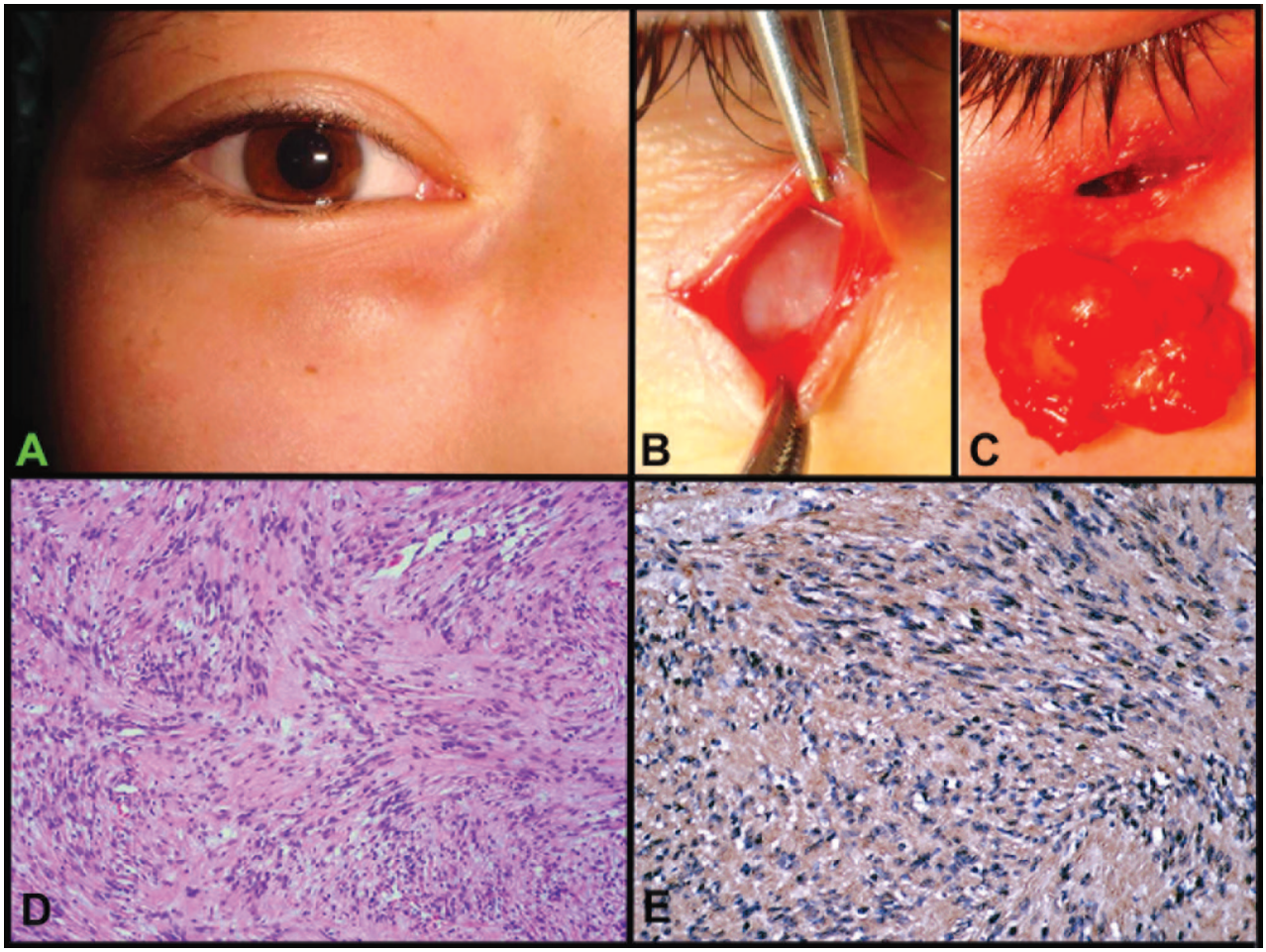

A, Round lower eyelid mass in a 13-year-old girl. **B**, Lesion seen with a smooth capsule beneath the skin and orbicularis. **C**, Gross appearance of the capsulated lesion after complete excision through a skin incision. **D**, Spindle cells arranged as a palisading fashion in an Antoni A area (hematoxylin-eosin, ×100). **E**, S-100 positivity in spindle cells (×200).

onstrated a positive reaction for S-100 protein and vimentin (Fig. E). These findings were consistent with a benign schwannoma. No further treatment was planned after the excision. Systemic examination revealed no clinical signs of neurofibromatosis or tuberous sclerosis. No local recurrence was observed at 1-year follow-up.

DISCUSSION

Schwannomas are tumors of the peripheral and cranial nerves that are frequently benign. They are often well-encapsulated lesions, which makes them easier to excise completely. The histopathologic appearance of schwannoma is distinctive, with 2 characteristic patterns: Antoni A areas are composed of compact spindle cells that are usually arranged in palisades and Antoni B areas consist of tumor cells scattered in a myxoid matrix. A strong positive reaction for S-100 protein, especially in hypercellular Antoni A areas, is strong evidence for a diagnosis of schwannoma.

Besides the eyelid, the orbit is a more common location for a schwannoma, accounting for ~1% for all orbital tumors.[1] The oculomotor, ciliary, and supraorbital nerves are the reported origins of tumors affecting the orbit. Branches of the supraorbital, supratrochlear, and infraorbital nerves are the presumed origins of eyelid schwannomas. There have been no reports of malignant transformation after total excision of an eyelid schwannoma; thus, total excision seems to be curative.

Schwannoma of the eyelid was first reported in 1960,[2] and after that, 7 cases of eyelid schwannomas have been described in adults who ranged in age from 18 to 66 years.[2–9] This is the second report of a schwannoma in a child and the first of a schwannoma in the lower eyelid of a child. The first reported case of a schwannoma in a child was that of a slowly enlarging, painless mass in the upper eyelid of an 8-year-old boy that recurred 2 years after the primary excision.[3] Chalazion is a common form of the eyelid mass in children, but schwannoma should be considered in cases of a slowly growing, mobile, firm lesion, or when attempted drainage of a lesion proves unsuccessful. Also, schwannoma should be considered in the differential diagnosis of spindle-cell tumors of the eyelid and periocular region.

The schwannoma is a classic feature of neurofibromatosis, a disorder that primarily affects cell growth of neural tissues. There is a lack of knowledge regarding the positive predictive value of a solitary schwannoma of ocular tissues for future neurofibromatosis. However, screening of these patients for neurofibromatosis may be reasonable, especially when the schwannoma presents during childhood.

Schwannoma has rarely been a preoperative clinical diagnosis in an eyelid tumor; all reported cases have been diagnosed histopathologically, as in our case. Detailed histopathologic and immunohistochemical analyses of the lesion are essential for an accurate diagnosis and outcome prediction.

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Surgical Treatment of Familial Dacryocystocele and Lacrimal Puncta Agenesis

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Abstract: **Bilateral agenesis of the lacrimal puncta and enlargement of the nasolacrimal canal are rare anatomic variants. The authors present 2 familial cases: a 39-year-old woman with bilateral dacryocystocele and lacrimal puncta agenesis and her 46-year-old brother, who had a long history of epiphora and recurrent dacryocystitis, and also had bilateral lacrimal puncta agenesis and a left dacryocystocoele. The authors report the endoscopic, CT, and MRI findings, and describe the surgical treatments, by endoscopic dacryocystorhinostomy in the first case, and conjunctivodacryocystorhinostomy with Jones tube in the second case. At 24 months after surgery, both patients' symptoms were improved.**

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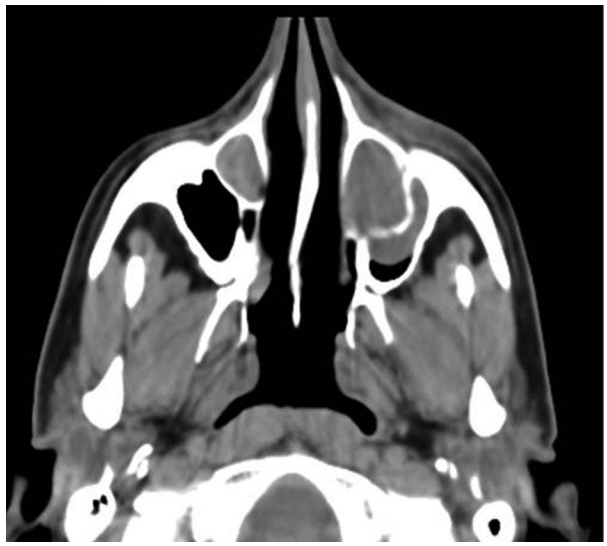

FIG. 1. Case 1, a 39-year-old woman. Preoperative axial CT shows enlargement of both nasolacrimal ducts: 29-mm diameter on the left and 17-mm diameter on the right.

CASE REPORT

We evaluated a 39-year-old woman with a longstanding history of chronic sinusitis symptoms and recurrent left dacryocystitis, without epiphora. Examination revealed bilateral lacrimal puncta agenesis and a left medial canthal mass covered with scar tissue that was caused by previous drainage. Nasal endoscopy showed a left nasal blockage consisting of an intranasal mass with purulent rhinorrea. CT (Fig. 1) showed a bilateral dacryocystocele that was subsequently confirmed by MRI. Under general anesthesia, functional endoscopic sinus surgery was performed, with marsupialization of both dacryocystoceles, opening widely both maxillary sinuses to the nasal cavity. Complete resolution of her symptoms was achieved. At the 24-month follow-up visit, CT (Fig. 2A, B) and nasal endoscopy (Fig. 2C) also showed her improvement.

We also evaluated her 46-year-old brother for recurrent dacryocystitis and left epiphora. His medical history included right-sided epiphora that had resolved after external dacryocystorhinostomy performed during childhood. Nasal endoscopy was normal. CT (Fig. 3A, B) showed a left dacryocystocele with concurrent maxillary mucocele. Examination showed bilateral lacrimal puncta agenesis (Fig. 3C) and a left medial canthal mass. Under general anesthesia, conjunctivodacryocystorhinostomy with Jones tube (19 mm size) was performed. Thereafter, his left epiphora and dacryocystitis were resolved. At the 24-month follow-up visit, his improvement was confirmed on CT.

DISCUSSION

Embryologically, the lacrimal system develops from a thickening of ectoderm within a groove between the lateral nasal process and the adjacent maxillary swelling. The ectoderm invaginates in the underlying mesenchyme and forms an epithelial cord, which progresses caudally. As it develops, the duct is invested by bone during ossification of the maxilla, forming the nasolacrimal canal.[1]

Congenital absence of lacrimal puncta may be an isolated finding or associated with other developmental

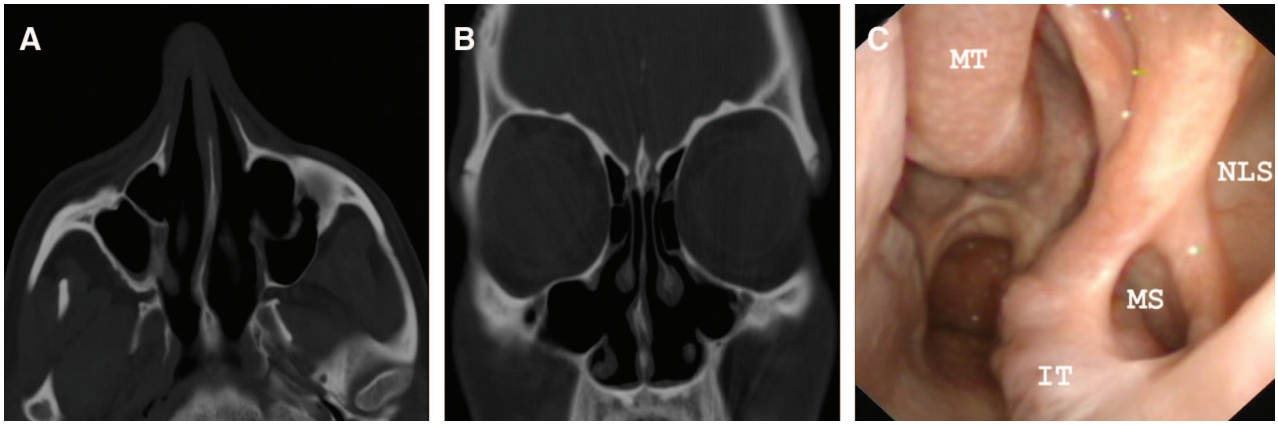

FIG. 2. Case 1. **A,** Axial CT shows marsupialization and enlargement of both dacryocystoceles. **B,** Coronal CT shows bilateral wide middle meatal antrostomy. **C,** During left nasal endoscopy, note the permeability of the nasolacrimal duct and maxillary sinus.

abnormalities.[2] Canalicular or nasolacrimal ducts can also be absent, thus predisposing to the formation of a congenital lacrimal mucocele in rare cases.[2] An enlargement of the nasolacrimal duct, so-called dacryocystocele, is a dilatation of the nasolacrimal duct that occurs after an obstruction of the proximal (valve of Rosenmuller) and distal (valve of Hasner) parts of the lacrimal drainage system.[3] There are few reported cases of enlarged nasolacrimal ducts, with an average diameter of 14 mm in the series by Rheeman and Meyer[4] and 30 mm in the case reported by Schloegel and Sindwani[1] On CT, our first case had a left nasolacrimal duct of 29 mm in diameter and a right nasolacrimal duct of 17 mm, whereas the second case had a duct of 19 mm of diameter.

The pathogenesis of dacryocystocele remains unclear. As reported,[1] it may form as the result of chronic obstruction within the nasolacrimal system (both proximally and distally), which leads to an increased intraluminal pressure enough to cause dilation of the nasolacrimal duct by a bone remodeling process. This chronic enlargement of the lacrimal system results in obstruction of the normal drainage of the maxillary sinus, leading to the development of mucocele and chronic sinusitis, as demonstrated by our cases.

Multidisciplinary surgical treatment was successful in both patients. The first case was treated with functional endoscopic sinus surgery, with marsupialization of both dacryocystoceles and maxillary mucoceles. The second patient underwent conjunctivodacryocystorhinostomy with Jones tube. Both approaches were effective; the first was directed toward relief of chronic rhinorrea and nasal obstruction with absence of epiphora,[5] whereas the second was designed to resolve epiphora in the absence of lacrimal puncta and canaliculi.[6]

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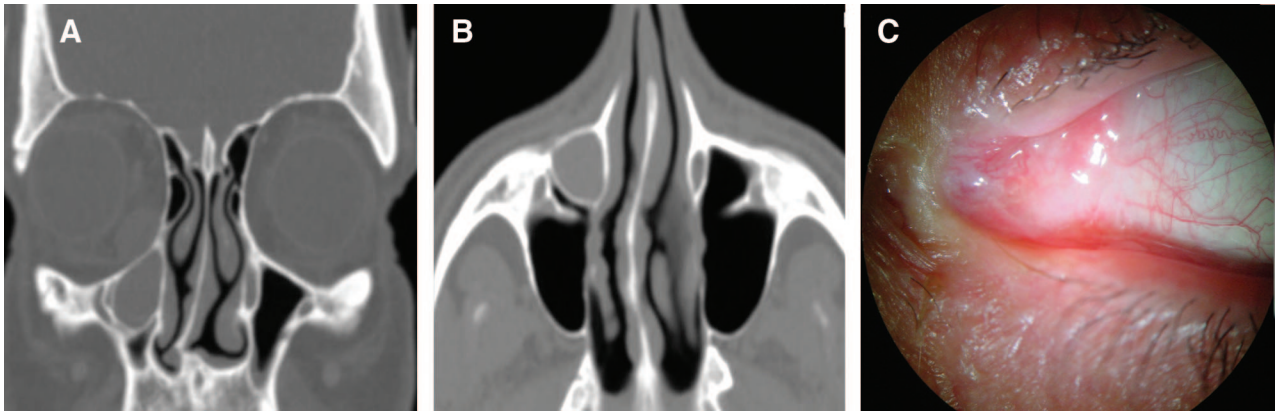

FIG. 3. Case 2, a 46-year-old man. **A,** Coronal CT shows enlargement of the left nasolacrimal duct (19 mm in diameter). **B,** Axial CT shows left maxillary dacryocystocoele. **C,** Left lacrimal puncta agenesis.

Transorbital Puncture of the Cavernous Sinus to Treat a Dural Carotid-Cavernous Sinus Fistula

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*and Alan A. McNab, M.B.B.S., F.R.A.N.Z.C.O.**

Abstract: **A 35-year old woman presented with left ocular injection, proptosis, and reduced vision. Cerebral angiography demonstrated a dural carotid-cavernous fistula supplied by internal and external carotid branches. Following endovascular embolization, the external carotid supply was obliterated. A residual fistula supplied by a branch of the ophthalmic artery and draining by the ophthalmic veins resulted in progressive glaucoma. Compartmentalization of the cavernous sinus precluded access to the fistula via the cerebral venous sinuses. Attempted transarterial embolization via the ophthalmic artery was not achieved. The superior ophthalmic vein was partially thrombosed and an attempt at its surgical isolation was unsuccessful. In this patient, transorbital puncture of the cavernous sinus allowed complete obliteration of the fistula and resulted in prompt resolution of symptoms.**

Transorbital puncture of the cavernous sinus for the treatment of carotid-cavernous sinus fistula (CCF) is rarely reported and, to our knowledge, has not been described in the ophthalmic literature.[1–3]

CASE REPORT

A 34-year-old woman presented with a 2-year history of left proptosis, ocular injection, and reduced vision. Left visual acuity was 20/30, with a left relative afferent pupillary defect, 3 mm of proptosis, dilated episcleral vessels, and venous stasis retinopathy. Intraocular pressures were 14 mm Hg OD and 28 mm Hg OS.

Cerebral angiography demonstrated a left Barrow type D CCF, with cortical venous reflux. Transarterial embolization via an accessory meningeal artery feeder, together with transvenous embolization of the posterior cavernous sinus obliterated external carotid supply and cerebral venous reflux. Transvenous embolization of the anterior cavernous sinus via the inferior petrosal sinuses was not possible because of compartmentalization of the cavernous sinus. A residual fistula, supplied by a branch of the ophthalmic artery and draining by the ophthalmic veins, persisted (Fig. 1A, B). Further endovascular procedures, including attempted transarterial embolization via the ophthalmic artery, were unsuccessful. Surgical isolation of the anteriorly thrombosed superior ophthalmic vein via the upper eyelid was not achieved. Progressive visual field loss resulted from glaucoma refractory to medical management.

Transorbital puncture of the cavernous sinus was performed under general anesthesia. Using road-mapping from internal carotid artery (ICA) angiography and under fluoroscopic guidance, a 3.25-in 16-gauge catheter was advanced via the inferolateral orbit into the cavernous sinus via the superior orbital fissure (Fig. 2). The inner metal stylet was removed, a rotating hemostatic valve attached, and the cavernous sinus catheterized with a renegade microcatheter (Boston Scientific, Boston, MA, U.S.A.) and guidewire. Fibred platinum coils (GDC, Boston Scientific) were placed, with complete fistula obliteration confirmed on ICA angiography (Fig. 3A, B). The catheter was then withdrawn.

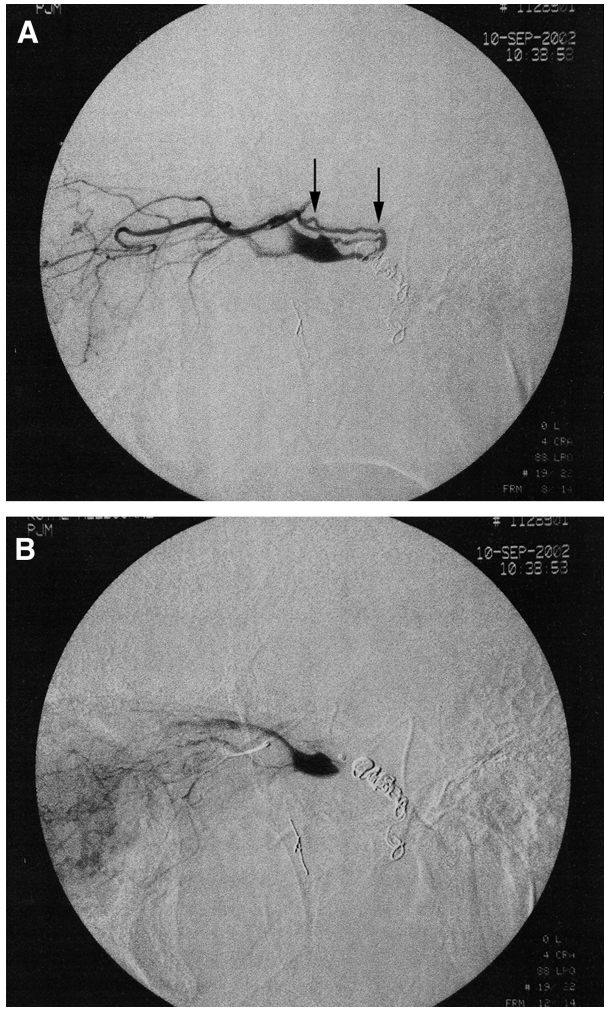

FIG. 1. Lateral view, anterior to left, early **(A)** and late **(B)** phase ophthalmic artery angiogram. The sole arterial supply of the residual fistula, following transarterial coil embolization, is the proximal recurrent meningeal branch of the ophthalmic artery (arrows) arising at an acute angle 2 mm from the internal carotid artery. The late phase shows the fistula draining anteriorly in the ophthalmic veins. The orbital veins are severely congested but only the deep superior ophthalmic vein is of significant caliber.

No procedural complications occurred. Visual acuity improved to 20/20 with complete resolution of ocular symptoms and signs. These findings were maintained over 4 years' follow-up.

DISCUSSION

Endovascular embolization is the established treatment for dural CCF. Through transfemoral venous access, the cavernous sinus can be approached via the cerebral venous sinuses, the superior ophthalmic vein, and the pterygoid plexus. Alternatively,

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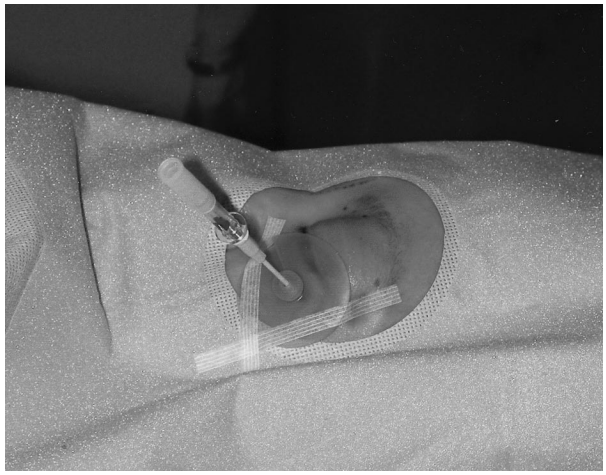

FIG. 2. Catheter (Angiocath, Becton Dickinson Infusion Therapy Systems, Inc., UT, U.S.A.) maintained in position using steristrips and a retention disk (Molnar, Cook Australia, Queensland, Australia), just prior to removing the inner metal needle. The inferolateral orbital puncture site is demonstrated.

direct cannulation of facial veins (facial, angular, superficial temporal, or frontal vein) may provide access. Access may not be possible when veins are narrow, tortuous, or thrombosed, or when the cavernous sinus is compartmentalized. Surgical isolation of the superior ophthalmic vein in the anterior orbit is a well-described alternative. Accessing CCFs via surgically isolated dilated inferior ophthalmic veins has also been reported.[4] When transvenous and transarterial approaches are combined, most CCFs can be occluded with endovascular therapy.

Transorbital puncture of the cavernous sinus to treat CCF was first described through an anophthalmic orbit in 1979.[1] Teng et al.[2] reported this approach to obliterate direct CCFs in 11 patients, 10 of whom had prior occlusion of the ipsilateral ICA. The only complication observed was temporary ptosis in 2 patients. A series of 8 patients in whom this approach allowed obliteration of dural CCF, with a single procedure and without complication, has recently been reported.[3]

An alternative option in our case was a craniotomy with endovascular embolization or direct exposure of the cavernous sinus. Krisht et al.[5] described a pretemporal, extradural approach to the superior orbital fissure with cannulation of the orbital venous drainage channels. A lateral orbitotomy to access the deep superior ophthalmic vein has been described.[6] These approaches allow direct visualization of the puncture site but are more invasive.

Potential complications of transorbital puncture include orbital hemorrhage, globe perforation, ICA puncture (thereby causing a direct CCF), and trauma to cranial nerves II to VI. Orbital hemorrhage has not occurred in the 20 patients reported using this approach and is potentially treatable in the presence of an orbital surgeon. The risks of globe injury should be comparable with peribulbar injections. With modern imaging guidance, avoiding the optic canal and ICA is technically feasible.

In cases of dural CCF where standard endovascular treatment has failed and orbital venous channels are not sufficiently dilated to allow their surgical isolation in the anterior orbit, transorbital puncture is a useful approach to the anterior cavernous sinus and avoids the potential complications associated with a craniotomy or deep orbital dissection.

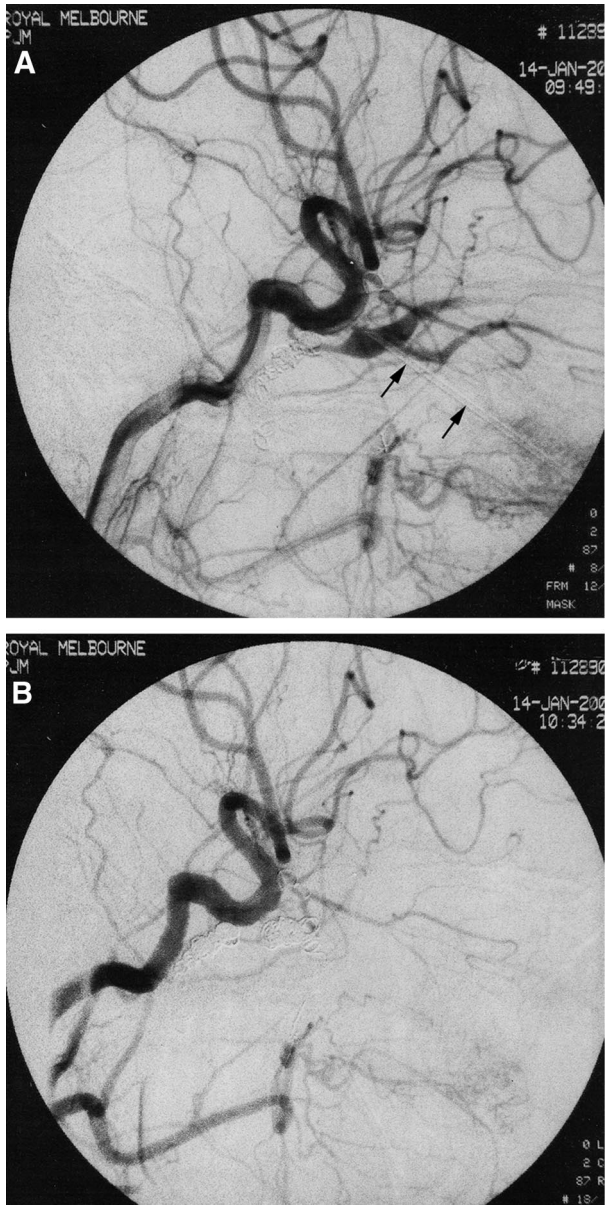

FIG. 3. **A,** Lateral view common carotid artery angiogram showing the catheter tip in relation to the cavernous sinus, superior ophthalmic vein and internal carotid artery. The arrows indicate the line of the catheter. **B,** After delivery of 4 fiber platinum coils, the fistula no longer opacifies.

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Pituitary Macroadenoma Presenting With Brow Mass and Acromegaly

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Abstract: **A 62-year-old woman presented with a brow mass and acromegaly. Biopsy of the lesion showed fibrous dysplasia. Radiologic imaging revealed a pituitary macroadenoma. The patient underwent transsphenoidal pituitary resection and radiation treatment, followed by medicalmanagement of endocrinopathy.**

Pituitary tumors account for 10% to 15% of all primary intracranial tumors.[1] They are usually associated with symptoms due to local compression or hormonal hypersecretion. Typically women are affected more frequently than men. Growth hormone (GH)-secreting tumors represent about 30% of secreting pituitary tumors, of which 75% are macroadenomas.[1] Presentation of pituitary tumors to ophthalmologists is usually related to visual field loss from compression of the optic chiasm. Other associated symptoms secondary to hormonal hypersecretion, such as galactorrhea, infertility, or loss of libido, are often revealed from a detailed history.

We present an atypical case of a patient with a pituitary macroadenoma who presented with a brow mass and acromegaly.

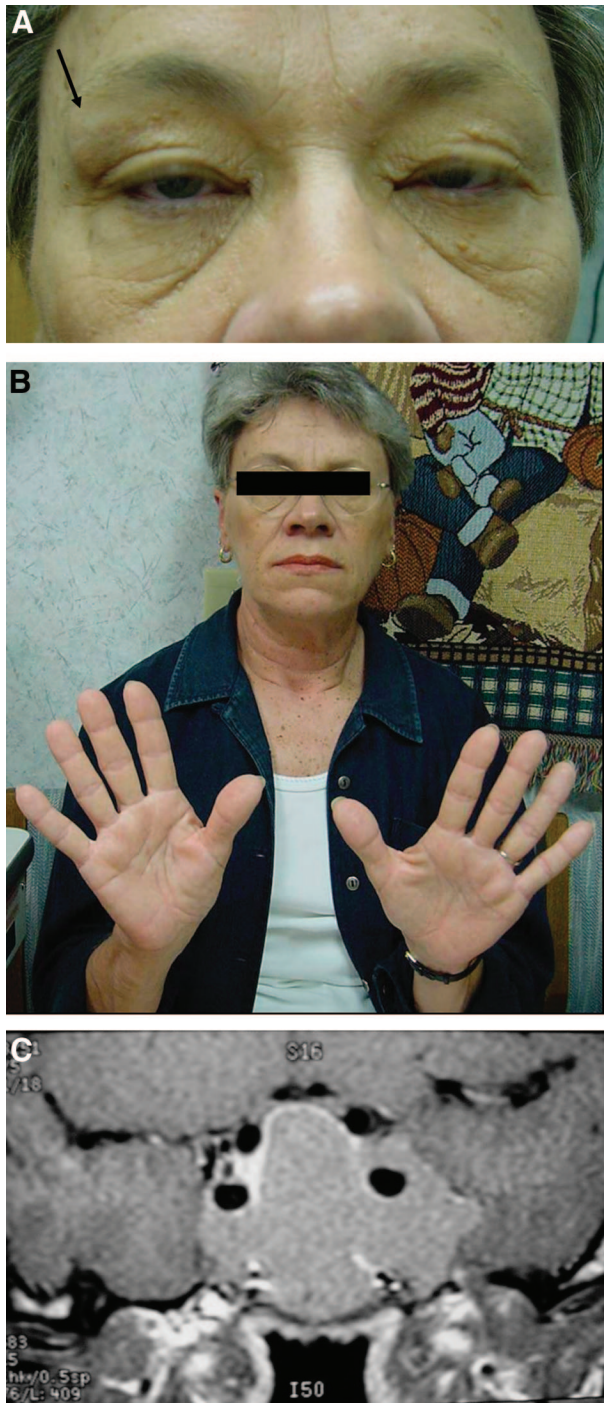
CASE REPORT

A 62-year-old white woman was referred by her general ophthalmologist for evaluation of a right brow mass. The patient had first noticed the lesion ~2 years prior to presentation. The mass was not painful or inflamed, nor had it significantly changed in size over time. Her medical history was significant for thyroidectomy due to goiter, and she was taking thyroid hormone replacement medication. On review of systems, she noted enlargement of her feet from a size 10 to a size 11 over the past 2 years. She also noticed that her rings no longer fit on her fingers. She had no family history of cancer.

Her best-corrected visual acuity was 20/25 OU. Her pupillary examination, confrontational visual fields, and extraocular motility were grossly normal. Slit-lamp examination was also unremarkable. There was a small nodular mass located on the right superolateral orbital rim just inferior to the tail of the eyebrow that was firm and nonmobile (Fig. A). She also had enlarged hands and feet, consistentwith acromegaly (Fig. B).

A biopsy of the brow lesion was performed using an eyelid crease incision. The mass was isolated at the superolateral orbital rim and was discovered to be a bony lesion. The periosteum was incised, and a chisel and mallet were used to shave off a piece for biopsy, which subsequently revealed a diagnosis of fibrous dysplasia. CT and MRI showed a pituitary macroadenoma measuring 5 × 5 × 3.5 cm that involved the entire clivus and invaded both cavernous sinuses and Meckel caves (Fig. C).

The patient was referred for neurosurgical and endocrinologic evaluation. Laboratory values revealed elevated levels of GH at 162 ng/ml (normal, 0–5) and insulin-like growth



A, Right lateral brow mass (arrow). **B,** Acromegaly with enlargement of hands, nose, and jaw. **C,** MRI with gadolinium demonstrates pituitary macroadenoma.

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factor-1 at 900 ng/ml (normal, 71–290). Systemic workup did not reveal other sites of fibrous dysplasia. The patient underwent transsphenoidal debulking of the pituitary macroadenoma, which resulted in about 75% of the tumor being removed. She subsequently received 4,500 cGy of fractionated stereotactic radiation therapy administered over a 5-week course. Medical treatment with Sandostatin (octreotide) was also given to suppress residual elevation of GH and insulin-like growth factor-1. However, despite maximal dosage of Sandostatin, she had persistent elevation of GH and insulin-like growth factor-1. She was then switched to Somavert (pegvisomant), a GH-receptor antagonist, and her GH and insulin-like growth factor-1 levels fell dramatically. Her acromegaly improved and the patient continues to be followed.

DISCUSSION

Pituitary macroadenoma presenting with a brow mass due to fibrous dysplasia and acromegaly has not been previously described, to our knowledge. Histopathologic examination by 2 pathologists from different institutions confirmed this brow mass to be fibrous dysplasia.

It is possible that this fibrous dysplasia lesion occurred independently of the GH-secreting pituitary adenoma, and that the hormonal changes from the pituitary adenoma caused the preexisting fibrous dysplasia lesion to grow. However, the combination of fibrous dysplasia, pituitary adenoma, and hypersecretory endocrinopathy in this patient may constitute this case as an atypical variant of McCune-Albright syndrome (MAS).

Classically, MAS presents in a child with precocious puberty, café-au-lait spots, and fibrous dysplasia. Our patient is obviously much older than the typical MAS patient and did not present with a history of precocious puberty or café-au-lait spots. However, Bhansali et al.[2,3] describe 2 patients who presented in their third decade with the diagnosis of MAS without precocious puberty or skin lesions. They also cite numerous other references in their review of the literature of MAS patients who presented in adulthood, even as late as the fifth decade, many of whom presented with fibrous dysplasia and some form of hypersecretory endocrinopathy. In their discussion, they state, "Two of the following lesions, namely poly/monostotic fibrous dysplasia, cutaneous pigmented macules and hypersecretory endocrinopathies are suffice to make the diagnosis of MAS." Our patient had both fibrous dysplasia and a hypersecretory endocrinopathy, which may constitute this as an atypical case of MAS.

Acromegaly is a rare pituitary disease caused by a GH-secreting tumor. It can be easily diagnosed on physical examination, however, one must recognize it when seeing it. Serious cardiac complications can arise from acromegaly, making its diagnosis even more critical. Excessive serum levels of GH over time induce cardiomyopathy, which can lead to arrhythmias, valvular dysfunction, and heart failure.[4]

In summary, we present an unusual case of a pituitary macroadenoma that presented to an ophthalmologist with a brow mass secondary to fibrous dysplasia and acromegaly. It is important for ophthalmologists to remember to obtain a thorough history and conduct a full physical examination to recognize acromegaly and other signs of systemic disease.

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Ectopic Meningioma Anterior to the Lacrimal Gland Fossa

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Abstract: **A 66-year-old man reported a slowly growing tumor on the lateral edge of his left upper eyelid. This lesion was hard but movable on palpation. A neoplasm of the lacrimal gland was suspected. CT showed a highly calcified lesion at the left upper eyelid. Resection of the tumor was performed, which was located just behind the orbital septum and in front of the lacrimal gland. Anatomopathologic investigation of the excised specimen with immunohistochemistry revealed a benign meningioma of a meningotheliomatous type, containing multiple bone elements. An ectopic orbital meningioma is rare, and this is the first case of a unique lateral localization of this lesion. Therefore, it should be included in the differential diagnosis of a lacrimal gland tumor.**

Primary and secondary meningiomas of the orbit arise from arachnoid cells of the optic nerve and cranial dura, respectively. Both are relatively uncommon tumors.[1] Apart from these are ectopic orbital meningiomas. These very rare tumors are neither connected to nor arise from the optic nerve sheath, and are not secondary extensions from the skull base.

CASE REPORT

A 66-year-old man was referred with a growing orbital mass protruding in his left upper eyelid during the past 4 years (Fig. 1A). Medical history was unremarkable except for head trauma 40 years before without radiologic evidence of a skull fracture. Visual acuity was 20/20 OU. Slit lamp, fundoscopy, and ocular motility were normal. A firm mass not adherent to bone or skin was palpable trough the left upper eyelid. CT of the orbit showed an extensively calcified mass located at the anterior edge of the lacrimal fossa without hyperostosis or involvement of the adjacent orbital bone (Fig. 1B, C). Paranasal sinuses were normal and symmetric.

A translid surgical approach revealed a well-circumscribed hard, whitish mass within the preaponeurotic fat just anterior to the orbital part of the lacrimal gland. The tumor was excised under frozen section control and measured 12 × 10 ×

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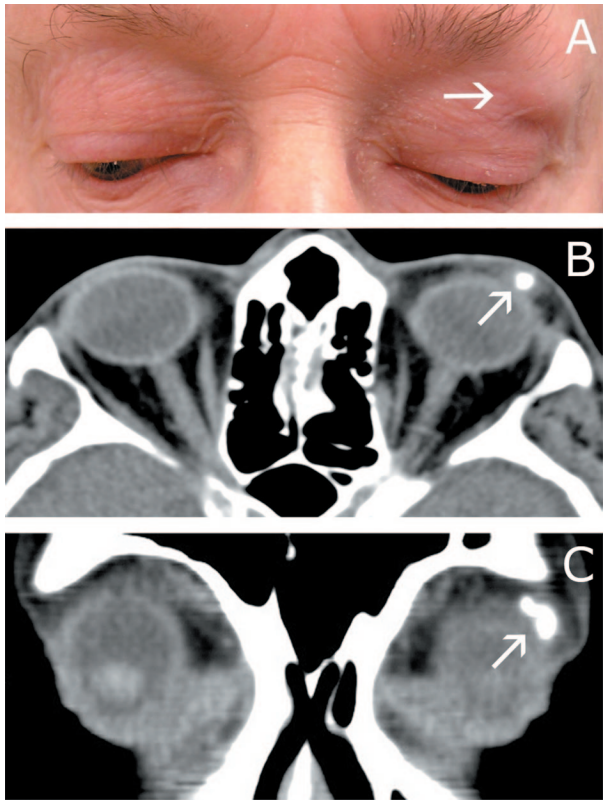

FIG. 1. **A,** Clinical appearance of tumor protruding through left upper eyelid (arrow). **B,** Axial CT shows a calcified mass (arrow) at the lateral edge of left eye without signs of bone or skin infiltration. **C,** Coronal view shows tumor (arrow).

7 mm. Microscopy revealed it to consist of cells displaying a meningothelial morphology and a syncytial growth pattern with nuclei showing little pleomorphism (Fig. 2A, B). Dispersed in the tumor were areas of osseous metaplasia (Fig. 2A, D). Immunohistochemistry with epithelial membrane antigen and vimentin positivity confirmed the meningothelial nature of the cells (Fig. 2C). Other epithelial markers including cytokeratin were negative, excluding an adenoma. Ki 67 immunostaining was performed to assess the proliferation rate of the lesion, with a labeling index of 5%. During a follow-up of 15 months no recurrence was noted.

DISCUSSION

Ectopic orbital meningiomas are rare. Farah et al.[2] stated that only 5 published cases of these tumors had sufficient clinical, radiologic, and histologic evidence to support their diagnosis. Patient age ranged from 7 to 66 years.[2–5] Our patient is the oldest person ever reported to have such a lesion. All previously reported cases were located along the medial orbital wall, whereas the tumor in this case was localized at the anterior edge of the lacrimal fossa mimicking a lacrimal gland tumor.

Many theories have been proposed regarding the origin of ectopic orbital meningiomas. Tan and Lim[4] proposed 3 possible ways: 1) from occasional arachnoid "nests" in the orbit; 2) from the optic nerve sheath with loss of connection before discovery; and 3) from smaller nerves endowed with arachnoid cells. In our case, neither small nerves were identified histologically, nor was the tumor attached to the periosteum. Marquardt and Zimmerman[6] suggested ectopic orbital meningiomas should be considered as an extension of intracranial meninges. Finally, Farah et al.[2] suggested

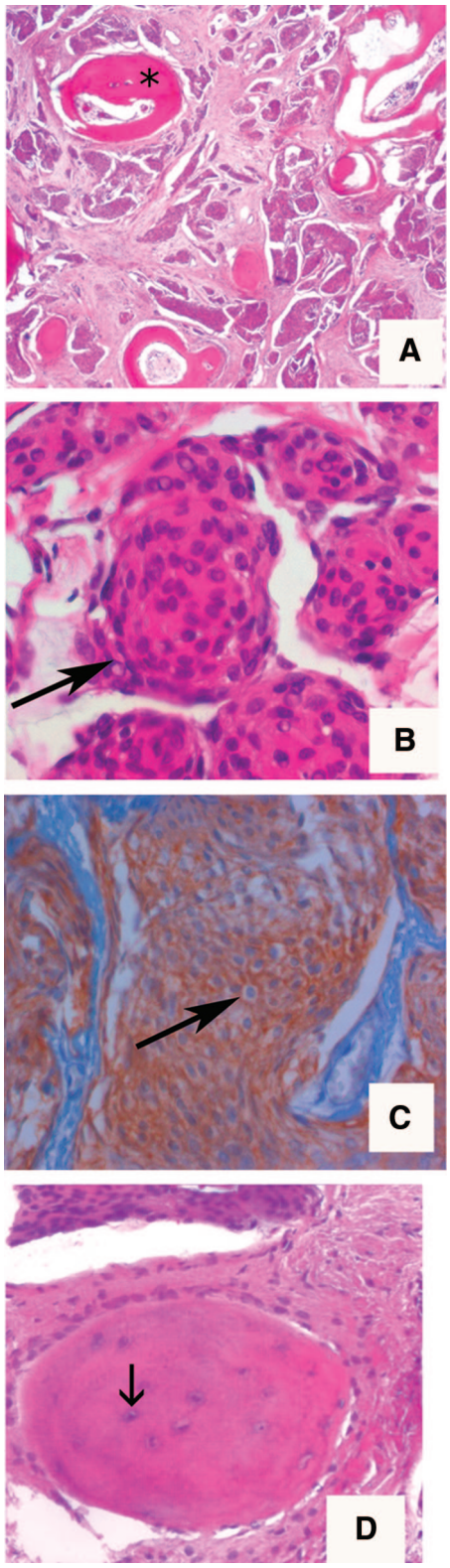

FIG. 2. **A,** Hematoxylin-eosin staining of the specimen shows meningotheliomatous meningioma and areas of osseous metaplasia (asterisk) (×20). **B,** High magnification of the tumoral cell aggregates (arrow) (hematoxylin-eosin, ×400). **C,** High magnification epithelial membrane antigen stain of tumoral cells (arrow) (×400). **D,** Magnification of osseous metaplasia (arrow) characterizing ectopic meningioma (×200).

an association with paranasal sinus asymmetry and head trauma. Paranasal sinus asymmetry was found in 4 of the 5 previously reported cases, although not in our case. Three of 6 patients (including ours) had a history of head trauma.[2,3] Of the other 3, one patient had a history of traumatic cataract.[5] In 1938, Cushing and Eisenhardt[7] stated that meningiomas can be caused by head trauma with resultant meningeal irritation. As such, skull fracture with trapping of extracranial meningeal tissue followed by its cicatricial process, has also been proposed as etiology for cutaneous meningioma of the face.[8]

Given the distant location from the optic nerve, the theory of evolutionary migration from the optic nerve sheath seems rather unlikely. The most plausible etiologic factor to explain a possible implantation of cells from the intracranial cavity in this case was the history of head trauma without evidence of a skull fracture.

In conclusion, ectopic orbital meningiomas are extremely rare. All previously reported cases were located near the medial orbital wall. This case of ectopic orbital meningioma is not only in the oldest person ever reported, but also represents the only tumor located laterally in the orbit. The remote location away from the optic nerve suggests an origin from ectopic arachnoidal cells triggered by head trauma.

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Orbital Leiomyoma: Histopathologic and Immunohistochemical Findings of a Rare Tumor

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Abstract: **A 51-year-old man was referred for evaluation of a right orbital hemangioma. Ophthalmologic examination was unremarkable except for 1 mm of proptosis OD. CT revealed a 10-mm lesion with evidence of growth from 6 to 10 mm within a year. The tumor did not compromise other orbital structures. An excisional biopsy was performed. On the basis of the histopathologic and immunohistochemical findings, the diagnosis of orbital leiomyoma was established. Orbital leiomyoma is a slow-growing tumor that can be located anywhere in the orbit. Posterior tumors are believed to originate from smooth muscle cells of vessel walls; anterior lesions may arise from the capsulopalpebral or Müller muscle. Although there are no unique features that help the radiologist to exclude other benign lesions of the orbit, the histopathologic diagnosis using immunohistochemical markers is usually straightforward. Attention to the cytologic features that exclude the malignant variant is of utmost relevance for proper diagnosis and patient counseling.**

Leiomyoma is a benign spindle-cell tumor that rarely involves the orbit. The tumor is presumably derived from the muscular layer of blood vessels.[1] Arat et al.[2] recently reviewed 26 cases of orbital leiomyoma, although only 8 were confirmed by immunohistochemistry, including 4 cases presented by these authors. The most common clinical presentation is a painless, slowly progressing proptosis. We report a patient who presented with conjunctival hemorrhage that prompted a clinical investigation that ultimately disclosed the orbital tumor.

CASE REPORT

A 51-year-old man was referred for evaluation of an orbital hemangioma OD. His medical history was relevant for an episode of spontaneous, painless temporal subconjunctival hemorrhage, and inferior eyelid ecchymosis 1 year before. CT performed at that time revealed a 6-mm, well-defined mass in the inferotemporal right orbit, just posterior to the globe. Ophthalmologic examination was unremarkable except for 1 mm of proptosis OD. CT was repeated and the lesion was again observed with clear evidence of growth; now, 1 year later, it measured 10 mm in diameter (Fig. 1A). The tumor did not compromise other orbital structures. An excisional biopsy was performed.

Histopathologically, the tumor was encapsulated, composed of spindle-shaped, benign-appearing cells arranged in fascicles (Fig. 1B). Multiple thin-walled vessels could also be seen interspersed with the neoplastic cells (Fig. 1C). The cells had cigar-shaped nuclei with blunt edges and eosinophilic cytoplasm (Fig. 1D). Some microscopic areas of hyalinized eosinophilic collagen matrix were seen interspersed between dead and preserved cells, a pattern consistent with hyaline necrosis (Fig. 2A). Other focal areas showed increased cellularity with mild degree of nuclear pleomorphism (Fig. 2B). Although larger, mildly atypical cells showed minimal variation in nuclear size and shape, and nucleoli were small. A single mitotic figure was seen, despite evaluation of several levels of the specimen (<1 mitosis/50 high power fields). The tumor stained red with Masson trichrome, which also highlighted the connective tissue of the capsule in blue (Fig. 3A). Reticulin was present around each individual cell, a pattern previously described in this particular entity (Fig. 3B).[1] Immunohistochemistry was positive for smooth muscle actin (Fig. 3C), desmin (Fig. 3D), vimentin, and muscle-specific actin. CD68, HMB-45, S-100, and cytokeratins were all negative. CD34 was positive in the vascular component of the tumor. On the basis of the aforementioned findings, the diagnosis of orbital leiomyoma was established. After 10 months of follow-up, no recurrence was seen.

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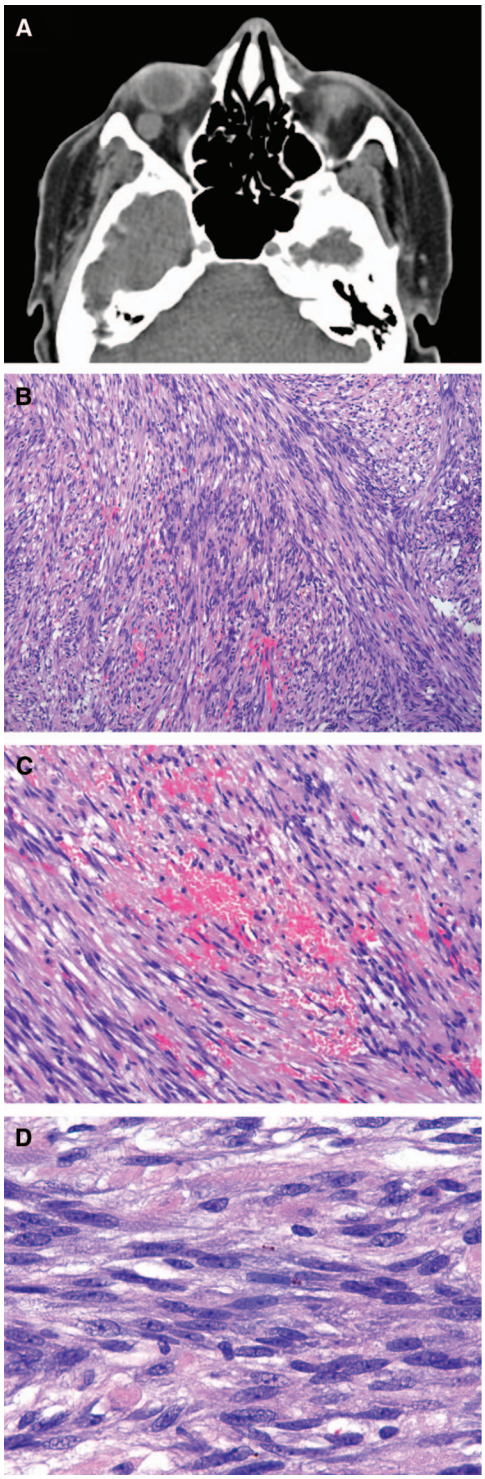

FIG. 1. **A,** CT reveals a 10-mm well-defined lesion in the inferotemporal orbit, just posterior to the globe. **B,** The tumor was composed of spindle cells arranged in fascicles (hematoxylin-eosin, original magnification ×100). **C,** Numerous thin-walled vessels were seen interspersed with the neoplastic cells (hematoxylin-eosin, original magnification ×400). **D,** The cigar-shaped nuclei, with blunt ends, characteristic of leiomyoma (hematoxylin-eosin, original magnification ×640).

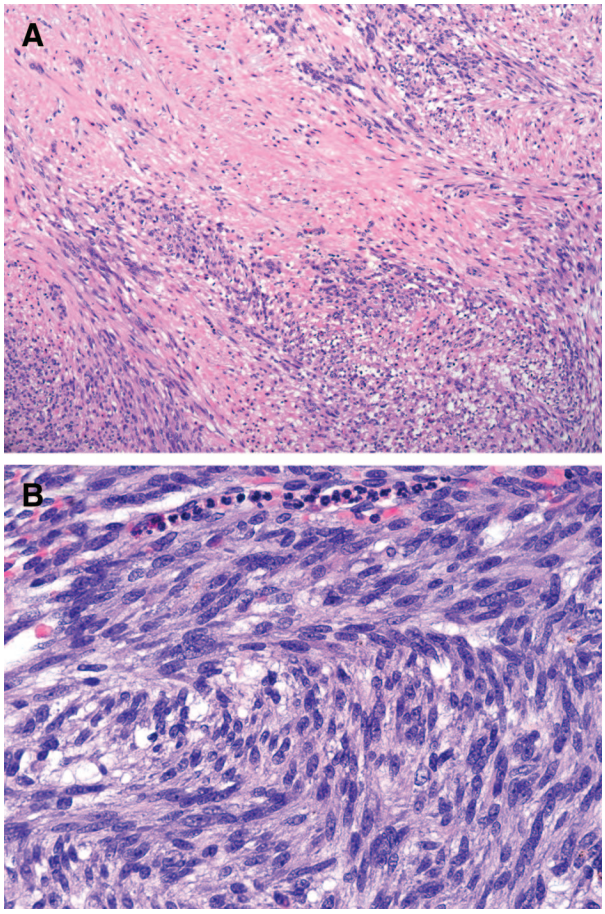

FIG. 2. **A,** Hyalinized eosinophilic collagen matrix interspersed between dead and preserved cells, a pattern consistent with hyaline necrosis (hematoxylin-eosin, original magnification ×100). **B,** Area showing increased cellularity with a mild degree of nuclear pleomorphism. Larger, mildly atypical cells show minimal variation in nuclear size and shape, and nucleoli are small (hematoxylin-eosin, original magnification ×400).

DISCUSSION

Orbital leiomyoma is a slow-growing tumor that can be located anywhere in the orbit. Posterior tumors are believed to originate from smooth muscle cells of vessel walls; anterior lesions may arise from the capsulopalpebral or Müller muscle.[1,2] The most common presentation of the disease is a slowly progressive proptosis, without inflammation or pain.[2] The tumor can disturb extraocular motility in some cases. Imaging studies are useful to determine the extension of the lesion. However, there are no unique features that help the radiologist to exclude other benign lesions of the orbit.[3]

Complete excision is the treatment of choice. If the tumor cannot be completely removed, close follow-up of the patient is recommended. Radiation is discouraged not only because the tumor is radioresistant but also for the possibility of sarcomatous transformation.[2] Recently, an orbital leiomyoma was treated with goserelin, a GnRH analog, after histopathologic confirmation.[4] The tumor regressed completely, without recurrence during a 5-year follow-up period.

The histopathologic diagnosis using immunohistochemical markers is usually straightforward. The absence of nuclear pleo-

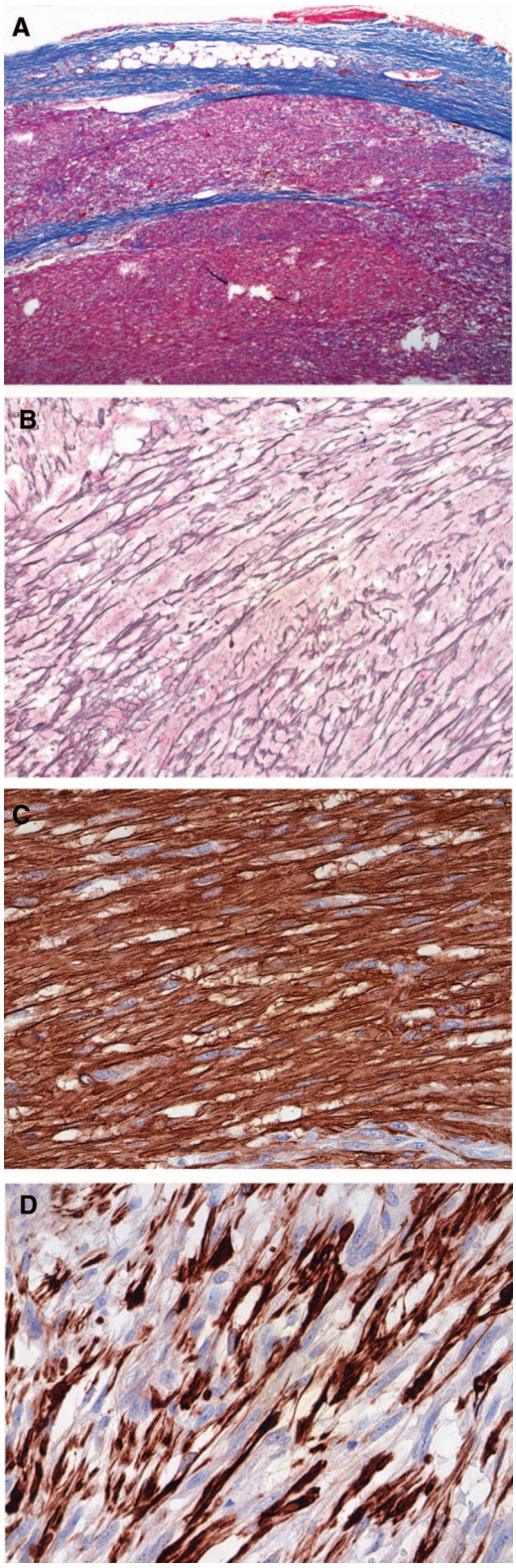

FIG. 3. **A,** Masson trichrome stained the tumor red, and the capsule blue (original magnification ×50). **B,** The reticulin deposition around individual cells is highlighted by special stain (original magnification ×200). **C,** Diffuse and strong positivity for smooth muscle actin is seen (original magnification ×400). **D,** Desmin was also positive in the neoplastic cells (original magnification ×400).

morphism, hyperchromatism, giant cells, or mitotic figures helps the pathologist to distinguish leiomyoma from leiomyosarcoma, its malignant counterpart.[5] However, in some cases such distinction is not so clear. A recent review proposed that the biologically benign group of smooth-muscle tumors can be defined by stringent histopathologic criteria: lack of nuclear atypia (moderate or marked) and coagulative necrosis and low mitotic count (<1 or 1–4 mitoses/50 High Power Fields).[6] In our case, the tumor was deemed benign after considering the slow growth rate, the presence of a well-defined capsule, a low mitotic count, and the absence of significant cellular atypia and coagulative necrosis.

To our knowledge, bleeding is an unreported presentation of an orbital leiomyoma. Because conjunctival bleeding is such a common event, we cannot rule out the possibility that the bleeding was unrelated to the tumor, the latter being only an incidental finding. However, considering the number of thin-walled vessels that were seen within the tumor, it is plausible to imagine that some of these lesions may bleed. As some orbital leiomyomas are located in the posterior orbit, the bleeding may not have been evident in previous cases. Another possible explanation would be the compression of orbital veins by the mass with secondary pressure increase in conjunctival and eyelid capillaries.

In summary, we presented a case of orbital leiomyoma emphasizing histopathologic features. Attention to the cytologic features that exclude the malignant variant is of utmost relevance for proper diagnosis and patient counseling.

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Ewing Sarcoma Presenting as a Subconjunctival Mass

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Abstract: **A 14-year-old previously healthy boy presented for evaluation of a subconjunctival mass that had been present for 2 to 3 years, but had recently started to enlarge. Imaging**

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demonstrated an anterior orbital lesion that enhanced with gadolinium. An excisional biopsy was performed and revealed highly malignant-appearing small, blue, round tumor cells. Immuohistochemical stains were strongly reactive with CD99 (MIC2 and O13), CD56, and retained INI-1 reactivity. The EWS-FLI1 chimeric fusion gene transcript was also detected by reverse transcription-polymerase chain reaction analysis, confirming Ewing sarcoma. Full-body CT, bone scan, and bone marrow biopsy were all negative, proving this to be a premetastatic, primary orbital tumor arising from soft tissue.

Extraskeletal Ewing sarcoma (EES) is a rare and highly malignant neoplasm that is part of the Ewing sarcoma family of tumors (ESFT). Both clinically and pathologically indistinguishable from Ewing sarcoma of bone, EES can arise in the soft tissues at any location, and is included in the differential of small, blue, round cell tumors.[1,2] While EES is most often seen in the trunk, extremities, and retroperitoneum,[1,2] the head and neck are rare primary sites for this neoplasm.[1–3] There has been only one case of primary orbital involvement reported in the literature.[4] We report a second case of primary orbital EES, presenting as a vascular subconjunctival mass.

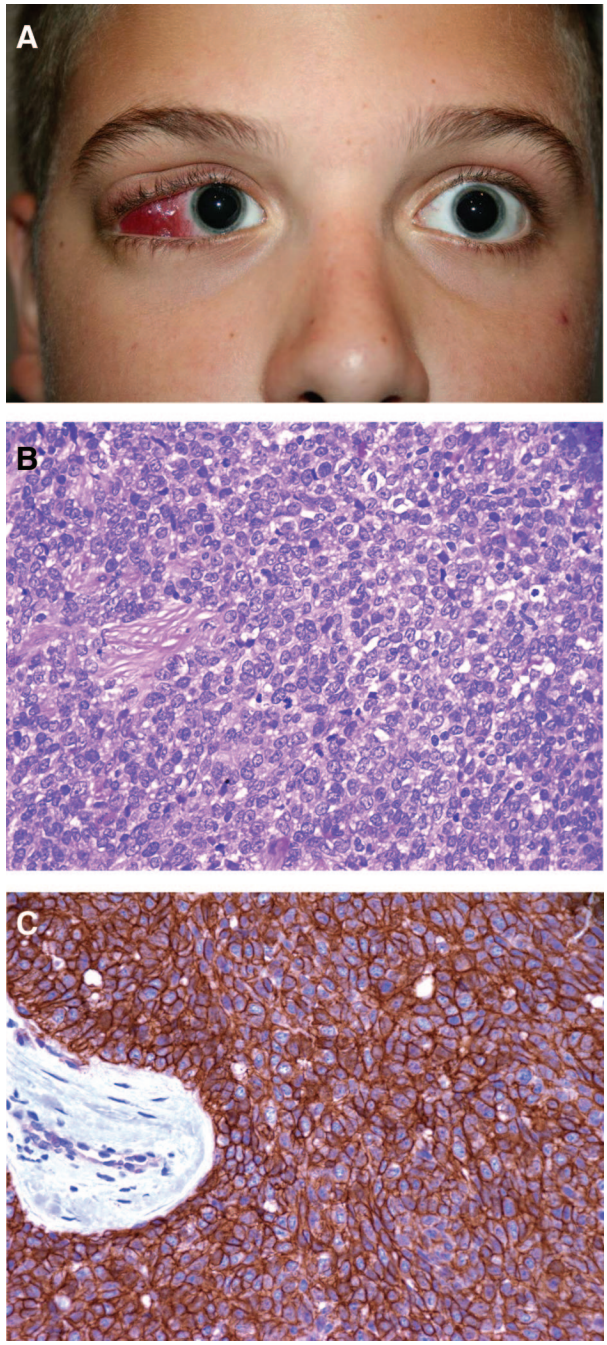
CASE REPORT

A pediatrician referred a healthy 14-year-old boy for evaluation of a painless subconjunctival mass in his right eye which, by history, had been present for 2 to 3 years, but had recently begun to enlarge. Examination revealed a firm, vascular-appearing mass in the temporal fornix of the right eye, measuring 17 mm × 15 mm (Fig. A). Visual acuity was 20/20 OU, motility was full, and the patient did not complain of diplopia. MRI was limited because of orthodontic hardware, but it showed an anterior orbital lesion that enhanced with gadolinium. The patient underwent an excisional biopsy 3 weeks after initial evaluation, at which time an interim increase in the mass size was noted (23 mm × 17 mm). The mass was dissected from the overlying conjunctiva and peeled free of the scleral surface and lateral rectus muscle; lacrimal gland was thought to be uninvolved.

Histopathology revealed cells with scant eosinophilic cytoplasm, prominent nucleoli, and frequent mitotic figures (Fig. B). No pseudorosettes or neurosecretory granules were noted. The cells stained weakly for vimentin and strongly for CD99 (MIC 2 and O-13) (Fig. C) and CD56, and were nonreactive with S-100, desmin, synaptophysin, and all B- and T-cell lymphocyte and myeloid markers. Fluorescence in situ hybridization (FISH) analysis revealed the EWS/FLI-1 translocation, which is diagnostic for the Ewing family of neoplasms.

The patient underwent systemic evaluation, including a full-body bone scan and CT, and repeat MRI after removal of his oral braces. He also underwent a bone marrow biopsy. All test results were normal except for residual MRI enhancement posterior to the right lacrimal gland. On the basis of these results, the patient was entered in a treatment protocol that included 14 cycles of vincristine, cytoxan, and doxorubicin alternating with ifosfamide and etoposide. After 6 cycles, imaging demonstrated postoperative changes versus residual tumor enhancement posterior to the right lacrimal gland. A multidisciplinary decision was made to forgo further surgery in favor of proton therapy to the area of enhancement.

The patient has been tolerating his treatment well, and has no evidence of recurrence or metastasis in the 10-month follow-up period since treatment was initiated.



A, Clinical photograph. **B,** Orbital biopsy specimen shows cells with scant eosinophilic cytoplasm, prominent nucleoli, and frequent mitotic figures (hematoxylin-eosin, ×640). **C,** Tumor cells demonstrating immunoreactivity of the tumor cells for the O13 antibody, also known as the Ewing/PNET antigen (×640).

DISCUSSION

The ESFT, a group which includes Ewing sarcoma of bone, EES, peripheral primitive neuroectodermal tumor of bone, and Askin tumor, comprises neoplasms of neuroectodermal origin. These tumors are characterized pathologically as small, round, blue cell tumors, cytogenetically by a t(11;22) or t(21;22) translocation, and molecularly by the presence of chimeric transcripts resulting from the fusion of the EWS gene

with genes that encode specific transcription factors.[1–3] Within the ESFT there is a continuum of neuronal differentiation, from minimal evidence (Ewing sarcoma of bone and EES) to more pronounced (peripheral primitive neuroectodermal tumor of bone).[1–3] This degree of neuronal differentiation has been used for histopathologic subclassification; however, because the behavior, prognosis, and treatment seem to be similar for all subsets of ESFT, this histopathologic subclassification may not be clinically significant.[2,3]

Although Ewing sarcoma of bone is recognized as the second most common primary malignancy of bone, 40% of primary ESFT occur in soft tissue. The differential diagnosis of such tumors in the head and neck area, which account for 4% to 18% of all ESFT,[1–3] includes lymphoma, rhabdomyosarcoma, and neuroblastoma. Primary orbital variants arising in soft tissue are even rarer: there are 8 reported cases of peripheral primitive neuroectodermal tumor of bone[5–11] and only one of EES[4] (Table, http://links.lww.com/A649). Interestingly, many of these patients are adults, with primary soft-tissue tumors that arose intraconally or adjacent to an orbital wall. To our knowledge, this is the first reported case of an ESFT presenting as a subconjunctival mass.

The history and clinical presentation of our patient is unusual. As with other malignant orbital tumors, ESFT typically present with rapid onset due to the space constraints of the bony orbit. The history of a subconjunctival mass present for 2 to 3 years seems at odds with the malignant nature of this tumor on biopsy. However, the highly visible location in our patient may offer an unusual opportunity to appreciate the possible pathogenesis of ESFT: the acquisition of specific genetic mutations may be a slow, controlled process occurring over the course of years until, as in our patient, some critical point is reached, and unchecked, rapid growth ensues.

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Primary Cutaneous Anaplastic Large Cell Lymphoma of the Medial Canthus and Orbit

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Abstract: **A 59-year-old man sought evaluation for a rapidly enlarging, nontender, ulcerated right medial canthal lesion unresponsive to antibiotics. Biopsy revealed CD30+ anaplastic large cell lymphoma. CT demonstrated contiguous spread in the orbit. Systemic evaluation for lymphoma was negative, and he underwent local radiotherapy. The lesion regressed completely, and he has remained disease free for 7 months. CD30+ anaplastic large cell lymphoma of the periocular skin and orbit are usually distinct, exceedingly rare entities; no reported cases had simultaneous involvement of both tissues. The authors present the first reported case, to their knowledge, of simultaneous skin and orbital involvement by anaplastic large cell lymphoma.**

Only 1% to 3% of cases of ocular adnexal lymphoma are non-B-cell lymphomas, the majority representing involvement by systemic T-cell lymphoma or progression of mycosis fungoides. Primary cutaneous (C-) anaplastic large cell lymphoma (ALCL) and systemic ALCL, both usually of T-cell origin, rarely involve the periocular skin with only 3 of each entity reported.[1–5] None of these cases of skin involvement showed orbital extension. Orbital ALCL is likewise rare, with only one reported case in which there was direct extension from the paranasal sinuses.[6] We report an exceptionally rare case of ALCL involving the skin and orbit.

CASE REPORT

A 59-year-old man with ulcerative colitis presented with a 12-day-old ulcerated, nontender right medial canthal lesion. Initially appearing as a small wound, the lesion failed to respond to tobramycin/dexamethasone ointment. Progression prompted an unsuccessful 3-day course of intravenous ampicillin/sulbactam. Upon referral to the University of Michigan, a 5-mm ulcerated lesion was present in the superior right medial canthus and 14 mm of surrounding induration extended in the eyelids. Visual acuity was 20/25 OD and 20/20 OS. The ocular examination was otherwise unremarkable. Two days later, the ulcer measured 7 mm with 25 × 18 mm of induration. The differential diagnosis included spider bite, as supported by the patient's history, and cutaneous anthrax from cleaning gutters, but pyoderma gangrenosum in ulcerative colitis was the presumptive diagnosis and prednisone (40 mg/day) was prescribed. A punch biopsy was obtained.

Two weeks later the ulceration and induration measured 14 mm and 33 mm, respectively (Fig. 1). Histopathology of the biopsy revealed ALCL composed of large, malignant CD3+ and CD30+ lymphoid cells that were negative for CD15, CD43, CD56, anaplastic lymphoma kinase 1, and epithelial membrane antigen (Fig. 2). The differential diagnosis of lymphomatoid papulosis was eliminated as other skin lesions were absent. CT showed an infiltrative extraconal mass

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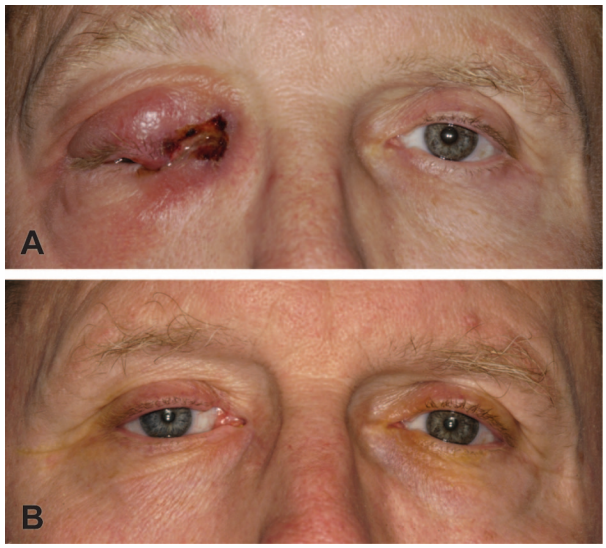

FIG. 1. The patient before **(A)** and after **(B)** radiotherapy.

of the medial orbit extending in the cheek, forehead, and nasal bridge (Fig. 3). Systemic disease was not detected.

The patient underwent radiotherapy with a total dose of 31 Gy. He remains clinically and radiographically disease free after 7 months. Ocular findings at last follow-up were mild right blepharoptosis and epiphora.

DISCUSSION

C-ALCL, systemic ALCL, and lymphomatoid papulosis exhibit similar histopathology with CD30+ large, anaplastic lymphoid cells. Lymphomatoid papulosis differs from ALCL by the presence of recurrent, multiple red-brown papulonodular lesions that regress spontaneously in 3 to 8 weeks. C-ALCL consists of a single, ulcerated lesion that grows rapidly, but does not extend in deep tissues. Lymphomatoid papulosis has a very favorable prognosis; only 4% of patients develop malignant lymphoma.[7] The distinction between C-ALCL and systemic ALCL with secondary skin involvement is difficult, but important. C-ALCL has a better prognosis and shows good response to complete excision or radiotherapy alone, whereas systemic ALCL requires chemotherapy.[8] Histopathologic features do not distinguish C-ALCL from systemic ALCL with secondary cutaneous involvement, but clinical features are helpful. C-ALCL affects older patients (median age, 61 years) than systemic ALCL (median age, 24 years),[8] while a negative systemic evaluation renders the possibility of secondary skin involvement unlikely. It is impossible to determine conclusively whether our patient's tumor was a C-ALCL with secondary orbital invasion, or a systemic ALCL of the orbit with secondary cutaneous spread. If it was systemic, our case would be the first reported case isolated to the periocular region.

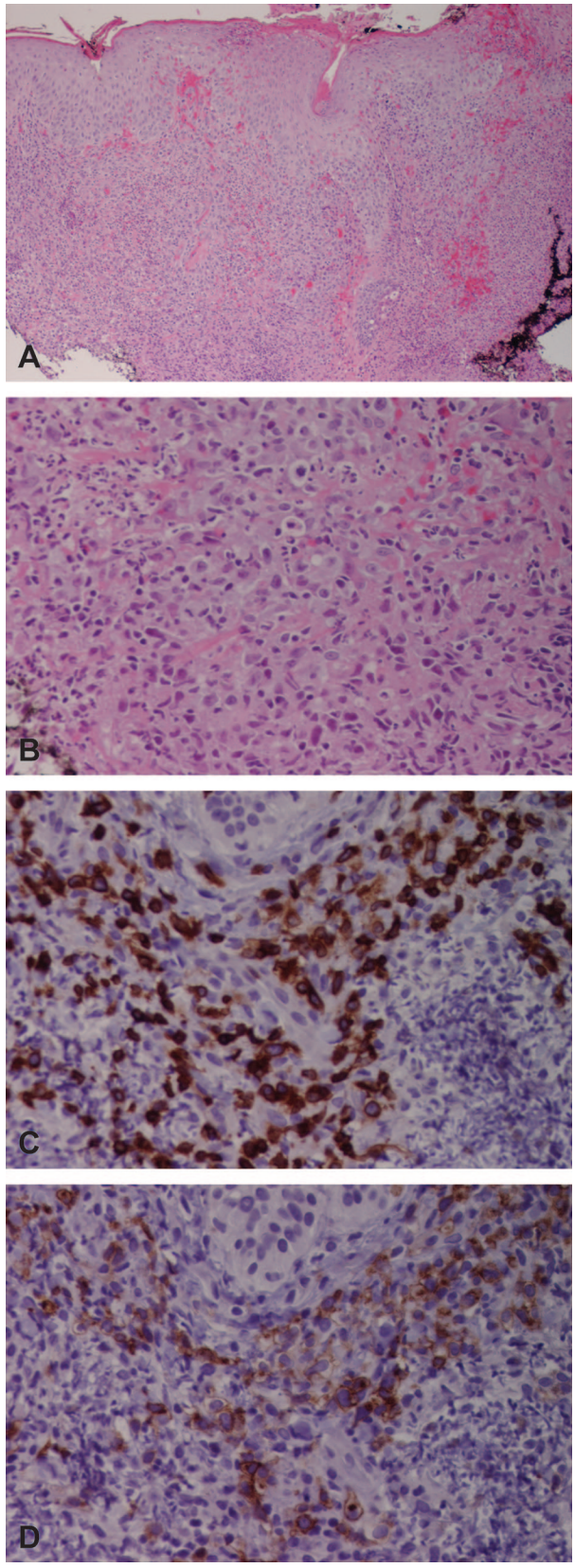

FIG. 2.

FIG. 2. **A,** Biopsy showing dense dermal infiltrate with overlying reactive epithelial hyperplasia (hematoxylin-eosin, ×100). **B,** Large, atypical lymphocytes interspersed with small benign lymphocytes; the malignant lymphocytes contain vesicular, often convoluted nuclei, and prominent nucleoli (hematoxylin-eosin, ×400). **C,** The lymphoma cells stain intensely for CD3 (immunoperoxidase with hematoxylin counterstain, ×400. **D,** Lymphoma cells demonstrate intense perinuclear and cell membrane immunoreactivity for CD30 (immunoperoxidase with hematoxylin counterstain, ×400).

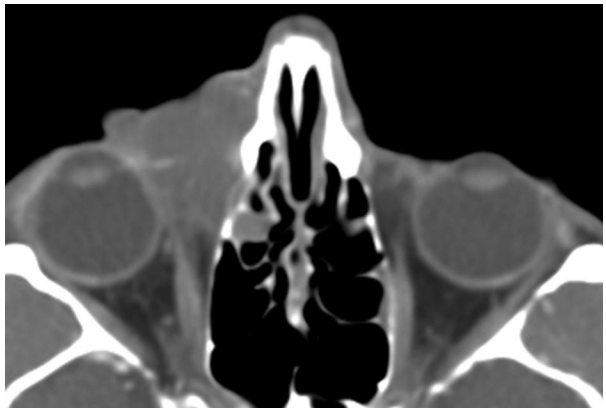

FIG. 3. Orbital CT shows an ill-defined right medial canthal mass with infiltration of anterior orbital tissues.

We favor the diagnosis of C-ALCL due to tumor location, typical clinical presentation of a rapidly enlarging, ulcerated skin lesion, and lack of systemic involvement. Thus, to our knowledge, this is the fourth reported case of periocular C-ALCL without systemic involvement, and the first with contiguous orbital spread. Segal et al.[4] reported an eyelid C-ALCL of a 10-year-old boy without systemic involvement, who underwent incomplete excision and chemotherapy (HM-91 protocol) and remained disease free for 9 months. Tanzi et al.[5] reported an eyelid C-ALCL of a 45-year-old woman who underwent complete excision, but was lost to follow-up before undergoing systemic evaluation. Coupland et al.[2] reported an eyelid C-ALCL of a 48-year-old woman without systemic involvement who underwent biopsy, chemotherapy, and radiotherapy. She remained disease free for 4 years.

A common feature of C-ALCL is favorable outcome. In a large study, survival was 96% for C-ALCL compared with 24% for systemic ALCL with secondary cutaneous involvement.[7] The recommended treatment for C-ALCL is radiotherapy or complete excision, as only cases with multiple lesions are treated with systemic chemotherapy. Spontaneous resolution may occur in some patients.[7] In our case, radiotherapy was used because of clinical extension of the tumor and resulted in complete regression, as would be expected for C-ALCL.

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Idiopathic Sclerosing Orbital Inflammation: Two Cases Presenting With Paresthesia

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Abstract: **The authors report 2 patients with idiopathic sclerosing inflammation of the orbit who presented with periorbital paresthesia in the trigeminal nerve distribution. The diagnosis in both cases was confirmed with biopsy and both patients responded to corticosteroid treatment. Although periorbital paresthesia is usually a sign of malignancy, these cases illustrate that it may also occur in patients with sclerosing orbital inflammation.**

Idiopathic sclerosing inflammation of orbit is a distinct subtype of orbital inflammatory disease characterized by a cicatricial inflammation associated with mass effect and involvement of orbital structures.[1] Though extraorbital extension through the superior and inferior orbital fissures is well documented,[2,3] sensorineural disturbances as a consequence of trigeminal nerve involvement are rare. We report 2 patients with sclerosing idiopathic orbital inflammation presenting with paresthesia in the trigeminal nerve distribution, which resolved with steroid treatment.

CASE REPORTS

Case 1. A 56-year-old white woman had a 3-month history of numbness of the left upper incisors followed by altered sensation over the left nasolabial fold and retrobulbar pain. On examination, visual acuity was 6/6 OU. There was 3 mm of left nonaxial proptosis with 3 mm of superior globe displacement (Fig. 1A) and extraocular movements were normal. A firm mass was palpable at the lateral inferior orbital rim and sensation was decreased over the distribution of the maxillary division of trigeminal nerve distribution with absent corneal sensation. CT demonstrated a well-defined homogenous soft tissue mass occupying the inferolateral orbit and extending through the infraorbital fissure in the infratemporal and pterygopalatine fossae, with involvement of the infraorbital canal (Fig. 1C). The left maxillary sinus was opaque with irregular thickening of the roof of the sinus.

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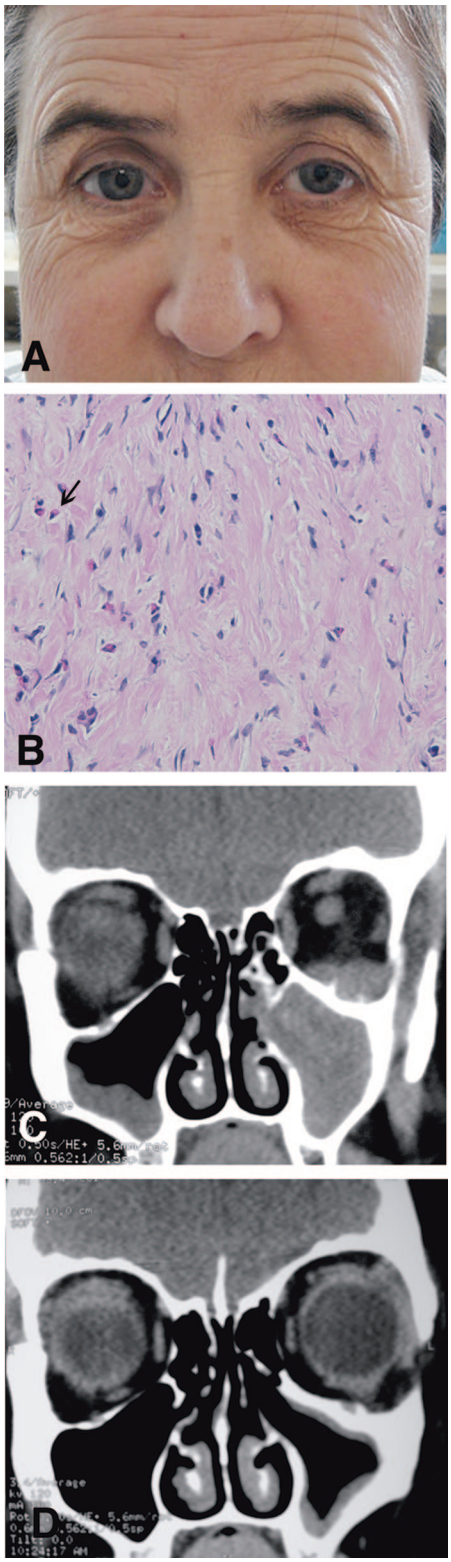

FIG. 1. Case 1. **A,** Clinical photograph shows left axial proptosis with superior globe displacement. **B,** Photomicrograph of orbital biopsy illustrates a fibrotic background in which scattered lymphocytes and eosinophils (arrow) are seen (hematoxylin-eosin, ×200). **C,** CT (pretreatment) shows a left inferior orbital mass with involvement of the infraorbital canal together with left maxillary sinus opacification. **D,** CT following treatment with intraorbital corticosteroid injection shows near-complete resolution of the disease.

Biopsy of the mass via a swinging lower eyelid approach revealed sclerosing orbital inflammation characterized by fibrosis and a chronic inflammatory cell infiltrate admixed with a few lymphoid aggregates (Fig. 1B). Special stains for microorganisms (Gram, periodic acid-Schiff and Gomori methenamine silver stains) were negative. Though the maxillary sinus was not biopsied, the thickening of the roof suggested involvement with the same process.[4] The patient was treated with orbital floor corticosteroid injections (1 ml triamcinolone acetonide 40 mg/ml, monthly for 3 doses). Her symptoms and signs resolved 1 week after the first injection and a significant decrease in the size of the lesion was noted on imaging (Fig. 1D). Two months after the last injection she remained symptom free.

Case 2. A 69-year-old white woman presented with a 4-month history of swelling and redness of the right upper eyelid associated with severe periocular pain, followed 2 months later by paresthesia over the right frontal and maxillary areas. On examination, visual acuity was 6/6 OU. Five millimeters of nonaxial proptosis with 2 mm of inferomedial globe displacement was noted (Fig. 2A) with limitation of right eye elevation. Sensation was decreased over the distribution of the ophthalmic, zygomatico-facial, and zygomatico-temporal branches of the trigeminal nerve, with intact corneal sensation. CT revealed a homogenous soft tissue mass extending from the lacrimal fossa along the lateral wall (Fig. 2B) and through the inferior orbital fissure (Fig. 2C).

Biopsy via a superior eyelid crease incision confirmed sclerosing orbital inflammation (Fig. 2D). The patient was treated with intraorbital injection of triamcinolone acetonide 40 mg/ml (2 doses a month apart) with little response. She was then started on oral prednisolone 75 mg to which she responded dramatically with complete resolution of symptoms and signs within a few weeks. She remains on a tapering dose of prednisolone.

DISCUSSION

Sclerosing idiopathic orbital inflammation is an uncommon subtype of idiopathic orbital inflammation that usually manifests with painful proptosis.[1] Diplopia, blurred vision, and restricted movements are other common findings but subjective sensory disturbances are rare, with only 2 cases previously reported in the literature.[3,5] In fact, sensory dysfunction is uncommon in orbital disease; Rose and Wright[6] in a review of their orbital cases found an incidence of periorbital sensory loss in 3.3% patients. However, subjective sensory loss (as in our patients) was seen in only 0.46% of patients. While malignancies such as adenoid cystic carcinoma and squamous cell carcinoma are known to cause sensory disturbance through perineural spread, Rose and Wright[6] found that 3% of their cases with sensory loss were secondary to orbital inflammatory disease. Periorbital paresthesia has also been reported in association with fungal sino-orbital disease.[7] The sensory loss in sclerosing idiopathic orbital inflammation is most likely secondary to mechanical entrapment of the nerves and the resolution of symptoms and signs following steroid treatment support this theory. It is difficult to explain the loss of corneal sensation in Case 1, but Rose and Wright[6] have also noted the lack of correlation between disease location and corneal hypoesthesia. Hence, in summary, clinicians should be aware of the potential for sclerosing idiopathic orbital inflammation to present with paresthesia due to involvement of sensory nerves.

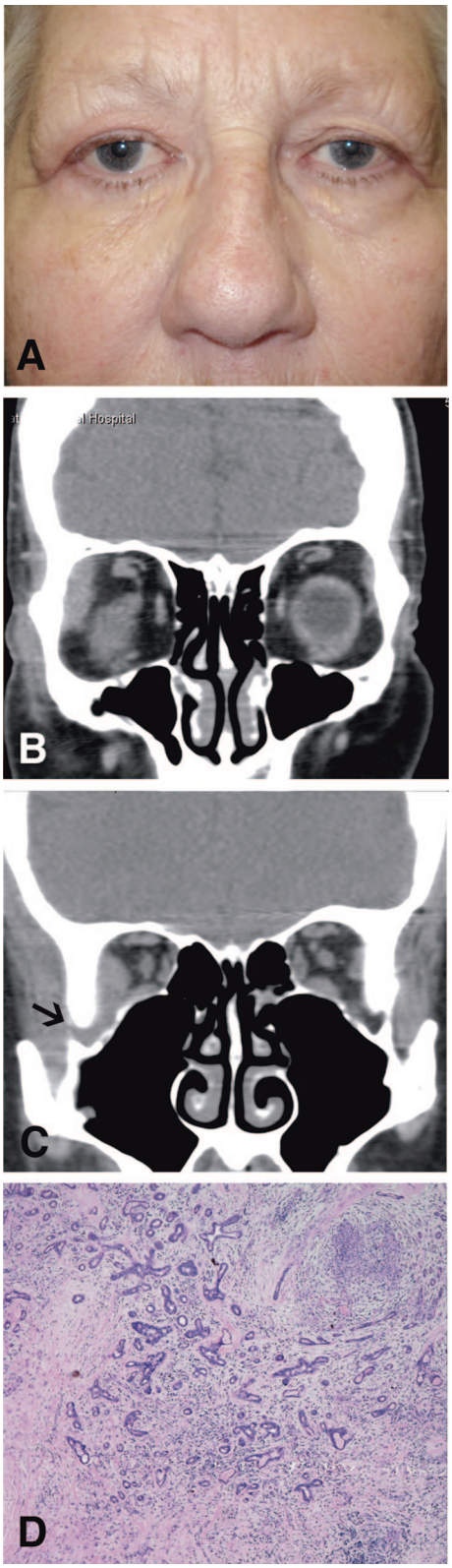

FIG. 2. Case 2. **A,** Clinical photograph shows right nonaxial proptosis. **B** and **C,** CT shows an orbital lesion occupying the right lateral wall of the orbit **(B)** and extending through the inferior orbital fissure **(C)** (arrow). **D,** Photomicrograph of orbital biopsy illustrates sclerosing inflammation with atrophic, disorganized lacrimal gland acini, and foci of inflammatory cells (hematoxylin-eosin, ×100).

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Exaggerated Postsurgical Inflammation in a Patient With Insufficiently Treated Addison Disease

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Abstract: **A patient with Addison disease developed fever, pain, and marked orbital inflammation 3 days after evisceration in the setting of perforated corneal ulcer. He was treated for presumed orbital cellulitis without improvement. Increasing the corticosteroid dose for his Addison disease resulted in complete resolution of the inflammation.**

Patients with Addison disease have reduced cortisol production. A patient with inadequately treated Addison disease who underwent evisceration in the setting of bacterial keratitis with perforation developed exaggerated postoperative inflammation that mimicked orbital cellulitis.

CASE REPORT

A 47-year-old man developed an ulcer within a corneal transplant OS. Perforation developed 3 days later despite hourly topical vancomycin and tobramycin and negative corneal cultures. His ocular history (OS) included congenital cataract, amblyopia, glaucoma requiring Ahmed valve (6 years prior) and penetrating keratoplasty for bullous keratopathy (3 years prior). His medical history included Addison disease. His medications included oral dexamethasone and fludrocortisone.

Examination OS revealed hand motions acuity, a perforated corneal ulcer, flat anterior chamber without hypopyon, and marked conjunctival injection. Evisceration with removal of the Ahmed valve was performed. Absolute alcohol was used to denature residual pigment in the scleral

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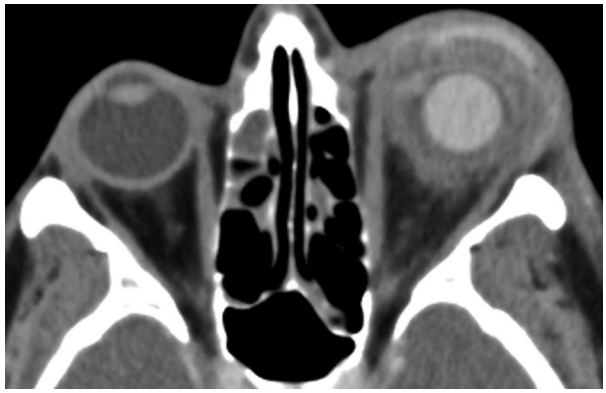

FIG. 1. Axial CT shows marked soft tissue swelling involving the anterior orbit and eyelids. No abscess is evident.

bed and this was followed by copious irrigation with saline. Posterior sclerotomies were not created. A 16-mm polymethylmethacrylate ball was placed. No purulence was found and cultures were negative. Buvipicaine without hyaluroinidase was administered for postoperative analgesia. His daily dexamethasone dose was increased from 0.375 to 1.0 mg the evening before surgery. He resumed his normal dexamethasone dose the evening of surgery and was discharged on oral cephalexin and erythromycin ointment.

Three days after surgery he developed a fever (101°F), orbital pain, marked eyelid erythema and edema, and intense conjunctival injection and chemosis without discharge. His white blood cell count was 8,200/mm^3. CT demonstrated soft-tissue swelling within the anterior orbit and eyelids, but no abscess (Fig. 1). He was treated with intravenous vancomycin and oral levofloxacin for presumed orbital cellulitis. Two days later, his condition had not improved. Repeat CT was unchanged. His temperature remained at 99°F to 100°F and he was orthostatic. His white blood cell count remained normal, although eosinophils were elevated (6%). Serum sodium was 133 mg/dl. The implant was removed. There was no sign of infection and cultures were negative. Scleral biopsy showed no bacteria or fungi, although pathology of the removed cornea confirmed initial bacterial keratitis (Fig. 2). The patient's orthostatic hypotension, fever, hyponatremia, and eosinophilia suggested that his Addison disease was undertreated.[1] The daily dexamethasone dose was increased from 0.375 to 6 mg. The next day his eyelid edema and erythema were markedly improved. He continued taking an elevated dexamethasone dose until a silicone implant was placed at 1 week. Dexamethasone was then tapered over 2 weeks to 0.5 mg daily. Two months after surgery, his orbit remained normal.

DISCUSSION

Our leading diagnosis on admission was orbital cellulitis based on the history and clinical findings. However, orbital cellulitis in the setting of corneal ulcer is rare, and his normal white blood cell count, failure to respond to broad-spectrum antibiotics, and negative cultures led us to question an infectious process. We considered an allergic response to the implant but are unaware of such reactions to polymethylmethacrylate. Acute orbital inflammation after peribulbar or sub-Tenon anesthesia using hyaluronidase has been reported,[2,3] but our patient did not receive this.

We suspect the patient had an exaggerated local inflammatory response to evisceration because of his inadequately treated Addison disease.[4] We postulate that he could not mount the usual endogenous glucocorticoid response to surgical trauma,[5] so he developed sterile orbital inflammation that responded to increased dexamethasone.

Clinicians should be aware of this phenomenon in patients with Addison disease and work closely with the patient's endocrinologist to consider optimal perioperative corticosteroid dosing.

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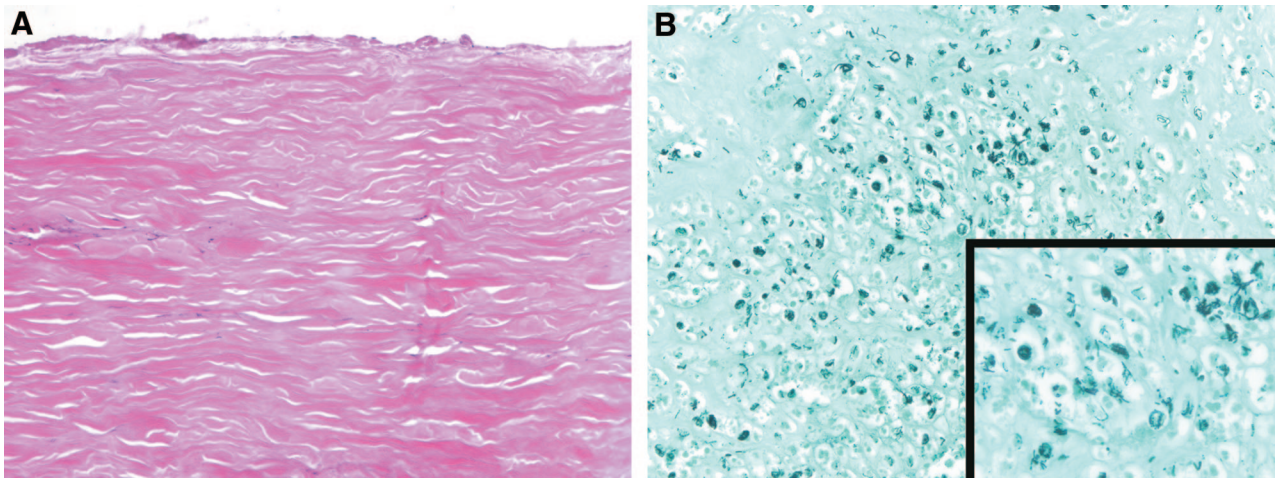

FIG. 2. **A,** Hematoxylin-eosin–stained section of sclera reveals absence of inflammation. Grocott methenamine silver, periodic acid Schiff, and Brown-Hopps staining did not demonstrate any microorganisms (×200). **B,** Cornea removed during evisceration contains abundant cocci often forming pseudofilaments that were beaded (indicating altered organisms from antimicrobial therapy) and best seen on silver staining. Brown-Hopps, periodic acid Schiff, and Fite stains were negative for fungi and mycobacterium species (Gomori methenamine silver, ×200, inset ×400).

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Histologic Features of Mesotherapy-induced Orbital Fat Inflammation

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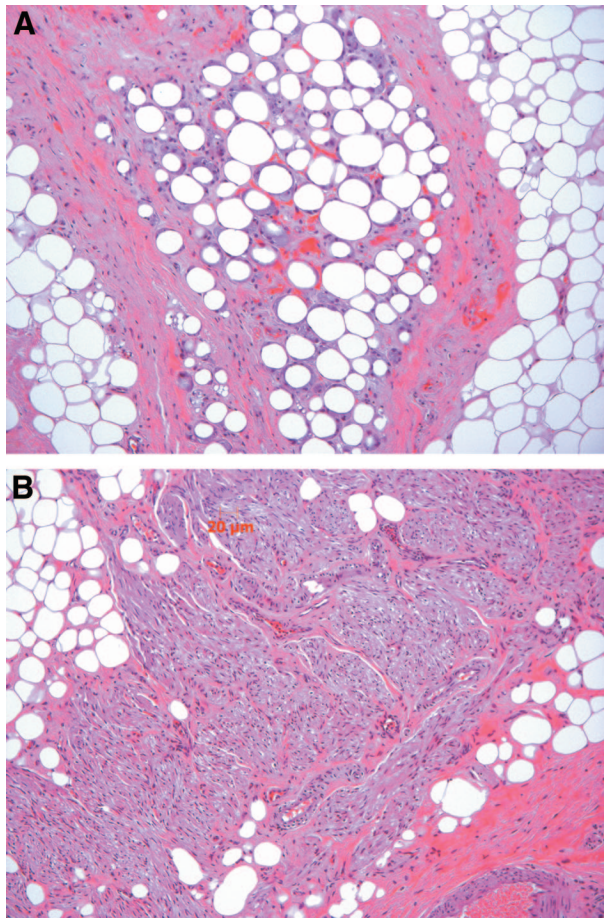
Abstract: **A 67-year-old man developed acute orbital inflammation after receiving cosmetic mesotherapy (Lipo-Dissolve) to the inferior orbital fat compartments. The injection was intended to cause lipolysis and shrinkage of fat lobules with subsequent cosmetic improvement. Injections of a mixture of bile salts, phospholipid, and alcohol preservative bilaterally in inferior orbital fat lobules led to an acute inflammatory reaction characterized histologically 12 days later by mild lymphocytic infiltration, fat necrosis, and fibrosis in the target areas. Benign proliferation of peripheral nerve trunks consistent with a traumatic neuroma was also noted histologically on one side. Inflammation including fat necrosis and traumatic neuroma are all possible consequences of mesotherapy.**

Mesotherapy, injection of various substances to dissolve fat without surgical incision, has been used as an alternative to liposuction for cosmetic purposes. Lipo-Dissolve is an increasingly popular mesotherapeutic agent and consists of a mixture of bile salts, a phospholipid, and an alcohol preservative.[1] It is purported to achieve lipolysis and the subsequent replacement of fat with fibrotic tissue through its detergent effects.[2–4] While the individual constituents are approved, the Food and Drug Administration has not established recommendations for the use of Lipo-Dissolve to reduce redundant subcutaneous or prolapsed orbital fat. Moreover, there are few data on the effects of Lipo-Dissolve on orbital fat compartments, which have a different embryologic origin and have been shown to have some biochemical and structural differences when compared with corporal fat.[5–7] To our knowledge, this is the first report of histologic findings of acute orbital inflammation due to Lipo-Dissolve injection in the inferior orbital fat compartments.

CASE REPORT

A 67-year-old white man presented with severe bilateral eye irritation, pain, tearing, and decreased vision. Four days prior, he elsewhere received Lipo-Dissolve, mesotherapy injections to his inferior orbital fat pads for cosmetic purposes. The injecting practitioner initiated oral prednisone (60 mg) 2 days after injection for the intense inflammation.



A, Photomicrograph shows an island of fat necrosis embedded in dense fibrous tissue with variable-sized lipocytes at various stages of engulfment by multinucleated foreign body giant cells (hematoxylin-eosin, original magnification ×20). **B,** Photomicrograph of benign nodular accumulation of peripheral nerve bundles consistent with a traumatic neuroma noted in excised orbital tissues associated with fat necrosis following cosmetic mesotherapy with bile salt-containing solution (hematoxylin-eosin, original magnification ×10).

This medication did not provide any subjective improvement and was discontinued.

Visual acuity was 20/30-2 OD and 20/30-1 OS. Pupil testing was normal with no afferent pupillary defect. Examination revealed bilateral orbital congestion with lower eyelid fullness, erythema, and warmth. Extraocular motility was slightly restricted in downgaze in proportion to the amount of edema and congestion, but full in other fields of gaze. Lower eyelid fat pad herniation was still evident. Moderate inferior bulbar chemosis caused poor eyelid apposition to the globe. An increased tear lake was observed overlying the chemosis and conjunctival injection, bilaterally. Mild superficial punctate keratitis was noted OU. Intraocular pressures were normal OU. The anterior and posterior segment examinations were otherwise normal.

A bilateral inferior exploratory orbitotomy was recommended and accepted by the patient for purposes of tissue diagnosis to exclude disease not related to the cosmetic injections such as inflammatory pseudotumor, or neoplasm and to debulk what was assumed to be a foreign body reaction. The

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patient was offered various alternatives to surgery including continued observation.

Light microscopy of excised tissues obtained from both orbits 12 days after mesotherapy injections revealed fat necrosis associated with mild lymphocytic infiltration. Prominent abnormally broad bands of fibrosis between lobules of orbital fat were present in all excised tissues. Islands of shrunken lipocytes and residual inflammatory cells were embedded in fibrous septae (Fig. A). Fat necrosis, characterized by variably sized lipocytes apparently being engulfed by multinucleated giant cells within fat lobules, was prominent in all tissues sampled (Fig. A). A benign neuroma (Fig. B) was present in excised tissue from one side.

The postoperative course was characterized by diminishing inflammation subsiding completely over 6 weeks after excisional biopsy. At 2 months postoperatively, the orbits were quiet with no epiphora, pain, redness, injection, or corneal pathology. Extraocular motility returned to normal and visual acuity improved to 20/20 OU.

DISCUSSION

Mesotherapy remains a controversial substitute for or complement to liposuction for general body cosmesis and the use in the treatment of prolapsed orbital fat protruding in the lower eyelids is especially uncertain. This case report demonstrates that mesotherapy, at least with the specific agent used—Lipo-Dissolve—may ignite a substantial inflammatory response when injected in orbital tissues. The traumatic neuroma noted could have been provoked by the injection itself or resulted from a combination of injection trauma and reaction to the mix of substances included, but likely contributed to the patient's discomfort.

Our findings are similar to previous reports on histology from body fat mesotherapy injections. Possible differences between mesoderm-derived subcutaneous body fat and neuro-ectodermal orbital fat, however, may limit the ability to extrapolate results from the treatment of corporal fat.[7–9] The indications for and long-term effects of mesotherapy on orbital tissues need further clarification.

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Orbital Abscesses Caused by *Fusobacterium necrophorum*

Monica Ralli, M.D., Mehryar Taban, M.D., Ronald Mancini, M.D., and Robert A. Goldberg, M.D.

Abstract: **The authors describe 2 cases of young men who presented with pansinusitis and *Fusobacterium necrophorum* orbital abscesses. The first patient had a complicated clinical course including epidural abscess formation and meningitis. He underwent surgical evacuation of the abscesses and sinus drainage and required long-term broad spectrum antibiotic therapy with full recovery. The second patient had a clinical course complicated by *F. necrophorum* septicemia. He required surgical evacuation of the abscess, sinus drainage, and long-term broad-spectrum antibiotic therapy and has remained asymptomatic since. *F. necrophorum* may be emerging as an increasingly prevalent pathogen in orbital abscesses and can portend a more aggressive clinical course where early surgical intervention may be prudent.**

Orbital cellulitis and abscesses often arise in children and young adults as extensions of adjacent paranasal sinusitis. Although *Staphylococcus* and *Streptococcus* species are most commonly implicated,[1] anaerobic bacteria have increasingly been recognized as important pathogens.[2] *Fusobacterium necrophorum* has been rarely associated with orbital abscesses and is the major pathogen associated with Lemierre syndrome, which is the constellation of anaerobic septicemia, internal jugular vein thrombosis, and septic emboli arising from infections of the head and neck.[3,4] We present 2 cases of orbital abscesses caused by *F. necrophorum*.

CASE REPORTS

Case 1. An 18-year-old man presented with 24 hours of right periorbital swelling and pain preceded by 2 weeks of fever, coryza, and malaise. He had received a dose of ceftriaxone 2 days before presentation. On examination, he had moderate right upper eyelid erythema and edema, hypoglobus, 2 mm of proptosis, and ~20% restriction of supraduction OD. His vision was 20/20 OU with no pupillary defects. CT demonstrated complete opacification of the bilateral maxillary, sphenoid, and ethmoid sinuses with a right frontal epidural abscess and right lateral orbital (extraconal) abscess (Fig. 1A, B).

He was initially treated with intravenous vancomycin, ceftriaxone, and clindamycin. On day 2, he acutely developed increased right retrorbital pain, nausea, and lethargy and underwent emergent evacuation of the right frontal epidural abscess, right orbital abscess drainage with Penrose drain placement, and right maxillary antrostomy and anterior ethmoidectomy (Fig. 1C). Postoperatively, the patient received intravenous vancomycin and meropenem and did

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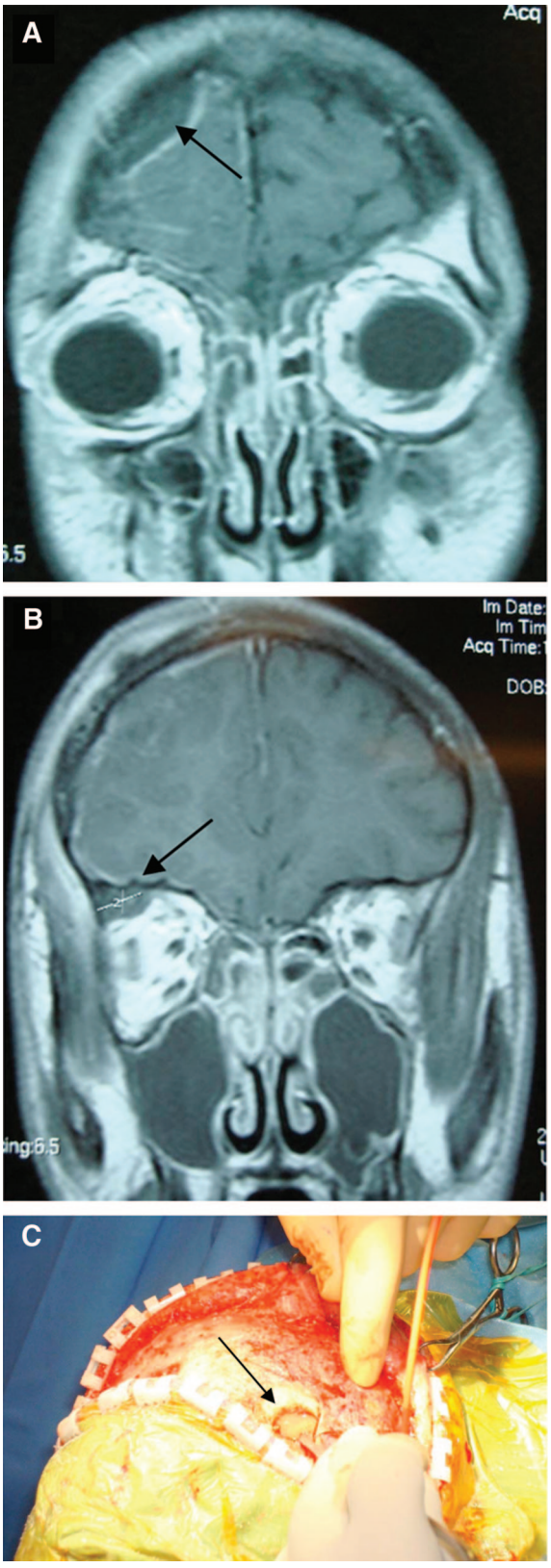

FIG. 1. Case 1. Contrast-enhanced MRI demonstrates **(A)** a 30 mm × 11 mm epidural abscess (arrow) in the right frontal region and **(B)** a 11 mm × 16 mm abscess (arrow) in the right lateral extraconal orbit with complete opacification of the paranasal sinuses. **(C),** Intraoperative drainage of the epidural abscess (arrow).

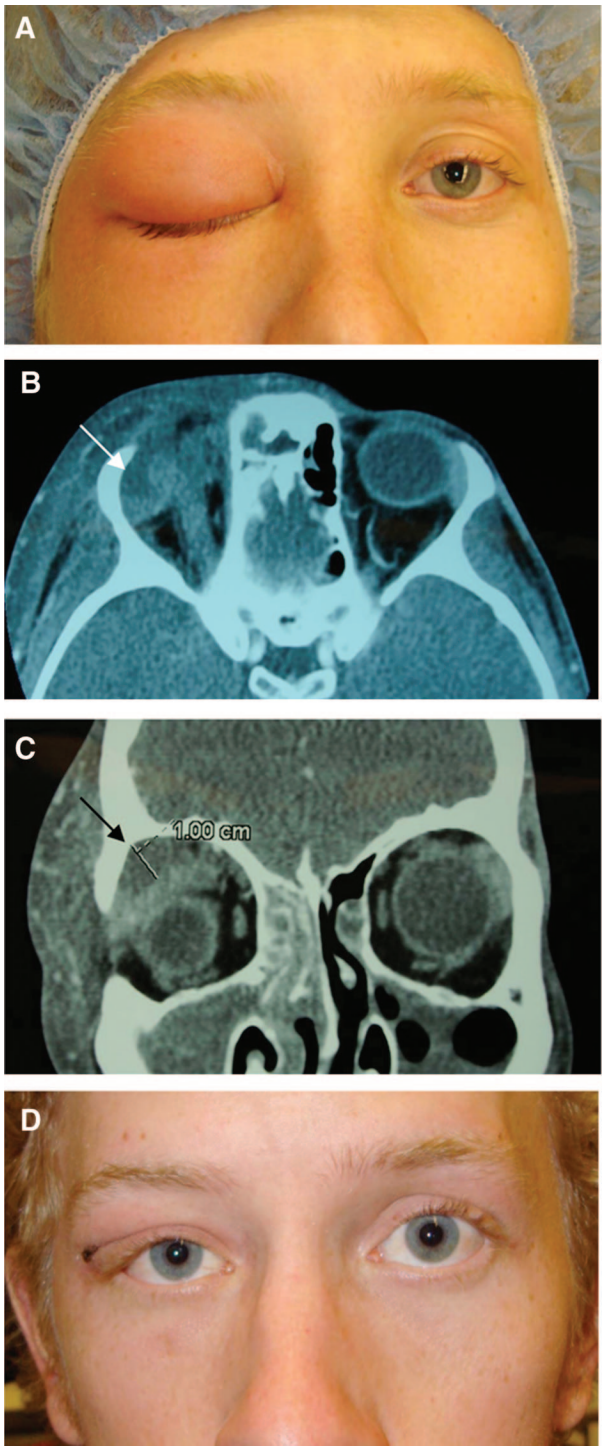

FIG. 2. Case 2. **(A)** A 20-year-old man on presentation with right orbital cellulitis. **(B)** Axial and **(C)** coronal CT of the right superolateral extraconal orbital abscess (arrows); 10 mm in greatest dimensions. **(D),** Two weeks after drainage of orbital abscess with marked resolution of orbital cellulitis.

well; the Penrose drain was removed on postoperative day 4. However, on postoperative day 5, the patient again decompensated with worsened headache, altered mental status, and nuchal rigidity. Imaging revealed leptomeningeal enhancement with interval decrease in sizes of the epidural and

orbital abscesses. Lumbar puncture was consistent with meningitis. CT venography demonstrated no venous sinus thrombosis, and no valvular vegetation or intracardiac shunts were seen on echocardiography. At this time, culture results from the intraoperative aspirates became available and demonstrated growth of *F. necrophorum*. All blood cultures were negative. Oral metronidazole was added to the patient's antibiotic regimen. He clinically improved with full extraocular motility and resolution of swelling and pain. The patient was discharged home on postoperative day 8 with intravenous vancomycin, meropenem, and oral metronidazole for 4 additional weeks and has remained asymptomatic.

Case 2. A 20-year-old man presented with 10 days of progressively worsening right periorbital swelling, pain, fever, coryza, and malaise, despite oral antibiotics. On examination, he had significant right upper and lower eyelid erythema and swelling, hypoglobus, and ~80% limitation of supraduction OD (Fig. 2A). Visual acuity was 20/20 OU with no afferent pupillary defects. CT demonstrated right pansinusitis, partial opacification of the left maxillary and ethmoid sinuses, and a loculated right superolateral orbital (extraconal) abscess (Fig. 2B, C). He underwent drainage of the orbital abscess with Penrose drain placement, debridement of necrotic periorbital soft tissues, right anterior and posterior ethmoidectomy, and maxillary antrostomy. Postoperatively, the patient received intravenous vancomycin and piperacillin/tazobactam. On day 3, blood cultures taken at admission demonstrated growth of *F. necrophorum*; cultures from the intraoperative aspirates remained negative. Oral metronidazole was added to the patient's antibiotic regimen. To evaluate for Lemierre syndrome given the *Fusobacterium* bacteremia, bilateral upper extremity venous duplex studies and MR venogram were performed. These studies showed no evidence of jugular vein thrombosis. The Penrose drain was removed on postoperative day 5, and the patient was discharged home with intravenous vancomycin, piperacillin/tazobactam, and oral metronidazole for 4 weeks. Two weeks after discharge, he had minimal right upper eyelid swelling, resolution of hypoglobus, and full extraocular motility (Fig. 2D).

DISCUSSION

F. necrophorum is a potentially virulent anaerobic gram-negative rod-shaped bacterium that normally inhabits the oral cavity. While *F. necrophorum* classically has been associated with pharyngitis, it has been implicated in several cases of mediastinitis, meningitis, cavitary pneumonia, gingivitis, and other necrotic soft tissue infections and Lemierre syndrome.[3] Three cases of orbital abscesses caused by *F. necrophorum* have been previously described in patients aged 13, 16, and 21 years.[5–7]

Neither of our cases fulfilled the criteria for Lemierre syndrome, although both had complicated clinical courses, with epidural abscess formation and meningitis in the first patient and septicemia in the second. The present report highlights the importance of *F. necrophorum* as an emerging pathogen in orbital abscesses and suggests that these patients may require earlier surgical intervention and more aggressive medical management.

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Osteogenesis With Hematopoiesis Simulating Infection in a Hydroxyapatite Orbital Implant

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Abstract: **A 28-year-old woman underwent secondary orbital implant surgery with placement of a hydroxyapatite implant. Over the next 7 years she underwent 3 drilling procedures. She began having copious discharge 1 year after the last drilling procedure. She was seen on numerous occasions with socket discharge, unresponsive to a variety of topical and oral antibiotics. Clinically, with the conjunctiva diffusely inflamed, the implant tender to touch, and the presence of a pyogenic granuloma, implant infection was suspected and the implant subsequently removed. Histopathologic assessment revealed widespread lamellar bone formation, including focal areas of marrow with active extramedullary hematopoiesis. There was no evidence of an inflammatory process or infection. Postoperatively the patient's symptoms and signs resolved. Extramedullary hematopoiesis within hydroxyapatite implants is rare. Porous orbital implant infection is also rare. Osteogenesis with extramedullary hematopoiesis simulating implant infection has not previously been reported.**

Infection of porous orbital implants is a rare yet feared complication that may be difficult to control without implant removal.[1–3] The clinical course is often prolonged because the early symptom of recurrent discharge, a common problem for implant recipients, may delay the diagnosis.[3] Osteogenesis and hematopoiesis within hydroxyapatite (HA) orbital implants (Bio-Eye, Integrated Orbital Implants, Inc., San Diego, CA, U.S.A.) is also rare.[4–6] Osteogenesis with hematopoiesis simulating an infection within an HA implant, to our knowledge, has not been reported in the English literature. We describe a case of a HA implant with widespread lamellar bone formation and active extramedullary hematopoiesis with recurrent socket discharge, conjunctival inflammation, pyogenic granuloma formation, and implant tenderness simulating an implant infection.

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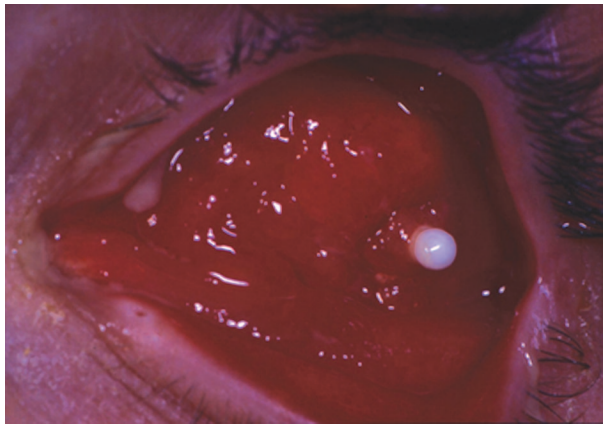

FIG. 1. Diffuse inflammation of conjunctiva, with purulent discharge and a pyogenic granuloma encircling a polycarbonate sleeve.

CASE REPORT

A 28-year-old woman underwent secondary orbital implant surgery with an HA implant to improve socket motility. Over the next 7 years, 3 drilling procedures were preformed to position her polycarbonate peg and sleeve peg system. Twelve months after her last drilling procedure she began experiencing purulent discharge from the socket. Despite numerous courses of topical and oral antibiotics, the copious discharge persisted and was accompanied by sanguineous discharge at times and socket tenderness. On examination, the bulbar and palpebral conjunctiva were grossly inflamed with a papillary reaction. Purulent discharge was present in the fornices and a pyogenic granuloma was identified encircling the polycarbonate sleeve (Fig. 1). The implant was tender to touch through the closed eyelids. Although conjunctival cultures were negative, implant infection was suspected and removal was recommended. The implant was removed and replaced with a polymethylmethacrylate sphere. A culture of the implant at the time of removal did not grow any organisms. Postoperatively, the discharge resolved, the socket inflammation cleared, and the patient's symptoms resolved. Histopathologic examination revealed widespread lamellar bone formation, including focal areas of marrow formation with active extramedullary hematopoiesis as confirmed by immunohistochemistry (Fig. 2). Dense fibrovascular septa and patchy areas of fat deposition also were seen intermixed with dissolved HA. Occasional multinucleated giant cells were seen in the vicinity of the dissolved HA; however, there were no other signs of inflammation. Special stains for microorganisms were negative.

DISCUSSION

HA orbital implants are commonly used in anophthalmic socket surgery. Infection after implantation is rare, yet feared, because implant removal is often required to treat the problem.[3] Early signs and symptoms of implant infection include discharge and conjunctival inflammation, but are nonspecific, as patients with an artificial eye occasionally have these findings. Persistent discharge and conjunctival inflammation despite multiple drops should raise suspicion of an implant infection.[1–3] Recurrent pyogenic granulomas and discomfort on palpation of the implant also should alert the clinician to possible implant infection.[1,3]

In the present case, implant infection was suspected clinically as the case had all the characteristic features.[3] Interestingly, no organisms were identified by culture or histopathology

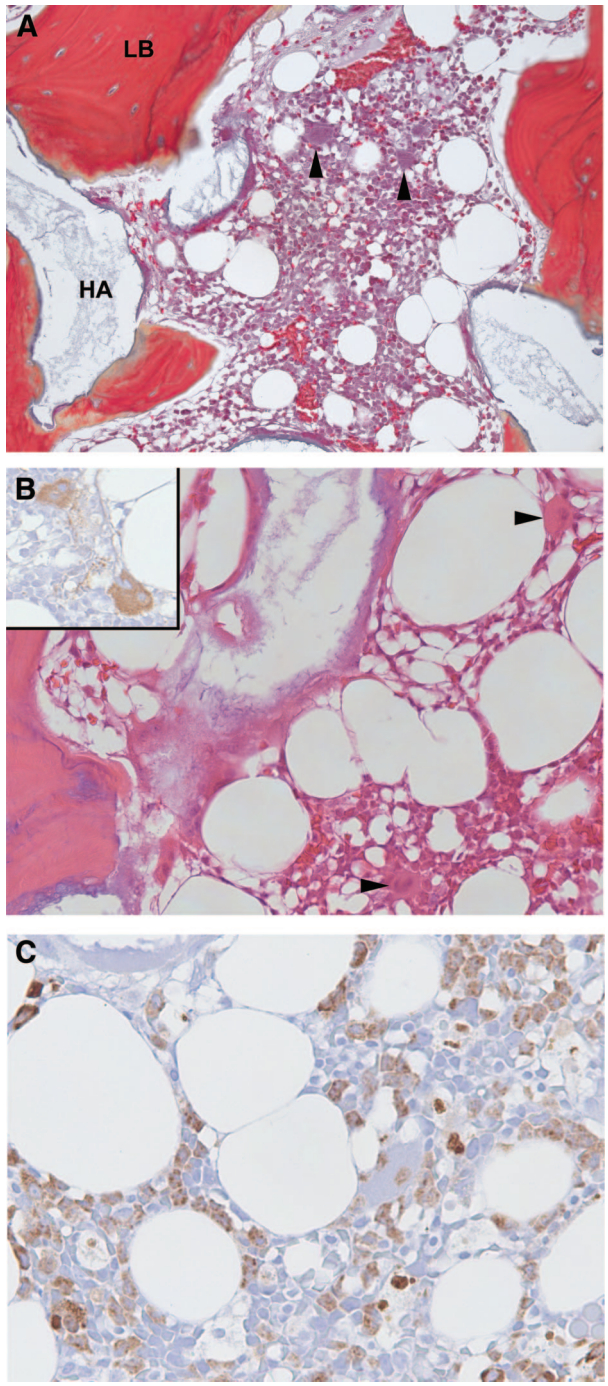

FIG. 2. **A**, Implant showing marrow with active extramedullary hematopoiesis including megakaryocytes (arrowheads) surrounded by lamellar bone (LB) and dissolved hydroxyapatite (HA) (Movat, ×200). **B**, High-power view of extramedullary hematopoiesis demonstrating megakaryocytes (arrowheads, hematoxylin-eosin, ×400). The inset shows positive cytoplasmic staining with factor VIII-related antigen characteristic of megakaryocytes (×320). **C**, Extramedullary hematopoiesis with granulocytic precursors showing positivity for myeloperoxidase (×640).

but rather widespread lamellar bone formation with foci of active extramedullary hematopoiesis. Bone formation in HA implants has been previously reported.[3–6] To our knowledge, extramedullary hematopoiesis has only been identified in the

case reported by Sires and Benda[6] and in the rabbit model of Lew et al.[7] The presence of bone has been attributed to an osteoinductive effect of HA.[5] Two possible mechanisms by which bone may form in HA implants were postulated by Green et al.[5]: transformation of circulating monocytes or osseous metaplasia of fibrocytes that migrate in the implant. Of 6 patients who underwent HA implant removal by other authors at 5, 7, 8, 8, 11, and 66 months that were found to have bone formation, only one had any clinical symptoms that may potentially have been attributed to the bone formation.[4–6] The implant was from a 70-year-old woman who had undergone enucleation.[5] Eight months later she had intractable pain and the HA implant was removed. Unfortunately, the authors did not state whether the patient's symptoms resolved, how extensive the bone formation was, or whether there was any hematopoiesis. It is therefore difficult to attribute the intractable pain to the bone formation. In 10 of 15 patients we previously reported who underwent HA implant removal because of signs and symptoms suggestive of implant infection, bone formation (without hematopoiesis) was documented.[3] In each case histopathologically, there was either acute or chronic inflammatory cells found with or without the presence of necrosis (abscess) or bacterial organisms.[3] In our patient, with extensive bone formation and hematopoiesis, copious discharge was the main symptom with implant tenderness being less significant. Clinically, she appeared to have an implant infection, but histopathologically there were no signs of implant infection, just the extramedullary hematopoiesis.

In summary, the most serious complication of porous implants is infection and implant removal is often required as treatment.[1–3] Recurrent discharge, pyogenic granuloma formation, and tenderness of the implant are hallmarks of an infected implant.[1–3] The present case indicates that these symptoms and signs also may be hallmarks of osteogenesis with extramedullary hematopoiesis in the implant. Alternatively, they may represent coincidental findings unrelated to this symptomatology. To our knowledge, implant removal is the only known treatment.

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