Radiologically Manifested Accessory Extraocular Muscles in Thyroid Eye Disease

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Abstract: **The authors describe 3 patients with Graves orbitopathy who all have accessory orbital structures on orbital imaging. These structures are isodense on CT and isointense on MRI to the extraocular muscles and have a similar anatomical appearance to accessory extraocular muscles previously described in the literature. The authors propose that Graves orbitopathy involves these accessory muscles and makes them visible on imaging.**

Accessory muscles in the human orbit are rare. They have been described in anatomical studies and in cases of restrictive strabismus.[1] Imaging of anomalous orbital structures has only been reported in a small number of these cases.[1–5] Only one of these cases has histologic confirmation of muscle tissue.[4]

Three types of accessory muscle have been described in the literature[1]: those arising from the extraocular muscles themselves, separate fibrous bands located beneath the recti, and discrete structures arising from the posterior orbit. Accessory muscles have not been described in Graves orbitopathy (GO). We describe a series of 3 patients with GO who have accessory orbital structures demonstrable on CT and MRI of the orbit.

CASE 1

A 67-year-old male was diagnosed with hyperthyroidism on the basis of a raised T4 and low thyroid stimulating hormone. He initially underwent medical treatment with carbimazole (20 mg/day). This dose was reduced during the following 12 months. He was subsequently diagnosed with GO after developing mild proptosis and intermittent vertical diplopia on upgaze. The magnetic resonance scan of his orbits demonstrated an accessory structure within both orbits originating from the orbital apex lateral to the optic nerve. This passed anteriorly in the intraconal orbit, medial to the lateral rectus muscle, and thinned at the lateral border of the inferior rectus insertion (Fig. 1). His orbitopathy improved spontaneously with resolution of his diplopia.

CASE 2

A 38-year-old female with known Graves disease was referred with signs of GO with increasing proptosis. Enlargement of the extraocular muscles was confirmed on CT scanning. Her clinical picture deteriorated as she developed dysthyroid optic neuropathy, which persisted despite a combination of oral prednisolone and azathioprine. She eventually required bilateral orbital decompression.

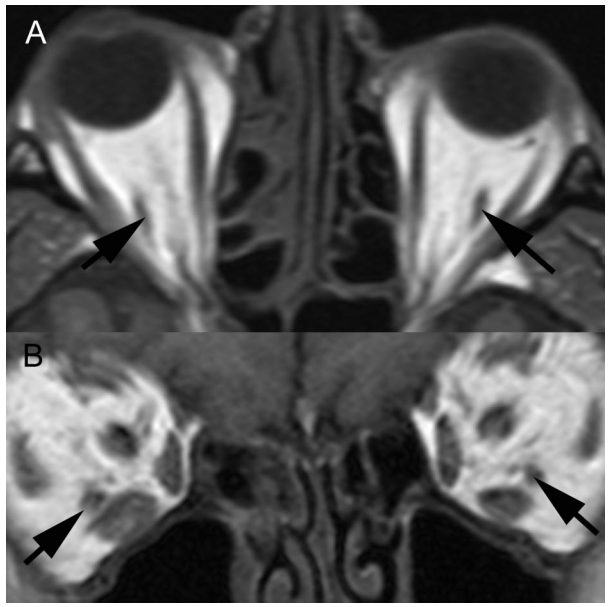

FIG. 1. T1W axial and coronal magnetic resonance images. The accessory extraocular structures in both orbits are shown (arrows).

Initial CT scan (Fig. 2A, B) demonstrated bilateral accessory orbital structures that originated from the orbital apex lateral to the optic nerve and passed anteriorly in the intraconal orbit medial to the lateral rectus insertion. Following medical treatment, MRI (Fig. 2C, D) showed a reduction in size of both the accessory structures and the extraocular muscles.

The patient developed a restrictive strabismus affecting the inferior rectus that required surgical correction. The insertion of the left orbital accessory structure (muscle) was isolated and biopsied. The histology demonstrated fibrous tissue.

CASE 3

A 42-year-old female presented with grossly asymmetric proptosis. The thyroid function tests at presentation showed a raised T4 (39.6 mmol/l) and low TSH (0.03 mmol/l). Features of GO were confirmed on MRI (Fig. 3A, B). She was treated with a combination of systemic immunosuppression and orbital radiotherapy.

MRI demonstrated a structure originating from the right orbital apex, which passed anteriorly in the intraconal orbit medial to the lateral rectus. It thinned to a vertical strap lying at the lateral border of the inferior rectus. Follow-up CT demonstrated a reduction in the proptosis and size of the extraocular muscles and of the accessory structure (Fig. 3C, D). She responded well to treatment and has inactive disease with persistent right proptosis and no diplopia.

DISCUSSION

All cases showed similar accessory orbital structures that were isointense to the extraocular muscles on all magnetic

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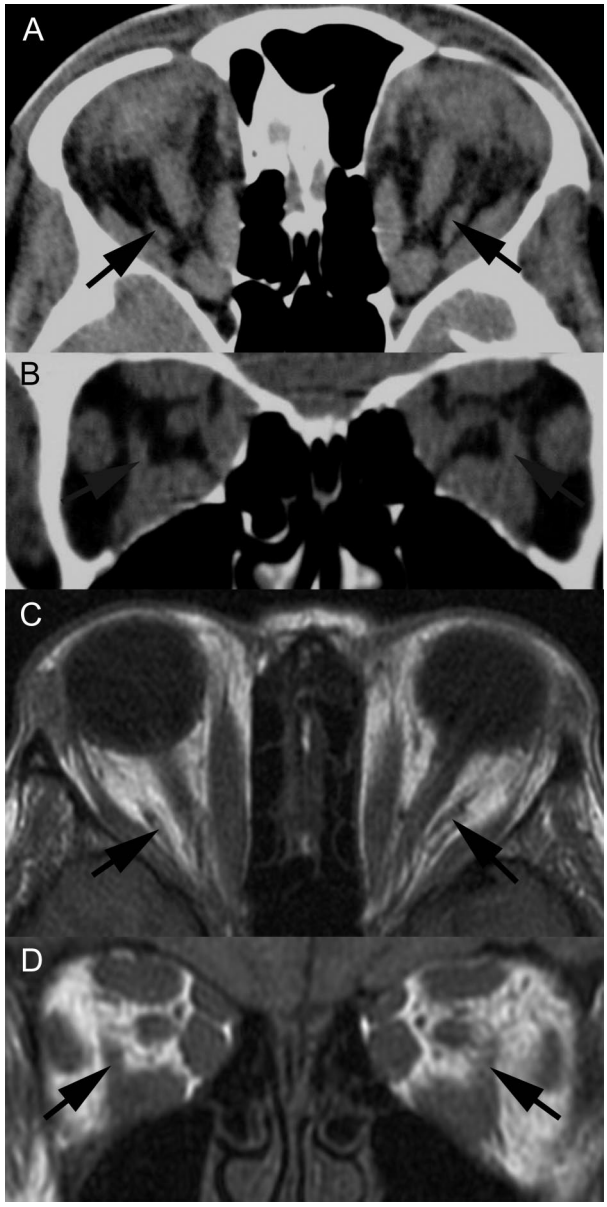

FIG. 2. Pretreatment: axial **(A)** and coronal **(B)** CT images demonstrating the accessory extraocular structures in both orbits (arrows). Posttreatment: axial T1W **(C)** and coronal T1W **(D)** magnetic resonance images demonstrating the accessory extraocular structures, which are isointense to the other extraocular muscles on all sequences (arrows).

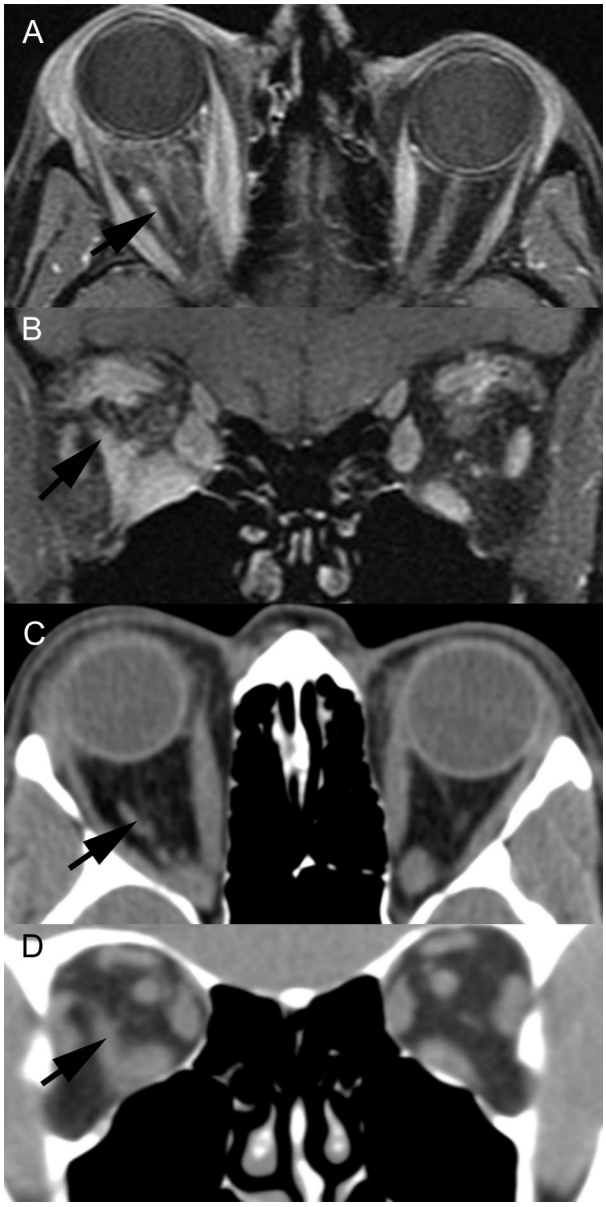

FIG. 3. Pretreatment: axial **(A)** and coronal **(B)** TI fat saturation postcontrast images. The extraocular structures enhance postcontrast. The accessory extraocular structure in the right orbit is shown (arrows). Posttreatment: axial **(C)** and coronal **(D)** CT images.

resonance sequences. The presence of a vestigial retractor bulbi was initially proposed. However, these accessory structures are not close enough to the optic nerve to be consistent with a retractor bulbi.[1] It is more likely that they represent accessory lateral rectus muscles, an almost invariable feature in macaque monkeys, although this was not confirmed histologically.[6] An accessory lateral rectus muscle is a rare radiologic finding with only 5 case reports of accessory muscles being identified radiologically. None of those were reported in the presence of GO, and only one had histologic confirmation that muscle fibres were present in the accessory structure biopsied.[4] We have described a series of 3 patients, all with similar accessory orbital structures. If these structures were indeed muscle, then we would expect them to enlarge with the other extraocular muscles in GO and decrease in size after treatment of the condition. This appears to have occurred in both Cases 2 and 3.

CONCLUSION

The reduction in size of the accessory orbital structures after treatment of the GO lends weight to the argument that these accessory orbital structures are indeed accessory muscles in vivo although histologic confirmation was not obtained. These cases suggest that GO may make accessory muscles manifest on imaging.

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A Unique Case of Dermolipoma Located in the Lower Eyelid

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Abstract: **A 16-year-old healthy girl was referred to the authors' clinic for a palpable mass in her right lower eyelid. The patient was treated under the initial diagnosis of chronic conjunctivitis and ectopic cilia in the medial part of her right lower eyelid at a local clinic for 1 year. A soft, nontender, 2.0 × 1.5 × 1.0-cm elliptical mass was palpable in the medial aspect of the patient's right lower eyelid. The medial part of the palpebral conjunctival surface was covered by keratinized epithelium with fine hairs. Histologic findings were consistent with a dermolipoma. The authors describe a very rare case of dermolipoma in the lower eyelid presenting with keratinized epithelium on the medial palpebral conjunctiva.**

A 16-year-old, otherwise healthy girl presented with discomfort and redness in her right eye for 1 year. She was diagnosed with chronic conjunctivitis and ectopic cilia in the medial part of her right lower eyelid at a local clinic. She was treated with topical antibiotics and repeated epilation of aberrant cilia intermittently, which provided only temporary relief. One month prior to referral, a palpable mass was discovered in her right lower eyelid.

Examination of the right eye revealed a nontender, soft, movable, 2.0 × 1.5 × 1.0-cm oval mass in the right lower eyelid. The mass was connected by keratinized palpebral conjunctiva with a few fine hairs on the surface. The keratinized epithelium occupied the medial portion of lower palpebral conjunctiva, extending horizontally from the lower punctum to the caruncle and vertically from the eyelid margin to the inferior fornix (Fig. 1). The other ophthalmologic examinations were unremarkable. An orbital CT scan revealed a 2.0 × 1.0-cm ovoid soft tissue mass with density of fat in her right lower eyelid (Fig. 2). The patient underwent excision of the mass. The mass was seen below the tarsal border, extending from the area of keratinized epithelium to the medial two thirds of the lower eyelid and to the fornix inferiorly (Fig. 3). The procedure was performed carefully to avoid damage to the inferior canaliculus. Once excised, the integrity of lacrimal canaliculus was confirmed by probing.

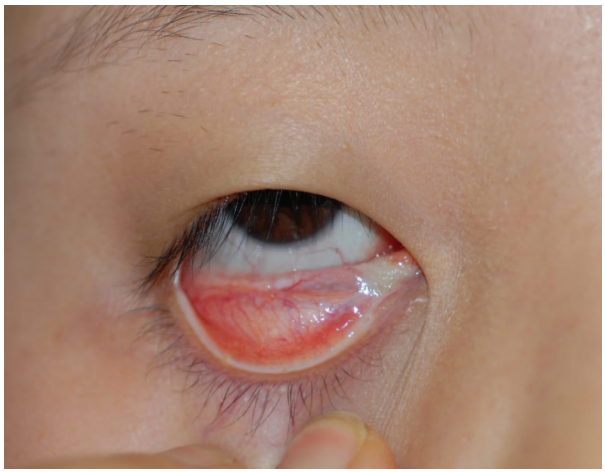

FIG. 1. The palpebral conjunctiva medial to punctum was covered by whitish keratinized epithelium with a few fine hairs.

Histologically, the mass showed keratinized squamous epithelium, and the underlying stroma contained collagenous tissue, pilosebaceous units, and large amount of adipose tissue in the deeper aspect (Fig. 4), consistent with a dermolipoma.

DISCUSSION

Dermolipomas consist of fatty tissue, bundles of dense collagen, and hair follicles covered with keratinized and nonkeratinized stratified squamous epithelium and may occasionally contain fine hairs on their surface. Dermolipomas are usually located in the superotemporal conjunctival fornix, near the lacrimal gland and lateral rectus muscle.[1–4]

Choristomas are congenital benign tumors consisting of elements of normal tissue in an abnormal location.[5] Dermolipomas are classified as choristomas and thought to arise from abnormal sequestration or implantation of surface ectoderm

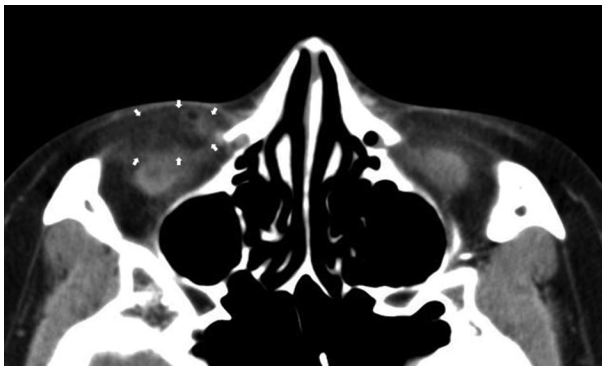

FIG. 2. Axial CT scans show a 2.0 × 1.0-cm ovoid soft tissue mass with density of fat in the right lower eyelid.

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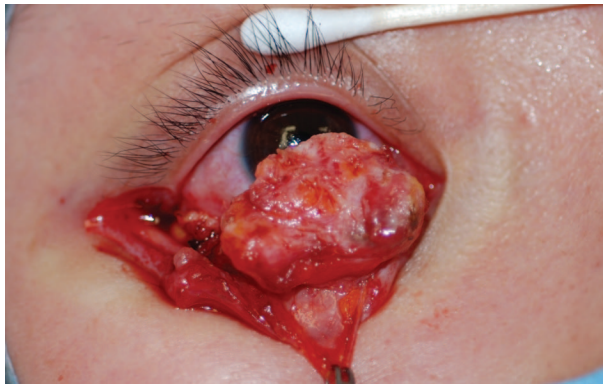

FIG. 3. Intraoperative photograph shows a fatty soft tissue mass measuring approximately 2 × 1.5 × 1.0 cm.

along embryonic lines of closure at between 3 and 5 weeks' gestation.[3,4]

While there have been previous reports of choristomas occurring on the lower eyelid, these have been of phakomatous choristomas and odontogenic choristomas.[6,7] A review of the literature failed to find any report of a dermolipoma of the lower eyelid, making our case the first to be reported. Dermolipomas are congenital and very common in children thus are usually diagnosed at a younger age. In our case, the adolescent patient was initially diagnosed 1 year ago and treated for chronic conjunctivitis and only recently discovered to have a lower eyelid palpable mass. We speculate that there may have been a progressive growth of the dermolipoma during puberty with the keratinized epithelium and fine hairs irritating the conjunctival surface.[3,4] Other reports have noted a history of increasing size often occurring at puberty.[8] Complete surgical excision of the dermolipoma is the recommended management, because it causes irritation and cosmetic complaint. In this procedure, every effort should be made to preserve otherwise healthy neighboring structures, especially the inferior canaliculus.

In conclusion, when evaluating patients presenting with a lower eyelid palpable mass with keratinized palpebral conjunctiva, dermolipoma should be included in the differential diagnosis.

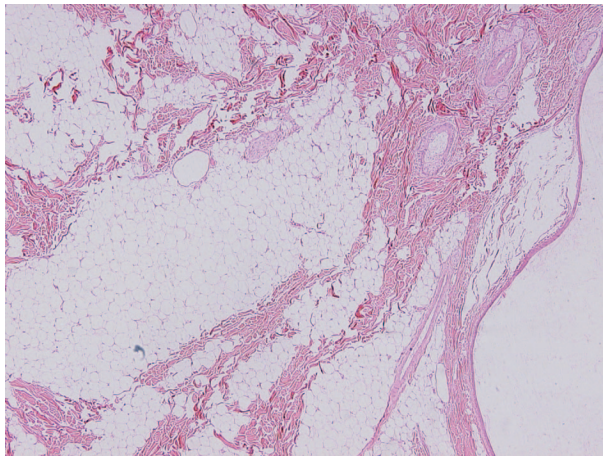

FIG. 4. Histopathologic examination of the mass revealed keratinized stratified squamous epithelium, collagenous tissue, pilosebaceous units, and large amount of adipose tissue in the underlying stroma (hematoxylin-eosin stain, ×40).

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Ocular Ischemia and Ischemic Oculomotor Nerve Palsy After Vascular Embolization of Injectable Calcium Hydroxylapatite Filler

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Abstract: **A healthy 25-year-old man who received a calcium hydroxylapatite filler injection for nose augmentation by a dermatologist suddenly developed blepharoptosis and orbital pain on the right side, associated with progressive visual disturbance of the right eye. Patchy necrosis at the nose and glabella, limitations of extraocular movements, and anterior segment ischemia, as evidenced by conjunctival injection, chemosis, corneal edema, dilated pupil, hyphema, and hypopyon, were noted. Orbital CT demonstrated linear deposits of a similar density to bone in the right medial orbit and eyelid, suggestive of multiple emboli along the conjunctival vessels. A provisional diagnosis of ocular ischemia and ischemic oculomotor nerve palsy secondary to vascular embolization was made. After 3 months, visual acuity, all intraocular inflammation, oculomotor nerve palsy, and skin necrosis resolved completely except for a dilated pupil.**

Facial soft tissue augmentation using various fillers is becoming increasingly common, having the distinct advantages of being an effective, safe, and less invasive nonsurgical method. However, there are a variety of possible complications, including hypersensitivity reaction, ecchymosis, contour abnormalities, inflammation or necrosis at the injection site, infection, and arterial occlusion or embolization.[1]

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Vascular embolization after periorbital filler injection is a rare but most serious complication.[2–4] To the best of our knowledge, anterior segment ischemia concurrent with vascular embolization after injection of calcium hydroxylapatite (CaHA, Radiesse, Bioform Inc., San Mateo, CA, U.S.A.) filler has not been reported previously.

CASE REPORT

A healthy 25-year-old man who received CaHA filler injection for nose augmentation by a dermatologist suddenly developed blepharoptosis and orbital pain on the right side. The procedure was stopped promptly, and an attempt to remove the injected material by aspiration was made. However, the orbital condition worsened, and he developed a central necrosis and surrounding reddish reticular pattern affecting the glabellar region, the bridge of the nose, and the right eyelid (Fig. 1A).

Eight hours after the procedure, he also complained of progressively decreased vision of the right eye. The initial visual acuity of the right eye was hand movement. The intraocular pressure was within normal limits. The right pupil was fully dilated, but a reverse afferent pupillary defect was not present. Right exotropia was noted with limitations of extraocular movements in medial, up, and down gaze (Fig. 1B). Slit lamp examination of the right eye showed conjunctival injection, chemosis, corneal edema, and severe anterior chamber reaction, including hyphema and hypopyon (Fig. 1C). Multiple emboli along the conjunctival vascular walls were noticeable (Fig. 1D). Fundoscopy could not be performed because of corneal edema, but B-scan ultrasonographic findings were normal. Orbital CT demonstrated linear deposits of a similar density to bone in the right medial orbit and eyelid, suggestive of multiple emboli along the conjunctival vessels (Fig. 2). The patient was diagnosed with anterior segment ischemia and oculomotor nerve palsy secondary to vascular embolization. He was initially treated with topical antibiotics and steroids, including intravenous antibiotics. The wound was soaked in a warm normal saline and covered with ointment and daily dressings. Cultures of aqueous humor and exudates of skin lesion were negative for organisms. After confirming the results of cultures, he was placed on a low dose of tapering oral corticosteroids.

After 5 weeks, visual acuity increased to 20/40 with pinhole in the right eye with improvement of corneal edema and anterior chamber inflammations. Limitations of extraocular movements, blepharoptosis, and skin necrosis resolved gradually. Fundoscopic examination showed many glittering spots, which were thought to be emboli in the nasal area, and fluorescein angiography revealed window defects due to choroidal ischemia in the same area. Three months later, visual acuity was 20/20 with pinhole in the right eye. All intraocular inflammation and oculomotor nerve palsy had completely resolved except for a fixed dilated pupil. The skin also recovered with minimal scar formation (Fig. 3).

DISCUSSION

Vascular embolization after periorbital filler injection rarely occurs but is a most serious complication. There are several reports of retinal or ophthalmic arterial occlusion following injection of autologous fat or hyaluronic acid (Restylane, Q-Med AB, Uppsala, Sweden) in the glabellar region.[2–4] It is hypothesized that the embolized material enters the ocular circulation through retrograde arteriolar flow after intraarterial injection in one of the peripheral

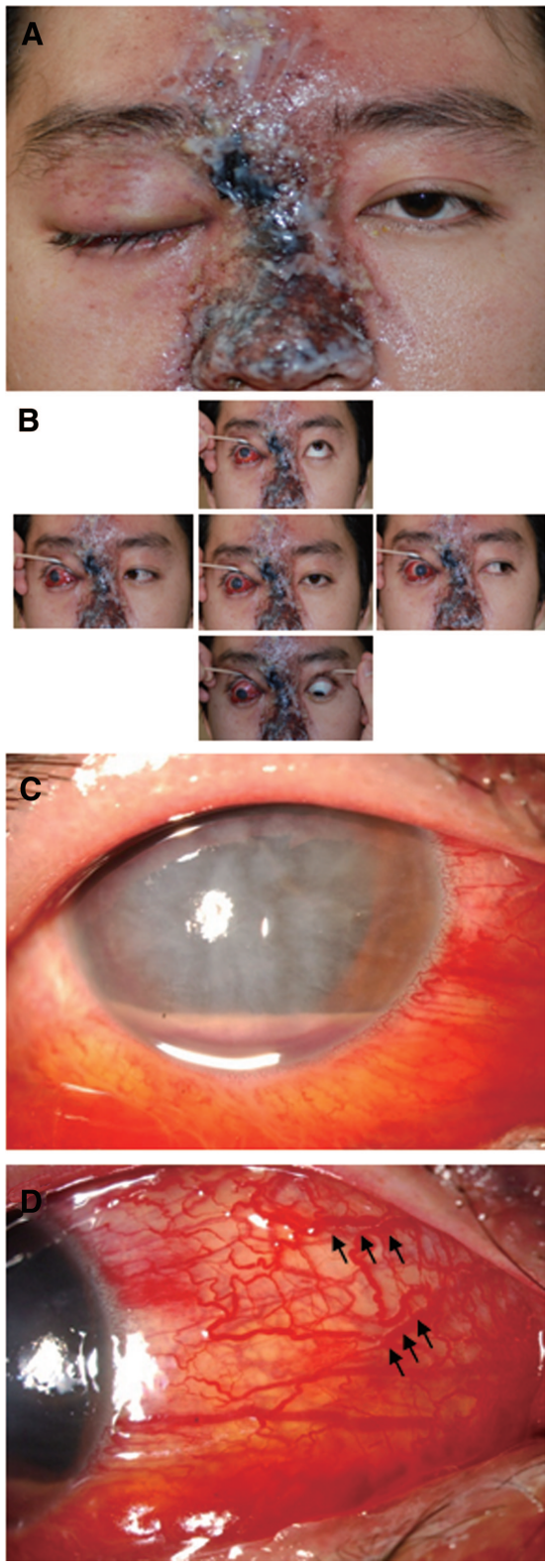

FIG. 1. **A,** Facial photograph shows patchy necrosis at the nose and complete blepharoptosis of right side. **B,** Right exotropia in primary gaze is noted with limitations of extraocular movements in medial, up, and down gaze. **C,** Slit lamp examination demonstrates corneal edema, aneterior chamber reaction with hyphema and hypopyon, and a mydriatic pupil. **D,** Multiple, white emboli are seen along the conjunctival vessels (arrows).

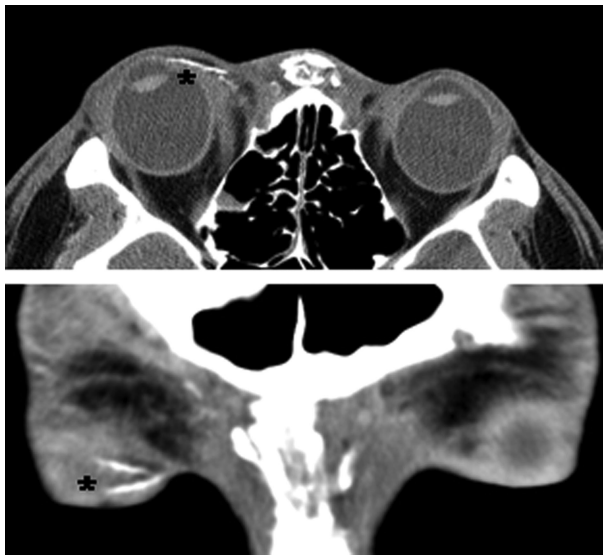

FIG. 2. Orbital CT scan with axial and coronal views demonstrates linear deposits of similar density to bone in the right medial orbit and eyelid, suggestive of multiple emboli along the conjunctival vessels (asterisks).

branches of the ophthalmic artery. However, to the best of our knowledge, anterior segment ischemia has not been previously reported with filler injection.

Anterior segment ischemia is a serious complication of strabismus and retinal detachment surgery and is also reported in patients undergoing other ocular procedures such as cyclocryotherapy and laser photocoagulation.[5] Blood supply to the anterior segment of the eye is derived from the long posterior ciliary arteries, the anterior ciliary arteries, and the conjunctival arteries. Therefore, embolization involving these vessels following periorbital injection can induce anterior segment ischemia as a result of the vascular insufficiency of the eye.

In our patient, the CaHA particles probably entered the dorsal nasal artery, a terminal branch of the ophthalmic artery. We postulate that the emboli moved in a retrograde fashion to the ophthalmic artery under the pressure of injection and subsequently affected the anterior ciliary arteries, medial posterior ciliary artery, and branch to the oculomotor nerve. The emboli along the conjunctival vessels, severe inflammation of anterior chamber, and ischemic iris atrophy were consistent with anterior segment ischemia secondary to embolization of the anterior ciliary arteries. Ischemic oculomotor nerve palsy could explain complete blepharoptosis and right exotropia with limitations of extraocular movements in medial, up, and down gaze. Fortunately, anterior segment ischemia and ischemic oculomotor nerve palsy completely recovered except for a fixed dilated pupil. The final visual acuity was also minimally compromised because the choroidal involvement from posterior ischemia was limited to the nasal area, hence sparing the macula.

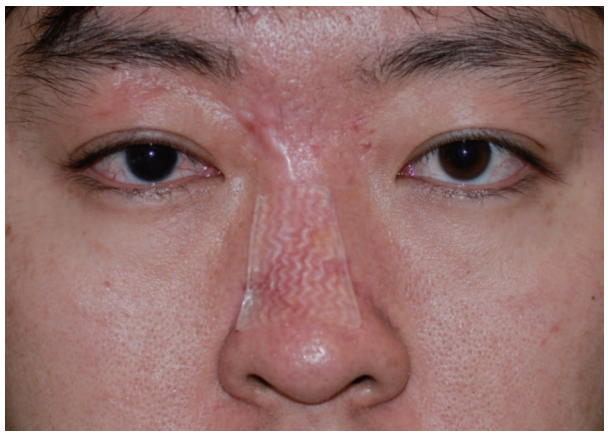

FIG. 3. Three months later, the skin is well recovered with minimal scar formation.

We report a case of anterior segment ischemia in conjunction with posterior segment and orbital ischemia after injection of CaHA filler for nose augmentation. Anterior segment ischemia is rare but should be kept in mind as one of the manifestations of vascular embolization. Physicians should be aware of injection techniques, such as aspiration before injection, injection on needle withdrawal, and injection of small aliquots of filler, to avoid vascular embolization.[1]

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Isolated Optic Nerve Infiltration in Systemic Lymphoma—A Case Report and Review of Literature

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Abstract: **Ocular involvement in non-Hodgkin lymphoma occurs either as primary ocular, central nervous system lymphoma or isolated intraocular lymphoma. Recurrence of systemic non-Hodgkin lymphoma in the form of CNS lymphoma has been reported. However, recurrence as an isolated optic nerve lesion without involving CNS has never been reported in the pediatric age group. We report a case of systemic diffuse large B-cell lymphoma in a 2-year-old female, which primarily occurred as suprapubic mass and later recurred in the form of isolated optic nerve infiltration, after remission of the primary disease. Early detection and prompt treatment resulted in complete reversal of the disease.**

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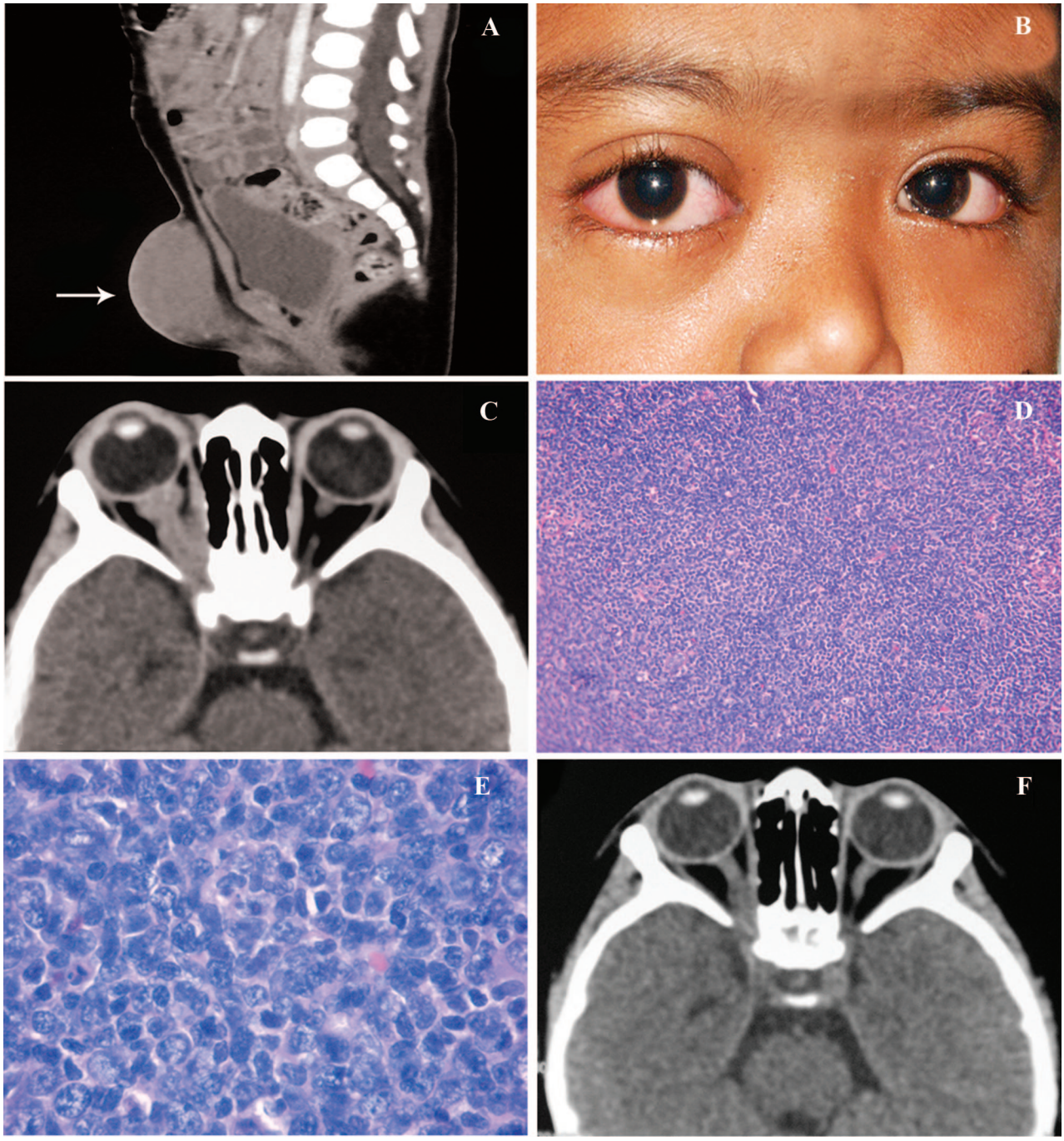
Central nervous system (CNS) involvement in non-Hodgkin lymphoma (NHL) is uncommon but occurs in about 10% of cases.[1] Of these, 5% may develop optic nerve infiltration, and this usually occurs in established CNS disease.[1] We report a case of systemic B-cell lymphoma causing early isolated optic nerve infiltration as the only sign of disease recurrence in a pediatric patient.

CASE REPORT

A 2-year-old female presented with a suprapubic mass of 1 month's duration. A CT scan of abdomen revealed multiple poorly enhancing hypodense areas replacing the renal parenchyma with enlargement of both kidneys, en-



A, CT (sagittal) of the abdomen showing a heterogeneously enhancing soft tissue lesion involving subcutaneous compartment of suprapubic region measuring 6.0 × 6.47 × 5.64 cm (white arrow). **B,** Photograph of the same patient in clinical remission 6 months later with axial proptosis. **C,** CT scan (axial) of the orbit showing a heterogeneously enhancing expansile lesion surrounding the right optic nerve with a central waist measuring about 2.46 × 1.07 cm and extending up to the intracanalicular part of the optic nerve. **D,** Low power view (×10) showing diffuse arrangement of lymphoid cells. **E,** High power view (×100) showing large atypical lymphoid cells. **F,** CT scan (axial) of the orbit after completion of 6 cycles of chemotherapy showing regression of the optic nerve lesion with residual disease at orbital apex measuring 1.31 × 0.81 cm.

larged inguinal lymph nodes, and a heterogeneously enhancing soft tissue lesion involving subcutaneous compartment of the suprapubic region (Fig. A). The patient underwent incision biopsy of the subcutaneous lesion with lymph node biopsy, and this revealed atypical large lymphoid cells arranged in diffuse pattern suggestive of diffuse large B-cell NHL (Fig. D, E). Immunohistochemistry was positive for CD 20 and CD 45 confirming the diagnosis of B-cell lymphoma. CD 10 was negative, and Ki-67 activity was 30% to 35%. The patient received 6 cycles of chemotherapy with adriamycin, vincristine, cyclophosphamide, and prednisolone. A repeat CT scan of the abdomen showed evidence of postoperative changes with complete regression of the enlarged inguinal lymph nodes, renal, and subcutaneous lesions.

Six months later, the patient presented with redness and pain in the right eye for 1 month. Anterior segment examination of the right eye revealed eyelid edema and conjunctival congestion with axial proptosis (Fig. B). Posterior segment examination revealed disc edema. The anterior and posterior examination of the left eye was normal. A CT scan of the orbit revealed a heterogeneously enhancing expansile lesion surrounding the right optic nerve with a central waist measuring about 2.46 × 1.07 cm and extending up to the intracanalicular part (Fig. C). A CT scan of the brain was normal.

A complete blood count and erythrocyte sedimentation rate were normal. The patient and her parents were screened for HIV, which was found to be negative. A bone marrow aspirate along with biopsy and cerebrospinal fluid (CSF) analysis revealed no malignant cells. Polymerase chain reaction on the CSF was negative for fungus. A CT scan of the chest was normal. Based on these findings, a presumptive diagnosis of lymphomatous infiltration of the optic nerve was made. The patient was treated with 6 cycles of chemotherapy (adriamycin, vincristine, cyclophosphamide, and prednisolone regime) and intrathecal methotrexate (8 mg) with each cycle. A repeat CT scan showed regression of the optic nerve lesion with residual lesion at orbital apex, suggesting a favorable response to chemotherapy (Fig. F). The patient is undergoing radiotherapy to the right optic nerve up to orbital apex (dosage 4000 cGy over 4 weeks) and is on close follow-up.

DISCUSSION

Optic nerve involvement by systemic lymphoma often occurs from direct invasion by tumor cells, which tend to infiltrate cranial and spinal nerves and their meninges[2] or may develop as a complication of leptomeningeal lymphomatosis or orbital lymphoma.[2]

Optic nerve involvement in NHL may occur in the form of lymphomatous infiltration, vincristine-related neuropathy, radionecrosis, and granulomatous or fungal infection secondary to immunosupression.[1] In our case, the 2 main diagnostic considerations were infiltrative optic neuropathy of lymphomatous origin and granulomatous or fungal infectious process. The characteristic thickening of optic nerve and resolution of optic nerve lesion following chemotherapy confirmed the diagnosis of recurrence.

When infiltrative optic neuropathy occurs in NHL, it usually is accompanied by CNS involvement, abnormal CSF, or pathologic bone marrow.[3] Previous reports have documented optic neuropathy with CNS involvement in patient with systemic lymphoma in clinical remission.[1,4] However, isolated optic nerve infiltration in a systemic NHL is very rare with only 2 cases reported to date. The first case was bilateral optic nerve infiltration in a 56-year-old male patient with systemic NHL and AIDS with CNS involvement in form of intracranial nodules in paraventricular distribution.[2] The second case reported was of optic nerve infiltration in a 55-year-old female patient with systemic lymphoma with presence of malignant cells in the CSF, indicating more diffuse lymphomatous meningitis.[5]

There were some unique features in our case. Lymphomatous optic nerve infiltration from systemic NHL in the pediatric age group and from this primary site has never been reported. In the previous reports, optic nerve infiltration was late in the course of the disease with CNS involvement in the relapse or with an abnormal CSF or bone marrow. Our patient developed optic nerve infiltration 6 months after completion of chemotherapy for systemic lymphoma with a normal CSF and bone marrow. Also, systemic or CNS relapse of lymphoma did not occur in our patient.

Isolated optic neuropathy in NHL is extremely rare. The management of such patients requires prompt investigation to exclude treatable causes, such as infection or drug toxicity.

This case demonstrates that infiltrative optic neuropathy may occur as the sole manifestation of disease recurrence in a patient with systemic NHL otherwise thought to be in clinical remission.

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Floppy Eyelid Syndrome and Ptosis in a Patient With Pachydermoperiostosis

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Abstract: **A patient with the rare condition of pachydermoperiostosis with secondary ptosis and floppy eyelid was successfully treated with a combination of levator advancement and an upper eyelid tarsal strip. This is the second report in the literature of combined floppy eyelid and ptosis with this condition and the first report of this surgical approach for its management. The histopathology of the excised eyelid tissue found changes consistent with both pachydermoperiostosis and floppy eyelid syndrome.**

Pachydermoperiostosis or primary hypertrophic osteoarthropathy is a rare inherited disorder characterized by digital clubbing, periostosis, and facial enlargement. Oph-

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thalmic features may include bilateral eyelid ptosis, while an associated floppy eyelid syndrome has been described once previously. We report a patient with both bilateral eyelid ptosis and floppy eyelid syndrome successfully treated with an upper eyelid lateral tarsal strip and levator advancement. The histopathologic findings are discussed.

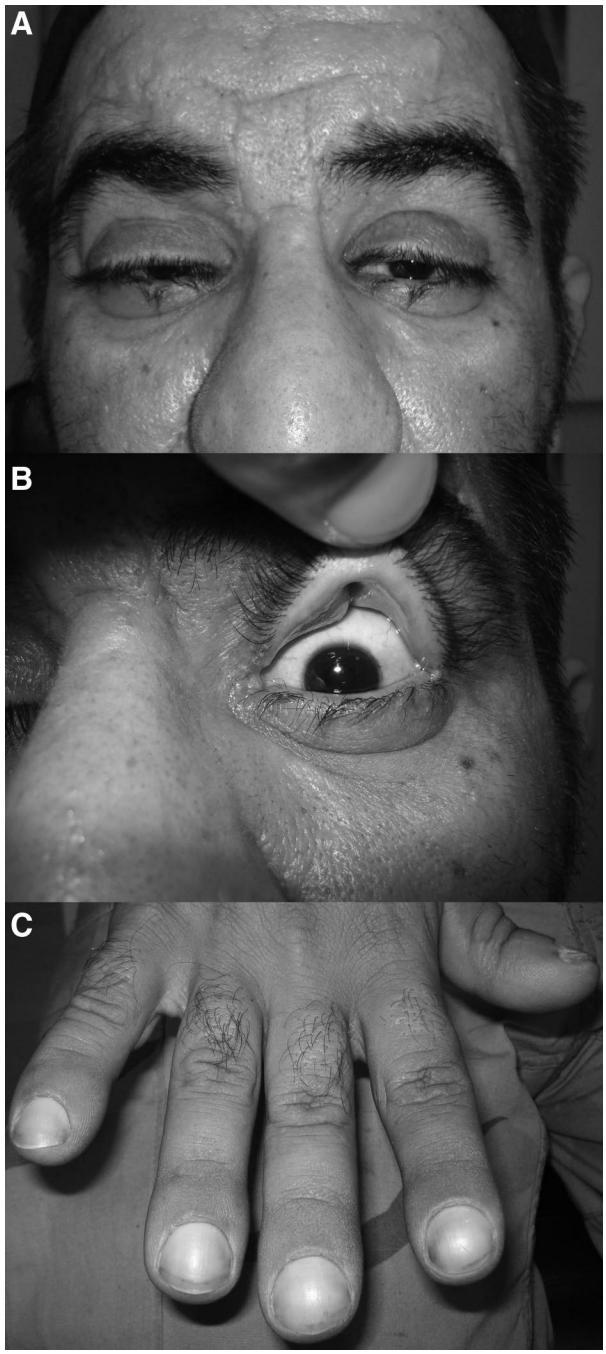

FIG. 1. **A,** 22-year-old male with thickened, oily facial skin, and prominent folds in the forehead and face. **B,** Upper eyelid is floppy and easily everted to demonstrate velvety, giant papillae on the palpebral conjunctiva. **C,** Large hands and clubbed fingers typical of pachydermoperiostosis.

CASE REPORT

A 29-year-old male was referred complaining of recurrent upper eyelid ptosis and ocular discharge and irritation on awakening. Both he and his brother were diagnosed with pachydermoperiostosis (PDP) in mid-adolescence. He was treated with upper eyelid blepharoplasties and levator advancement by a plastic surgeon 5 years previously and repeat left upper eyelid levator advancement and horizontal shortening with a central pentagonal excision by another ophthalmologist 1 year later. Two years later, he underwent bilateral refractive surgery (LASIK) and excision of bilateral nasolabial and glabellar rugae.

On presentation, he had markedly thickened, oily, and furrowed skin (Fig. 1A). His hands and feet were enlarged with distal clubbing (Fig. 1C). There were no signs of prognathism or macroglossia. He was not obese and had no history of obstructive sleep apnea or of sleeping with his face buried in the pillow. X-ray examinations showed cortical thickening of the long bones of the extremities with a thin periosteal reaction. Blood tests revealed a mild leukopenia and a normochromic normocytic anemia.

Visual acuity was 20/50 right and 20/30 left. His upper eyelid skin was elastotic, and the lids were thickened and overrode the lower ones. They were horizontally lax and could easily be everted, demonstrating a velvety, papillary conjunctival reaction (Fig. 1B). Both upper lids were ptotic with a margin-reflex distance of 0 mm right eye and 1 mm left eye. He had good levator function. Surgical scars were visible in both upper eyelid creases, and an irregularity was evident in the left upper eyelid centrally at the site of a previous wedge excision (Fig. 2B). His corneae had mild inferior punctate epithelial erosions. The remaining ocular examination was normal.

The diagnosis of bilateral residual ptosis and floppy eyelid syndrome associated with PDP was made. Bilateral upper eyelid horizontal tightening with tarsal strips was performed to remedy his floppy eyelid syndrome; because his eyelids remained severely ptotic following this maneuver, bilateral levator advancements were performed at the

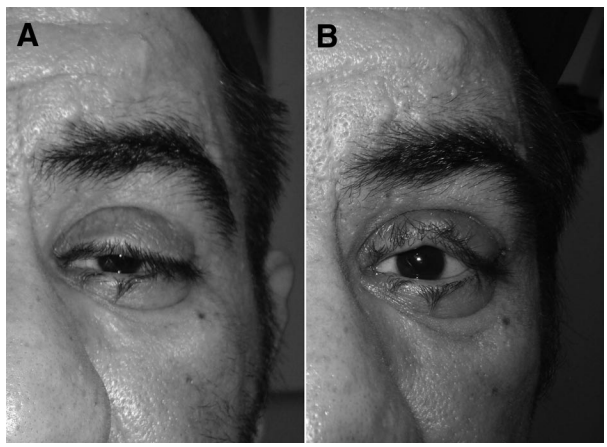

FIG. 2. **A,** Preoperative view of the left eyelid showing marked ptosis and thickened skin despite previous horizontal shortening with a central wedge excision and 2 previous attempted levator advancements. **B,** One week postoperative view of the left eyelid, demonstrating improvement of ptosis and horizontal laxity. Note the central peak from previous central wedge excision and the lateral canthal suture from his upper eyelid lateral tarsal strip.

same sitting (Fig. 2A, B). The patient was pleased with the improved field of vision and reduced morning discharge. Tissue from the left eyelid was submitted for histopathology and showed thickening of the tarsal plate, meibomian gland hyperplasia (Fig. 3A), thickening of the dermis, perivascular lymphocytic infiltration, and conspicuous giant conjunctival papillae (Fig. 3B).

DISCUSSION

Hypertrophic osteoarthropathy has been classified in 2 forms: primary and secondary.[1] Primary hypertrophic osteoarthropathy (PDP) is an autosomal dominant inherited disorder with no identifiable underlying disease. It was first described by Friedreich[2] in 1868. Secondary hypertrophic osteoarthropathy is associated with thoracoabdominal neoplasia or chronic disease. Both have similar clinical features. The skin and skeletal system are primarily affected with coarse facial wrinkles, cutis vertices gyrata, digital clubbing, "elephant feet," and thickening of the long bones caused by periostitis and osteogenesis.[1] Anemia, as described in our patient, has also been associated. Various conditions such as acromegaly, Paget disease, syphilis, and leprosy must be excluded.[1] PDP is more common in men and typically develops during puberty and progresses in adulthood. The underlying pathophysiology is unknown although altered peripheral blood flow leading to capillary stasis has been proposed.[1]

Thickened and ptotic upper eyelids are a common feature of PDP and have been previously described.[3,4] The floppy, easily everted upper eyelids and bilateral papillary conjunctival reaction seen in our case are features typical of floppy eyelid syndrome, first described by Culbertson and Ostler[5] in 1981. This association has only been previously described by Downes et al.[6] in 1989. Since our patient was not obese and had no history of obstructive sleep apnea and did not sleep with his face buried in the pillow, we speculate that the horizontal laxity results from the characteristic dermal changes rather than from mechanical stretching of the lids during sleep.

Management is essentially surgical. We performed a combined horizontal eyelid shortening with an upper eyelid tarsal strip and levator advancement on both upper eyelids with a satisfactory functional and aesthetic result at a follow-up visit 13 months later. A lateral tarsal strip procedure for horizontal eyelid shortening in PDP has not been previously described. Although only one group has reported an association with floppy eyelid syndrome,[6] other surgeons have performed wedge excisions to shorten the upper eyelid because levator advancement alone caused ectropion of the upper eyelid margin.[3,4]

Histopathologic findings of the eyelid in PDP include sebaceous gland hyperplasia, enlargement of sweat glands, thickening of the dermis with an increase in collagen content, deposition of mucin, and perivascular lymphocytic infiltration.[4] Our histopathologic findings are consistent with those previously described. Additionally, the conjunctival papillary reaction is consistent with the histopathologic findings in floppy eyelid syndrome.

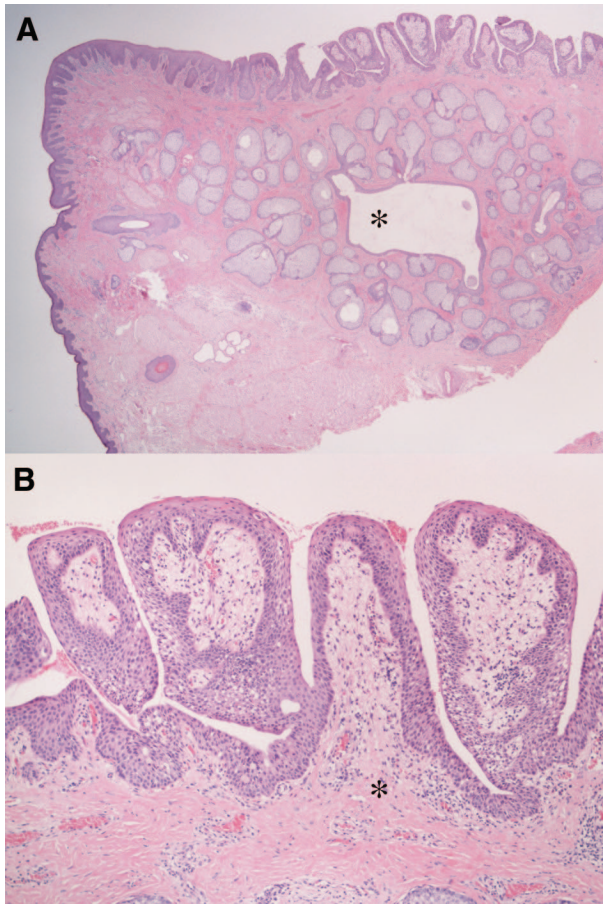

FIG. 3. **A,** Low power (×20) photomicrograph demonstrating hypertrophic sebaceous glands with dilated central duct (asterisk), consistent with previous descriptions of PDP. **B,** High power (×100) photomicrograph shows giant papillae in the palpebral conjunctiva with perivascular inflammation and thickened dermal collagen bundles (asterisk) consistent with previous descriptions of floppy eyelid.

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Pyoderma Gangrenosum of the Eyelids: Recurrence in a Skin Graft

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Abstract: **A 41-year-old woman presented with pyoderma gangrenosum involving the right eyelids. Over the course of 3 years, she developed progressive scarring of the eyelids with lagophthalmos and corneal exposure. Multiple reconstructive procedures were performed with varying degrees of success. Despite aggressive medical control of her disease, it recurred in grafted skin on her upper and lower**

eyelids as confirmed by tissue biopsy. Periocular pyoderma gangrenosum is a rare disease where the management may be complicated by disease recurrence in reconstructed tissues, including skin grafts as this case demonstrates.

Pyoderma gangrenosum (PG) is a neutrophilic dermatosis that clinically manifests as an ulcerating, often painful skin disease without infection. It is often associated with systemic disorders such as inflammatory bowel disease, connective tissue disorders, or diabetes mellitus. Its occurrence in periocular tissues is rare, and case reports are limited.[1,2] The disease is controlled with various systemic immunosuppressive agents, and rehabilitation of periocular tissues is ideally undertaken when the disease is quiescent.

The clinical course of a patient with periocular PG is presented, including the unusual feature of disease recurrence in a full-thickness skin graft of the upper and lower eyelids.

CASE REPORT

A 41-year-old black woman was referred by her dermatologist to the Ophthalmology Service at Grady Memorial Hospital for evaluation of PG involving the brow. She had been diagnosed with PG 8 years prior, and her disease had initially involved her scalp and progressed to involve her lower extremities. None of the areas affected had been subjected to prior trauma or surgery. She was otherwise healthy, without evidence of a known systemic disease commonly associated with PG. She had previously been treated with combinations of prednisone, cyclosporine, sulfasalazine, dapsone, azathioprine, and mycophenolate mofetil with varying degrees of disease control. Her initial ophthalmic examination was unremarkable with the exception of a well-circumscribed area of ulceration superior and lateral to the right lateral brow (Fig. 1).

She was medically managed by her dermatologist until she presented to the ophthalmology clinic 2 years later, noting that she could no longer close her right eye. At that time, she had punctate epithelial keratitis OD caused by cicatricial lagophthalmos associated with diffuse right upper eyelid ulceration. Aggressive lubrication was started, and a lateral tarsorraphy was performed after she developed a large epithelial defect.

Approximately 1 month later, she underwent right upper eyelid débridement and blepharotomy with full-thickness skin grafting. Over the next 13 months, she underwent 3 additional skin grafts to her right upper eyelid and 1 to her right lower eyelid with tarsorraphies due to recurrent cicatricial lagopthalmos (Fig. 2). During this time, her disease in other areas had not progressed.

She did well without evidence of active disease until her 6-month follow-up, when both the upper and lower eyelid grafts were found to be ulcerated, and there was significant exposure keratopathy with associated corneal scarring. She had repeat full-thickness skin grafting of both the right upper and lower eyelids. Biopsy of the prior skin grafts at that time showed a diffuse neutrophilic infiltrate with necrosis consistent with PG (Fig. 3).

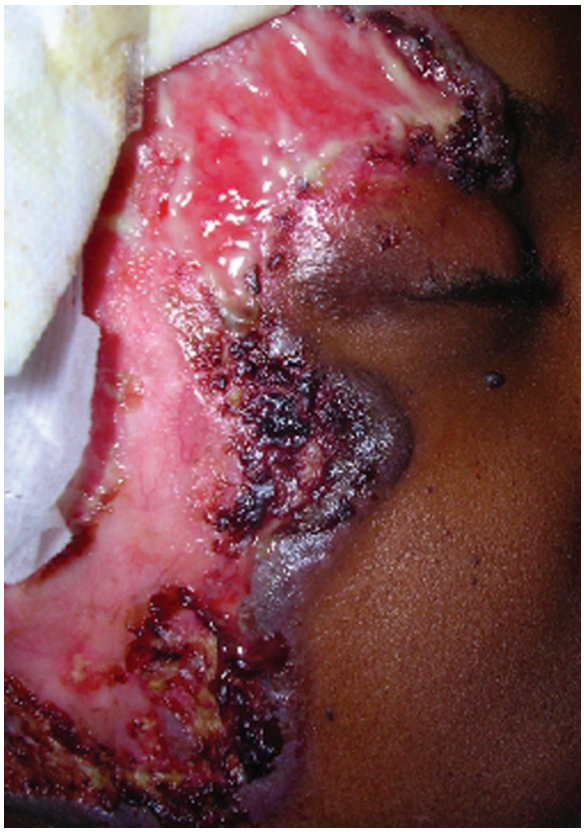

FIG. 1. The well-circumscribed area of ulceration on the right side of the face involves the brow and superior portion of the upper eyelid.

FIG. 2. The same patient 13 months later following rapid progression of her disease despite aggressive medical and surgical therapy.

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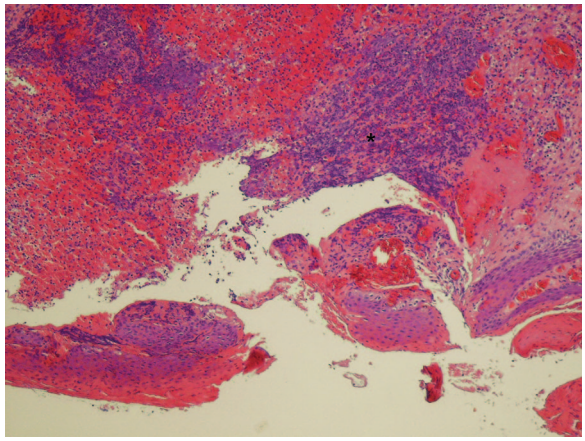

FIG. 3. The biopsy shows ulceration (*) and a neutrophilic infiltrate in the dermis with associated hemorrhage (hematoxylin-eosin, 25×).

DISCUSSION

This case highlights the difficulty of managing periocular PG. In general, surgery is used with great reluctance in patients with PG because of the risk of pathergy, a phenomenon in which progressive destruction of healthy-appearing skin may be triggered by minor trauma.[3] Our patient's lagophthalmos and associated corneal exposure necessitated intervention to prevent further ophthalmic complications, despite the risk of increased disease activity at the surgical sites. In our case, the patient's dermatologist was particularly concerned that PG would affect previously unaffected skin graft donor sites, but this never occurred despite numerous graft harvests from several separate sites.

In the largest series of periocular PG reported to date, Rose et al. describe 4 patients with varying degrees of eyelid involvement by PG. Of these, 2 underwent eyelid reconstruction using flaps or grafts, and the authors note that PG had not recurred at 6-year follow-up in one of these patients. Although we did not biopsy our patient's skin grafts during each of her eyelid surgeries, the clinical appearance was that of recurrent PG, and this was confirmed pathologically at her last surgery. While the phenomenon of pathergy would certainly lead us to expect this to be the norm in such cases, we were unable to find a prior report of recurrence of PG in an eyelid skin graft. Given the rarity of periocular PG, this is not unexpected. Pathologic confirmation in our case lends support to achieving maximal medical control of PG prior to undertaking surgical repair of the eyelids in such patients.

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Visual Improvement After Optic Nerve Sheath Decompression in a Case of Congenital Hydrocephalus and Persistent Visual Loss Despite Intracranial Pressure Correction Via Shunting

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Abstract: **Among the sequelae of persistent raised intracranial pressure (ICP) are ophthalmologic signs and symptoms, including cranial nerve palsies, visual field deficits, papilledema, and vision loss. Elevated pressure within the optic nerve sheath may not be relieved by shunt procedures, which can decrease generalized ICP. The authors present a case of acute visual loss in the setting of chronic hydrocephalus and multiple shunt revisions. Despite shunt correction resolving systemic symptoms of raised ICP, this child had persistent visual loss. Bilateral optic nerve sheath decompression was performed, and the visual acuity improved over the next 3 days. This case highlights the importance of routine ophthalmologic examination in patients with hydrocephalus and shunts and demonstrates the utility of optic nerve sheath decompression as a surgical intervention when shunting alone does not resolve visual loss.**

While often a lifesaving means of reducing intracranial pressure (ICP), shunting procedures have a predilection for malfunction.[1] Raised ICP from shunt malfunction can cause a variety of symptoms, including visual loss from persistent papilledema. Further complicating the issue of shunt malfunction is the possibility that correction of ICP via ventriculoperitoneal (VP) shunt may not resolve elevated optic nerve sheath pressure, papilledema, or vision loss. An effective valve may exist, separating the potential space surrounding the optic nerves from that surrounding the brain itself.[2] We describe a patient in whom VP shunt malfunction produced no light perception vision, papilledema, and bilateral sixth cranial nerve palsies. Correction of raised ICP via shunt revision improved ocular motility and papilledema, but vision loss remained unchanged. Subsequently, bilateral optic nerve sheath decompressions (ONSD) were performed with partial return of vision. This case underscores the importance of ophthalmologic examinations in patients with hydrocephalus and VP shunts to detect papilledema earlier, before the dramatic consequences are apparent. In addition, it demonstrates the utility of ONSD when shunting alone does not resolve visual loss in this context.

CASE PRESENTATION

A 3-year-old boy with history of congenital hydrocephalus status post–VP shunt placement and hypoplastic left heart

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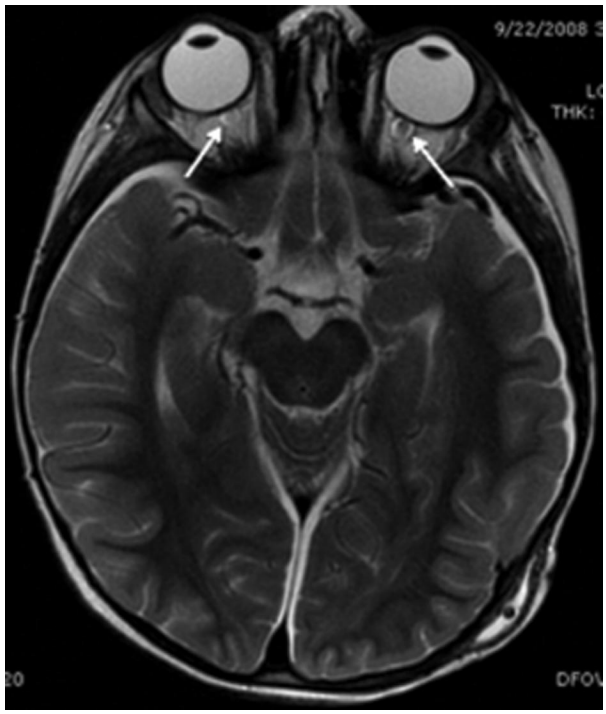

T2-weighted axial MRI of the brain showing prominent cerebrospinal fluid spaces surrounding the optic nerves (arrows).

syndrome presented with fever, headache, lethargy, and vomiting. Testing revealed an ICP of 56 cm H_20 with normal cerebrospinal fluid (CSF) constituents. No examination of the patient's fundus was performed, nor had it been in nearly 2 years. He underwent a shunt revision, but the symptoms persisted. A second shunt revision was performed with mild improvement of his systemic symptoms, but symptoms of elevated ICP returned several days later, accompanied by vision loss. On examination, the patient was found to have a visual acuity of no light perception in each eye, 7-mm fixed, dilated pupils, bilateral sixth cranial nerve palsies, and moderate to severe papilledema with elevation as well as obscuration of disc margin and vessels. He was treated with intravenous dexamethasone and acetazolamide and emergently underwent a complete valve and shunt replacement. After shunt revision, the patient's raised ICP symptoms resolved. There was mild improvement of the papilledema and resolution of the sixth nerve palsies, but the visual acuity remained no light perception OU on postoperative day 2. MRI of the brain revealed prominence of the CSF space surrounding the optic nerve heads (Fig.). There was no evidence of venous sinus thrombosis.

To reduce localized increased pressure on the optic nerves, bilateral ONSD was performed on postshunt day 3. A medial approach was used with a single longitudinal incision and use of tenotomy hook to lyse adhesions. A visible flow of CSF from the fenestration was evident, and the fenestrations were left open to freely communicate with the orbit.

The patient had an uncomplicated postoperative course with continued improvement of his previously noted signs and symptoms. In addition, on postdecompression day 3, which was also postshunt revision day 6, the patient's mother noted that he began tracking faces with his eyes, and he was able to identify some objects around his room. The visual acuity had improved to count fingers in the right eye and hand motions in the left eye.

DISCUSSION

Elevation of ICP can result in multiple neurologic and ophthalmologic sequelae, among them visual loss. Chou et al.[3] proposed that localized pressure on the optic nerve causes decreased axoplasmic flow within the nerve leading to ischemia. In a classic experiment, Seiff and Shah[4] used Bernoulli's equations of fluid dynamics to show that increasing CSF flow within or across the nerve sheath, via creation of an opening or fenestration, decreases pressure in this closed compartment. Combining this evidence with cases where localized pressure on the optic nerve was not decreased by ICP shunting alone led to the proposal that the subarachnoid space surrounding the optic nerves is separate from that encasing the brain itself.[2]

For years, this has been the rationale behind use of ONSD for treatment of visual loss in pseudotumor cerebri. Specifically, Kelman et al. showed that decompression could be used to treat visual loss even once generalized ICP has been corrected via lumboperitoneal shunting.[2] In a recent series of 3 cases, Ramsey et al.[6] demonstrated that visual function could be improved with ONSD in cases of elevated ICP. These patients, however, had not yet undergone shunt revisions for their malfunctions causing raised ICP.

We believe that in our patient, a functional valve existed between the intracranial CSF space and the optic nerve sheath space. Consequently, proven correction of ICP via VP shunting improved systemic and other neuro-ophthalmologic symptoms, but it did not fully resolve the papilledema or vision loss. We suspect that the patient had improvement in visual acuity post-ONSD due to increased flow across the nerve, thereby decreasing the optic nerve sheath pressure. This case demonstrates a role for nerve sheath decompression in cases of visual loss caused by raised ICP. Specifically, it reveals the utility of this procedure in cases where lowering of ICP via shunting of CSF has not decreased pressure on the optic nerves caused by hydrocephalus. Furthermore, it underscores the importance of routine visual assessment and fundus examinations, particularly in pediatric patients with hydrocephalus who are unable to communicate subtle visual impairment until significant visual loss has already occurred.

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Bilateral Orbital Compartment Syndrome and Blindness After Cerebral Aneurysm Repair Surgery

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Abstract: **A 56-year-old man who underwent surgical clipping of a cerebral aneurysm via a bifrontal craniotomy in a Jack-knife position complained of bilateral blindness immediately after surgery. Examinations 14 hours later revealed marked bilateral proptosis with eyelid edema and conjunctival chemosis. MRI demonstrated severe bilateral globe tenting with marked orbital tissue edema. Despite emergent lateral canthotomy and cantholysis, there was no visual recovery. This is a rare case of bilateral irreversible blindness after cerebrovascular surgery caused by orbital compartment syndrome and resulting in severe globe tenting and ocular ischemia. Attention to factors such as direct ocular pressure from skin flaps, congestion from head positioning, and adequate intraoperative eye protection and monitoring may reduce the risk of this complication or allow faster management when diagnosed.**

Perioperative visual loss during nonocular surgery is a devastating complication. The main causes are ischemic optic neuropathy, central retinal artery occlusion, and occipital lobe infarction in spinal, cardiovascular, and radical neck surgery.[1] However, acute orbital compartment syndrome with increased orbital pressure resulting in globe tenting (conical shape deformation)[2] is an extremely rare cause of perioperative visual loss. We found only 3 reports of unilateral orbital compartment syndrome with globe tenting,[3–5] but no such bilateral cases were previously reported.

CASE REPORT

A 56-year-old obese and hypertensive man was urgently admitted with subarachnoid hemorrhage from a ruptured aneurysm of the anterior communicating artery. Preoperative CT revealed a bilateral mild proptosis (Fig. 1A) and subarachnoid hemorrhage (Fig. 1B). The patient underwent urgent clipping surgery via a bifrontal craniotomy under general anesthesia, while in the head-down "Jack-knife" position for 9 hours. Surgery started with a coronal incision followed by folding the cranial skin flap over both eyes without any eye protection. The clipping surgery was successful without intraoperative injury to the ophthalmic artery, ophthalmic vein, or the cavernous sinus. Intraoperative systolic blood pressure was maintained at 100 to 160 mm Hg, without deliberate hypotension, vasopressors administration, or blood transfusions. Fluid infusion was estimated at 2900 ml and urinary volume at 1700 ml.

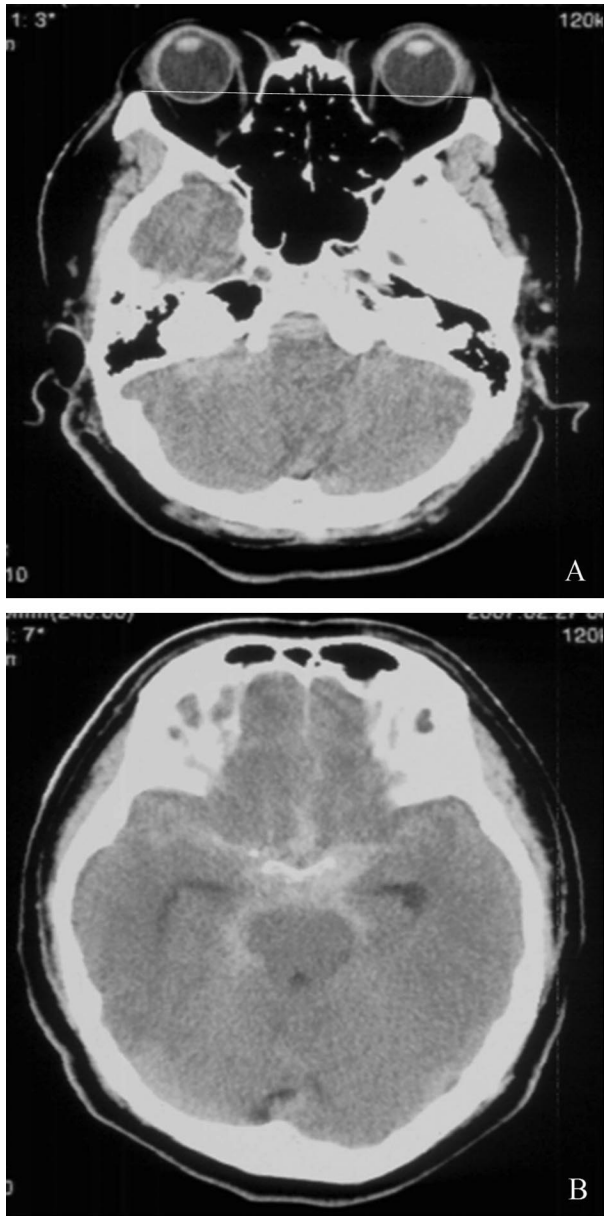

FIG. 1. Axial CT scans before clipping surgery (**A,** orbital image; **B,** brain image). Bilateral slightly prominent globes (the distance from the interzygomatic line to the posterior margin of the globe: 5 mm) are shown with no other orbital pathology **(A)**. Subarachnoid hemorrhage in the suprasellar cistern is demonstrated **(B)**.

Immediately after recovery from anesthesia, the patient complained of bilateral blindness and ocular pain. On referral to us 14 hours after surgery, ophthalmic examination revealed bilateral marked proptosis, eyelid edema, and conjunctival chemosis (Fig. 2). Excessive eyelid tension prevented eye opening. Urgent MRI showed severe bilateral globe tenting with an angle of 90° between the posterior sclerae on both sides of the optic nerve (Fig. 3A). In addition, there was bilateral orbital tissue edema, and the right superior ophthalmic vein was enlarged without cavernous sinus thrombosis (Fig. 3B).

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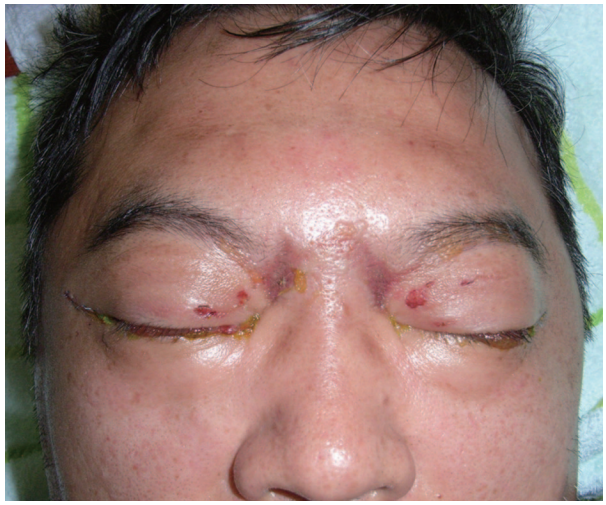

FIG. 2. Facial appearance after the clipping surgery. Proptosis, eyelid edema, and conjunctival chemosis are shown bilaterally.

An emergent lateral canthotomy and cantholysis were performed under local anesthesia, resulting in decreased eyelid tension. At intraoperative examination, vision was no light perception OU, and intraocular pressure was 24 mm Hg OD and 20 mm Hg OS. Exophthalmometry measured 20 mm OD and 22 mm OS. There was complete ophthalmoplegia. Ocular examination revealed bilateral Descemet's folds, dilated and nonreactive pupils, and macular cherry-red spots. Steroid pulse therapy (methylpredonisolone 1 g/day) was then administered for 3 days. Despite prompt resolution of periocular edema, neither patient's vision nor ocular motility improved. At last follow-up examination, 18 months later, the visual acuity was no light perception. Bilateral atrophies of the retinal pigment epithelia, choroids, and optic nerves were observed along with the choroidal pigmentations, which indicated the proximal involvement of the ophthalmic artery occlusion.

DISCUSSION

We present a rare case of bilateral blindness caused by orbital compartment syndrome with severe globe tenting as a complication of clipping surgery for a cerebral aneurysm.

Although bilateral visual loss is generally attributed to systemic etiologies,[1] local factors should also be considered. The present case involved several such significant factors; the first was preoperative bilateral prominent globes, which was not pathological, but could have been a predisposing risk factor for easier globe compression by the folded cranial skin flap.[1] Intraoperative head-down position and increased abdominal pressure during the Jack-knife position, and increased intracranial pressure, could also contribute to orbital congestion.[1] Then, the drastic changes in external pressure after skin flap unfolding and repositioning, and normalizing intracranial pressure, may have dilated vascular channels, which resulted in fluid transudation in the orbital space.[6] Large volume infusions may have also played a role in orbital congestion.[3]

Although the most common cause of bilateral perioperative visual loss is ischemic optic neuropathy, the degree of visual deterioration can sometimes be mild and may even gradually improve.[1] With orbital compartment syndrome, when globe tenting is severe (the posterior globe angle $<120°$), the visual prognosis is very poor despite urgent orbital decompression.[2] All reviewed cases of this syndrome with severe unilateral globe tenting were characterized by irreversible blindness.[3–5]

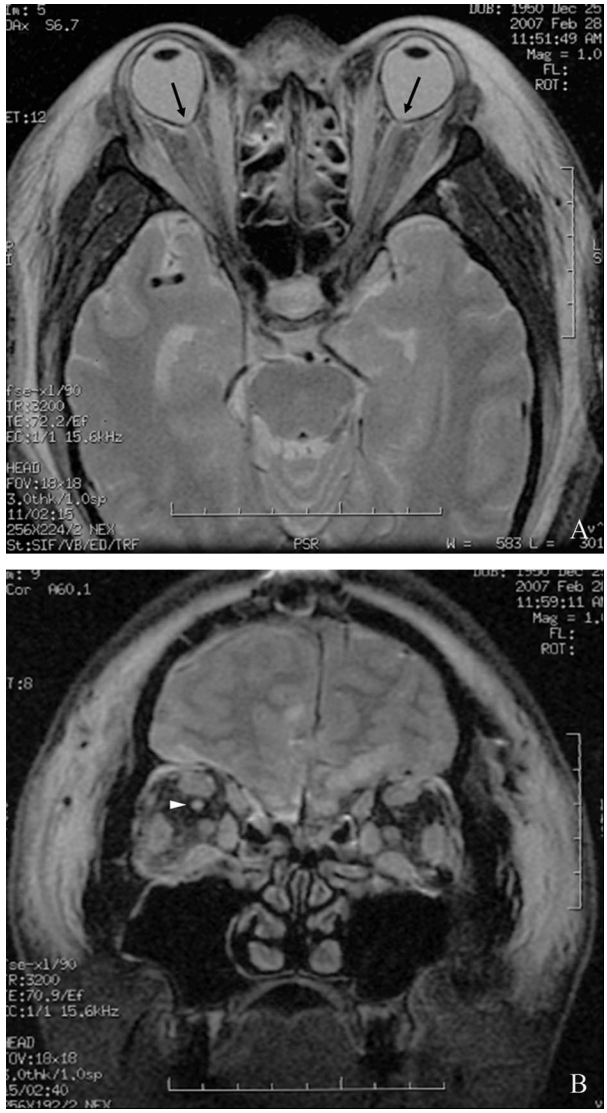

FIG. 3. T2-weighted magnetic resonance images after clipping surgery (**A,** axial view; **B,** coronal view). Severe bilateral globe tenting (**A,** arrow) and orbital tissue edema with the dilated right superior ophthalmic vein (**B,** arrow head) are seen.

Perioperative visual loss in cerebrovascular surgery was not given much attention previously. Perioperative vision loss largely occurred in spine surgeries,[1] while cerebrovascular disease is related to smoking or systemic diseases such as hypertension, diabetes mellitus, cardiovascular diseases, and obesity. These are all known risk factors for perioperative visual loss,[1] making it more important to monitor visual signs and symptoms in patients with these risk factors who are undergoing cerebrovascular surgery.

Physicians should be aware of this possible complication during cerebrovascular surgery. Attention to factors, such as direct ocular pressure from skin flaps, congestion from head positioning, and adequate intraoperative eye protection and monitoring, may reduce the risk or allow faster management.

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Spontaneous Orbital Floor Fracture in Thyroid Eye Disease

Mohsen Bahmani Kashkouli, M.D., and Farzad Pakdel, M.D.†*

Abstract: **Spontaneous orbital fracture is a rare entity. A 32-year-old man presented with thyroid eye disease and was found to have an orbital floor fracture on routine preoperative imaging without any antecedent relevant symptom or sign. To the best of the authors' knowledge, this is the first report of an asymptomatic orbital fracture in a patient with thyroid eye disease.**

Although traumatic blowout fracture of the orbital floor is a well-recognized entity caused by force impaction to orbital soft tissue or periorbital area, there are few nontraumatic causes including nose blowing[1] and neoplasm.[2] Previously, spontaneous orbital floor fracture has been reported in a patient with thyroid eye disease (TED) who developed pain, bruising, and infraorbital hypoesthesia without antecedent trauma.[3]

Here, we report a patient with TED who had asymptomatic spontaneous orbital floor fracture found on preoperative routine workup for cosmetic orbital decompression. To the best of our knowledge, this is the first asymptomatic spontaneous orbital fracture in a patient with TED.

CASE REPORT

The patient was a 32-year-old nonsmoking man who had Graves hyperthyroidism for 3 years and TED for 8 months (after receiving radioactive iodine treatment) before his referral for cosmetic orbital decompression in January 2009. He was taking oral levothyroxine and was biochemically and clinically euthyroid at the time of first visit. He had received 2 courses of oral steroid treatment starting with 50 mg/day and tapered off within 6 to 8 weeks during the past 8 months. On the first TED clinic visit, optic nerve function tests were within normal limits. Hertel exophthalmometry (base = 111) showed an eye protrusion of 27 mm on both eyes. There was a bilateral upper and lower eyelid retraction and a slight restriction of extreme upward gaze in both eyes but no complaint of diplopia. The clinical activity score was 1.

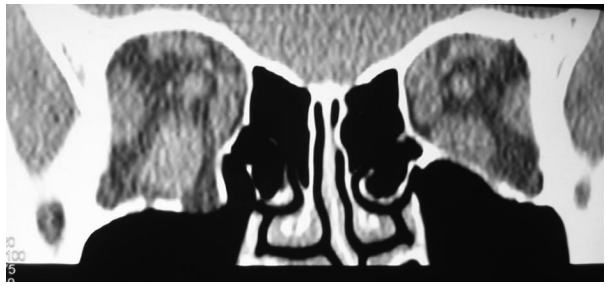

FIG. 1. CT scan of the mid-orbit section (coronal view) shows a right floor fracture medial to inferior orbital groove.

He was scheduled based on our protocol (burnt-out state for 4–6 months) for cosmetic orbital decompression. A conservative management, including eye lubricant, was followed during his preoperative time. In preparation for decompression, an orbital CT scan was requested in May 2009. CT scan revealed a right bony orbital floor fracture and protrusion of periorbital tissue in the maxillary sinus just medial to the inferior neurovascular bundle in the posteromedial part of the floor, without orbital and paranasal sinus pathologic findings (Fig. 1). The patient denied any orbital-facial trauma, sudden orbital pain, periorbital bruise, diplopia, or any sudden change in eye appearance. Hertel exophthalmometry showed 1-mm reduction of proptosis on the right side (Fig. 2). Cheek sensation test showed a mild decreased sensation on the right in comparison with the left side. Limitation of upward gaze was slightly more on the right side. Other examinations showed no significant change since the previous visit. Bone densitometry with dual energy x-ray absorptiometry method showed osteopenia of the lumbar spine. Blood tests for calcium, phosphorous, and alkaline phosphatase were within normal limits.

He underwent a successful bilateral simultaneous transeyelid orbital decompression. On dissection of the right orbital floor periosteum, a fracture was found medial to the neurovascular bundle with periorbita and fat entrapment but no inferior rectus entrapment or change in color.

DISCUSSION

Traumatic orbital floor fracture is assumed to result from either hydraulic (increased orbital pressure causing the floor

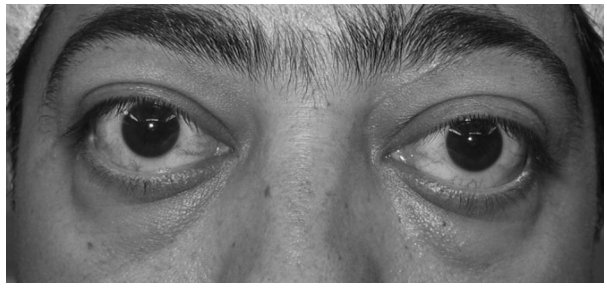

FIG. 2. Face photography of a patient with thyroid eye disease shows proptosis and eyelid retraction, less on the right side.

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fracture) and/or buckling (direct transmission of the force from the rim to the floor) mechanism.[4] In our patient without any history of trauma, explaining the mechanism of fracture, the hydraulic theory sounds more plausible. In addition, the naturally thin orbital floor can be further weakened by other local and systemic factors in TED.

In the presenting case, fracture of the orbital floor might be secondary to the local compressive effect of the enlarged inferior rectus muscle that could be accentuated by increased diffuse intraorbital pressure demonstrated in patients with TED.[5] Bone remodeling was evident in the medial orbital walls, supporting the idea of progressive effect of increased intraorbital pressure on the bony walls. Additionally, the CT scan showed greater enlargement of the right inferior rectus than of other extraocular muscles (Fig. 1).

Moreover, there is an imbalance between bone resorption and formation in hyperthyroid state that might result in decreased bone mineral density, hence an increased risk for osteoporotic and bone fracture.[6,7] Although antithyroid treatment is able to reduce this effect, previous hyperthyroidism is independently associated with an increased risk of fracture.[6,7] Furthermore, corticosteroid therapy increases the risk osteoporosis and bone fracture.[8] Our patient showed a mild osteopenic state in the bone densitometery test.

We believe in one or more of the following explanations for the spontaneous fracture in this case: increased intraorbital pressure may have exceeded the bone tolerance in the weakest point (postero-medial part of the floor) after a gradual remodeling of the floor; minor trauma such as rubbing of the eye, that was not recalled by the patient, might have pushed the globe back and increased the orbital pressure suddenly; steroid therapy may have compromised the bone density; and hyperthyroid state, low thyroid-stimulating hormone, and antithyroid therapy may have had a deleterious effect on the bone mass.[6]

Bhermi et al.[3] reported a case of Graves ophthalmopathy with proptosis, ocular motility restriction, and optic neuropathy, and the patient developed a sudden symptomatic obital floor fracture and soft tissue prolapsed in the maxillary sinus on awaking without a history of trauma. Absence of symptoms in our patient may be explained partly by a relatively small size of the fracture and possible occurrence of very mild and transient symptoms, which was not recalled by the patient.

In conclusion, although a patient with TED may develop asymptomatic spontaneous orbital floor fracture, review of literature and our presenting case propose the following signs to be considered as a clue for possible orbital fracture in patients with TED: sudden eyelid bruise and crepitations; unexpected unilateral limitation of movement in upward gaze; unexpected decrease of proptosis; and asymmetric cheek sensation. An orbital CT scan is recommended to evaluate for possible fracture.

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A Case of Acute Reversible Charles Bonnet Syndrome Following Postsurgical Unilateral Eye Patch Placement

Nicole Miriam Khadavi, B.S., Helen Lew, M.D.*, Robert Alan Goldberg, M.D.*†, and Ronald Mancini, M.D.*†*

Abstract: **A fully alert 70-year-old male with no significant medical or psychiatric history presented for surgical follow-up after uncomplicated right lower eyelid cicatricial ectropion surgery with postoperative unilateral, eye patch placement complaining of visual hallucinations. Preoperative unaided visual acuity was 20/20 in each eye. The patient described simple, nonformed and complex, formed images that were both static and animated. The images included crystal-like formations that appeared to bubble, green leaves against a vivid magenta backdrop, and an isolated hallucination of a lifelike plant with trembling leaves. These hallucinations began 2 days postoperatively and persisted 2 days following eye patch removal. The patient perceived the hallucinations multiple times a day over the 7-day period, without a stereotyped pattern. The images occurred when the eyes were open and ceased when they were closed. They were prompted by looking at a blank wall or white surface. The patient consistently recognized these images as unreal. They typically persisted for 1 to 2 minutes and could be extinguished by looking away. There were no associated auditory hallucinations, psychosis, or delirium and no history of visual, cognitive, or neurological deficit. The patient denied the use of hallucinogenic medications, including analgesics, or the initiation of any new medications. To the authors' knowledge, this is the first reported case of acute reversible CBS following unilateral eye patch placement. CBS may be a frightening postsurgical consequence of eye patch placement. It is important that the ophthalmic surgeon be aware of the potential for development of CBS and offer appropriate referral and reassurance should it occur.**

The Charles Bonnet syndrome (CBS) describes the development of visual hallucinations in patients with visual acuity loss or visual field deficits. Loss of input to the brain caused by a disorder in the visual system from the globe to the cortex may be the cause of CBS. Two commonly accepted theories for the visual hallucinations in CBS are the release and deprivation theories.[1] The release theory sug-

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gests that hallucinations result from anomalous signals to the visual cortex from lesions in the visual pathway. The deprivation theory implicates spontaneous image formation in the visual cortex in the setting of sensory deprivation as the cause of visual hallucination formation. More recently, it has been suggested that CBS hallucinations result from dynamic changes in visual perception and cease with stabilization or complete blindness.[2]

The demographic most commonly described in CBS are patients older than 64 years, socially isolated, of lower cognitive function, and with decreased bilateral visual acuity.[1] However, evidence for the role of social isolation and cognitive impairment in CBS is conflicting. The absence of cognitive impairment in CBS patients has been demonstrated and suggested as a criterion for diagnosis[3] despite studies, which identify early cognitive impairment as a risk factor.[4] It has been suggested that it is the perception of loneliness and personality traits such as shyness and low extraversion, rather than social isolation quantified by the number of social contacts, that is associated with CBS.[5] Age-related macular degeneration, diabetic retinopathy, dense cataracts, and glaucoma are the traditional underlying ocular pathologies that result in CBS visual hallucinations. While CBS is typically described in chronic visual disturbances, cases have been reported in acquired visual defects following neurosurgical procedures,[6,7] occipital lobe infarction,[8] enucleation,[9,10] temporal arteritis,[11,12] central nervous system tumors with and without optic nerve involvement,[13–15] acute anemia,[16] and multiple sclerosis.[17–20] In cases of acute visual deficits, CBS symptoms may develop immediately or with a latency of several days to months.[2]

Symptoms most commonly occur on awakening and may display a consistent pattern in terms of frequency, timing, and duration.[1] The most frequently perceived images are of people, plants, and geometric patterns that are typically in the central visual field (85%), in color (72%), and associated with moving parts (63%).[21] Here, we describe a clinical presentation consistent with CBS following postsurgical unilateral eye patch placement. To our knowledge, this is the first reported case of CBS in this setting.

CASE

A 70-year-old male with no significant medical or psychiatric history underwent right lower eyelid cicatricial ectropion with anterior lamellar shortage repair via full-thickness skin grafting, frost suture placement, and unilateral eye patch. His preoperative unaided visual acuity was 20/20 in each eye, and he had no history of ophthalmic pathology. He began to experience visual hallucinations on postoperative day 2. The patient described the hallucinations as vivid and lifelike with notably intense coloration. They typically occurred while focused on a blank white surface and could be induced to recur. The patient described 2 daily recurring images: the first was of a twisting vine with large green leaves against a vivid magenta background. He recounted that this image occurred on awakening when his gaze was focused at the upper corner of the blank white wall of his bedroom. The second was induced each time he looked in a basin of water such as the bathroom sink. The patient stated that he would visualize large white, black, and gold crystals that appeared to bubble. He also reported an isolated hallucination of a 4-foot-high plant with its leaves trembling as though in a light breeze, that appeared so lifelike that he was prompted to reach out and touch the plant despite realizing it was a hallucination. He denied any familiarity to these images. The images persisted for 1 to 2 minutes and occurred with eyes open. They could be extinguished by looking away or closing his eyes.

The patient lives independently, and the hallucinations always occurred when the patient was alone. There were no associated auditory hallucinations, psychosis, or delirium. He denied the use of hallucinogenic medications, including analgesia, or the initiation of any new medications. The patient reported anxiety surrounding the onset of the hallucinations and hesitancy to report the symptoms. He presented to his primary care physician on postoperative day 4, 2 days after the onset of hallucinations, and a complete physical examination and laboratory evaluation were performed without any abnormalities noted. The patch was left in place, and he was told to follow-up as scheduled with his surgeon. On examination by the operating surgeon on postoperative day 7, the patient was fully oriented, and no cognitive impairment was noted. He was neurologically intact and without visual deficit. The eye patch was removed, the likely diagnosis of CBS was discussed in depth with the patient. He was reassured and sent home with close follow-up. The patient reported almost immediate, progressive decline in the frequency of visual hallucinations with complete resolution at 2 days posteye patch removal. At 6 months postsurgery the patient denies any recurrent hallucinations or sequelae.

DISCUSSION

Despite more widespread recognition in cases of chronic visual impairment, CBS following acute deficit to the visual pathway has been reported.[6–19] It is hypothesized that resolution occurs with stabilization in visual symptoms.[2] However, the natural history of CBS has not been clearly elucidated, and there is no strong evidence for resolution of symptoms with duration of disease.[21] Here, we suggest that postsurgical eye patch placement may mimic acute visual deficit and results in development of the CBS. To our knowledge, this is the first reported case of acute reversible CBS following postsurgical unilateral eye patch placement. CBS is a clinical diagnosis of exclusion requiring appropriate ophthalmic and neurological workup. It can be differentiated from "black patch psychosis," which is a state of delirium occurring in the postoperative period when bilateral eye patches have been placed,[22] in that the hallucinations of CBS present in the absence of delirium. Although not functionally disabling, the hallucinations may be a source of significant anxiety. Behavioral modifications and treatment with antipsychotics, antiepileptics, and anticholinesterase inhibitors have been suggested although not proven.[23] It is important that the ophthalmic surgeon recognize CBS as a potential and frightening postoperative complication of eye patch placement and offer appropriate referral and reassurance should it occur.

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Recurrent Anophthalmic Socket Infections Due to Leakage of Reposited Infectious Fluid From a Microfractured Glass Orbital Implant

Christopher M. DeBacker, M.D.

Abstract: **Recurrent anophthalmic socket infections resulted in spontaneous extrusion of a spherical glass implant filled with sequestered fluid. Orbital imaging had revealed a fluid collection within the implant. While rarely used in developed countries, these implants are commonly used around the world due to their low cost and success in anophthalmic socket rehabilitation. Infection within an anophthalmic socket should alert the practitioner to the possibility of sequestered infectious fluid within these types of implant through a fractured breach.**

Orbital implants are essential for adequate rehabilitation of the anophthalmic socket. Alloplastic implants have been used for this purpose for well over a century with generally acceptable outcomes. Any foreign body in the orbit may act as a nidus for infection, however, and the present case describes a previously unreported etiology of implant-related infection in an anophthalmic socket.

CASE REPORT

An otherwise healthy 73-year-old man presented with signs of orbital cellulitis in his anophthalmic socket. The orbit was tense and tender to palpation, with difficulty retaining his prosthesis. The eye had been removed 36 years prior, secondary to trauma, and an implant was placed at the time of enucleation. He denied recent trauma to the affected side. A course of oral cephalexin was prescribed, and the infection resolved. Three weeks later, he presented with similar findings, and a second course of antibiotics led to resolution of the infection.

He developed a third bout of orbital cellulitis 3 weeks later. A 3-mm bulbar conjunctival fistula was seen inferiorly, and purulo-serous fluid was expressed with pressure. The transparent implant was visualized behind the dehiscence. A CT was obtained, which showed clear sinuses, soft tissue changes consistent with inflammation, and an air-fluid level within a hollow orbital implant (Fig. 1). The patient was informed that the implant would need to be removed. Two days later, the implant spontaneously extruded at home. He presented with quieting of the orbital inflammation. He brought the hollow glass implant that was filled with a yellow turbid fluid, which was slowly leaking through a small fracture in the wall of the implant (Fig. 2). The fluid was sent for culture and cytopathology. No growth was seen on bacterial culture, and cytology revealed macrophages and lymphocytes, but no malignant cells. The inflammation has since resolved, and future secondary implant placement is anticipated.

DISCUSSION

Hollow glass spherical orbital implants are rarely used today in the United States; however, they were one of the first implants developed for anophthalmic socket rehabilitation.[1] They are still used successfully in other parts of the world due to their low cost.[2] They are well tolerated with safety profiles similar to the current silicon and acrylic spheres. They are much lighter in weight compared

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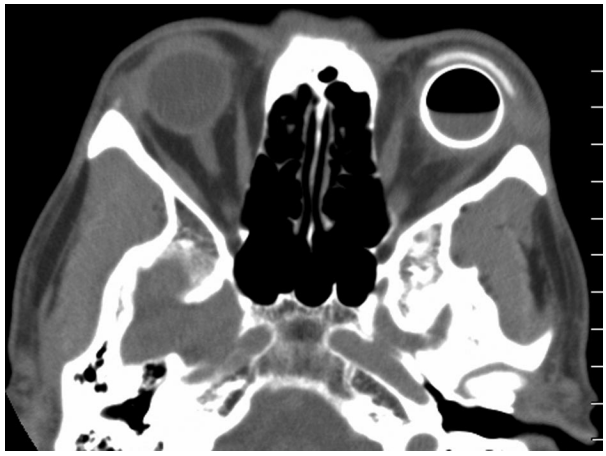

FIG. 1. Axial CT. Soft tissue changes consistent with inflammation and cellulitis are visible around the implant in the left orbit. The hollow glass sphere is readily visible with its internal air-fluid level.

with other implants, with very thin yet remarkably sturdy glass walls.

The patient had no issues with this implant for over 30 years. It is hypothesized that the recurrent orbital cellulitis was due to sequestration of fluid in the implant from an idiopathic microfracture in its wall, with bacterial colonization and growth in the stagnant fluid cavity, and subsequent fluid exchange between the implant and the orbit. Eventual fistula formation through the thinned inflamed bulbar conjunctiva led to rapid dehiscence and implant extrusion. Nonporous orbital implants are known to similarly extrude over short time frames once fistulous tracts develop.[3] An alternative explanation is that an unrecognized occult conjunctival fistula led to recurrent orbital infections, and the fluid accumulation within the fractured implant was incidental to this.

The current case describes recurrent orbital infections in an anophthalmic socket, which was felt to be due to sequestered infectious fluid in a hollow glass implant through a microfracture in the implant wall. Orbital cellulitis in an anophthalmic socket should alert the practitioner to this etiology when enucleation was performed with a hollow glass implant.

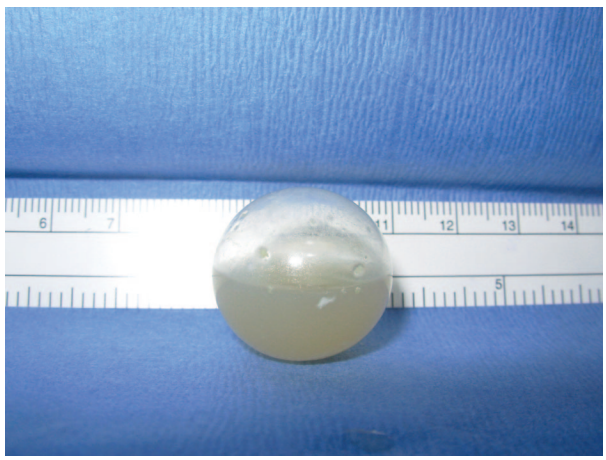

FIG. 2. Extruded glass orbital implant with sequestered purulo-serous fluid.

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Cutaneous *Sporothrix schenckii* of the Human Eyelid

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Abstract: **An 87-year-old patient presented with a 6-week history of an isolated progressive destructive nodular eyelid mass, secondary nodular and ulcerative lesions, and regional painful lymphadenopathy. After 4 weeks, fungal cultures demonstrated *Sporothrix schenckii*. *S. schenckii* is a rare dimorphic fungus that can occasionally involve the periocular skin. The authors' case demonstrates typical clinical features, emphasizes the delay in diagnosis, and shows effective treatment with oral itraconazole.**

An 87-year-old female presented with a 6-week history of a worsening painful nodular lesion of the temporal right eyelid that had progressed to include multiple secondary nodular and ulcerative "satellite" skin lesions (Fig. 1). Eyelid mattering, secondary cicatricial lower eyelid ectropion, and conjunctival injection were also present. The right submandibular lymph node was enlarged and painful. She denied any history of trauma, fevers, or chills. She was taking oral doxycycline and rifampin and was getting worse. These drugs had been prescribed elsewhere after cephalosporin antibiotics had failed to halt the progression of symptoms and findings. Her medical history included only hyperlipidemia and essential hypertension.

Excisional biopsy of the nodular eyelid lesion showed a granulomatous, focally necrotizing, dermatitis with hematoxylin-eosin and Gomori methamine silver stain. Acid-fast and Warthin-Starry stainings showed no evidence of mycobacterial or *Bartonella henslae* infection. Initial viral cultures for herpes simplex, aerobic and anaerobic bacterial cultures, and fungal cultures were all negative. CT showed only mild right periorbital edema, with no posterior abscess or extension. The patient was referred to an infectious disease specialist for further evaluation. A clinical suspicion of sporotrichosis or atypical mycobacterial infection was entertained. The patient was asked to stop rifampin and doxycycline and began twice daily doses of itraconazole 100 mg, clarithromycin 500 mg, and trimethoprim/sulfamethoxazole DS (160/800). While the initial tissue specimens showed atypical granulomatous inflammatory

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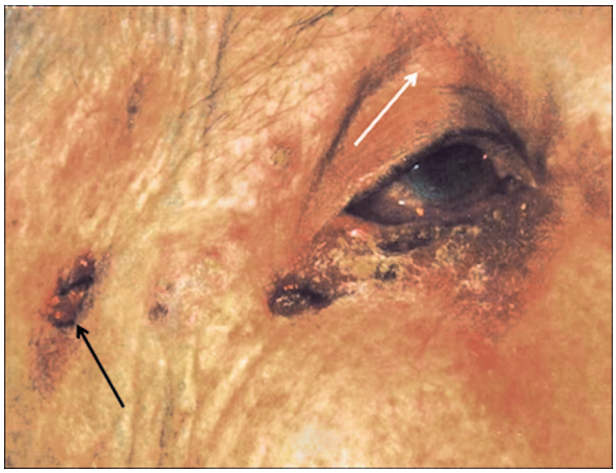

FIG. 1. Right lower eyelid ectropion with multiple satellite lesions including inflammatory nodules (white arrow) and suppurative lesions (black arrow).

changes (Fig. 2A, B), after 4 weeks, *Sporothrix schenckii* was isolated in fungal culture. Clarithromycin and trimethoprim/sulfamethoxazole were then discontinued. Itraconazole was continued for 6 months. The patient noted complete resolution of symptoms 5 months following the initial presentation.

DISCUSSION

Cutaneous sporotrichosis, classically described as "rose gardener" disease, often presents with a recent history of minor cutaneous trauma, ensuing peripheral nodular subcutaneous skin lesions, and subsequent extending lymphangitis. These lesions are usually not painful, and patients are generally otherwise healthy. Ophthalmic manifestations, relatively uncommon, have been noted in patients with disseminated infection, history of local trauma, or asymptomatic pulmonary innoculation.[1–6] Font and Jakobiec[7] described a case where the presentation of sporotrichosis was a retinochoroiditis with no prior history of systemic infection. Sporotrichosis has also been noted to cause a necrotizing ethmoid sinusitis, not unlike mucormycosis, in a diabetic patient with endophthalmitis and resulted in eventual evisceration.[8] Disseminated sporotrichosis may occur in immunocompromised persons.[9] Atraumatic ocular involvement has been seen with conjunctival lesions, and subclinical systemic infection has been documented to progress to endophthalmitis.[1] The drug of choice for most forms of sporotrichosis is itraconazole.[10] Terbinafine, potassium iodide, and fluconazole have also been used for cutaneous sporotrichosis.[9] Amphotericin B is reserved for disseminated or central nervous system sporotrichosis.[10] While relatively rare, cutaneous sporotrichosis should be considered in the differential diagnosis of periocular or eyelid lesions that progress despite oral antibiotics and even in patients with no history of trauma or a disseminated infection. Other key clinical features are development of ulcerative satellite lesions and regional lymphadenopathy, characteristic of a "sporotrichoid" pattern. The differential diagnosis for similarly presenting eyelid lesions includes herpes simplex, atypical mycobacteria, the Parinauds oculoglandular syndrome of *Bartonella henselae*, other fungal infections, and spider bite. Diagnosis of sporotrichosis through fungal culture with Sabouraud's agar, complete ophthalmic evaluation, and appropriate antifungal therapy can be potentially vision saving. Oral therapy with itraconazole is continued for a month after the disappearance of skin lesions in cutaneous sporotrichosis. While most organisms are usually identified by the laboratory within 2 to 3 weeks, *S. schenckii* may take longer. When fungal cultures do not demonstrate growth (i.e., for nearly 1 month in this case), repeat biopsy, culture, and continued monitoring are warranted.

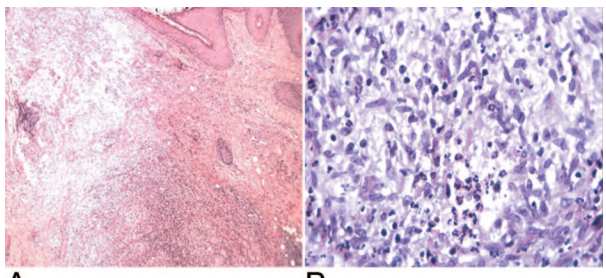

FIG. 2. Low- and high-magnification (**A**, **B**) images of right lower eyelid nodular lesion showing atypical inflammatory infiltrates.

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Perforating Globe Injury From Taser Trauma

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Abstract: **We report a case of a blinding, perforating globe injury from Taser trauma. There have been other instances**

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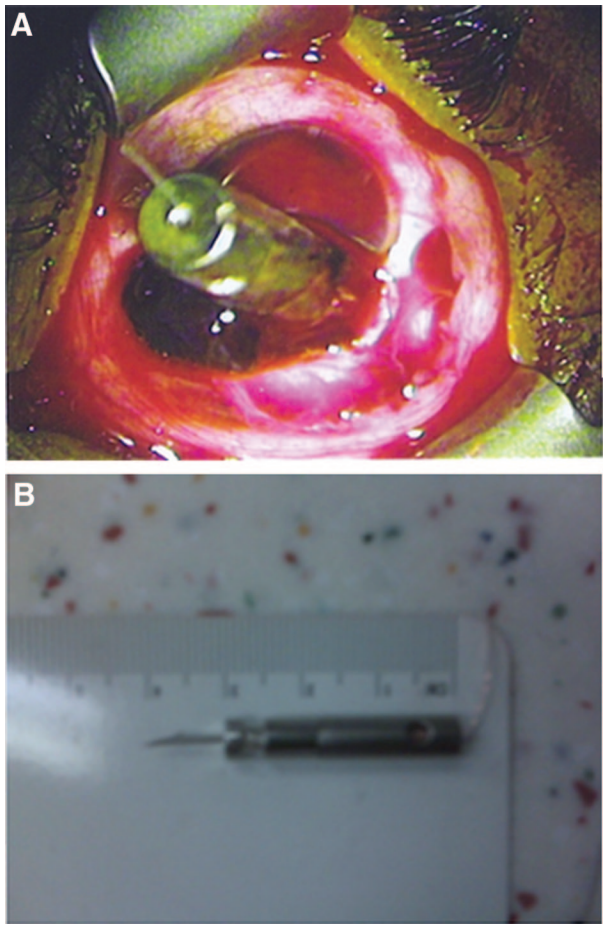

FIG. 1. **A,** External photograph of the patient's left eye, demonstrating the embedded dart and its resulting injuries. **B,** Photograph of the second dart from the pair that was given for examination prior to surgical planning. Note the sharp barb at the tip.

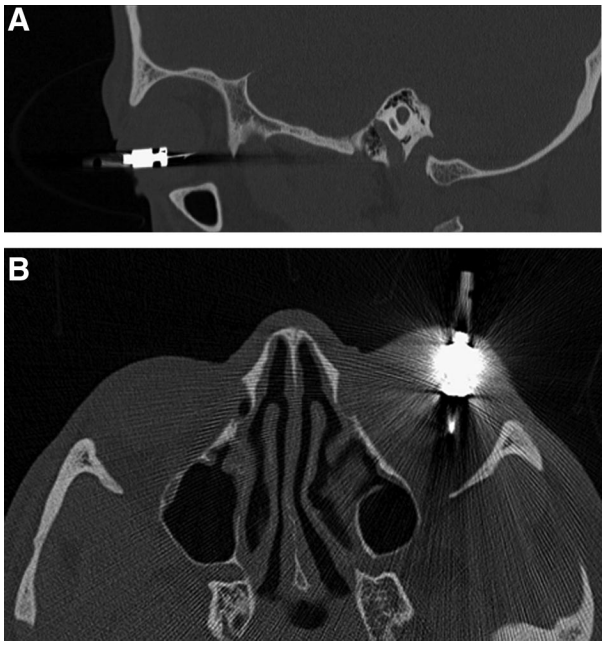

FIG. 2. **A,** CT with sagittal view illustrating the intact, penetrating dart. **B,** CT with axial view.

involving similar circumstances, but this traumatic event resulted in the loss of all meaningful vision and eventual enucleation. Despite meticulous planning and intervention, however, Taser trauma can result in devastating ocular injury.

A 26-year-old male sustained a left ocular injury from the tip of a Thomas A. Swift's Electric Rifle (Taser) dart after being involved in an incident with law enforcement personnel. Paramedics at the scene placed a small paper cup over the eye for protection and brought the patient to the local emergency room for emergent ophthalmic evaluation. On initial examination, the patient had no light perception vision in the left eye. He was found to have a cylindrical, metallic foreign body of approximately 2 cm in length and 0.5 cm in diameter protruding perpendicularly from the inferior limbus at the 6 o'clock position (Fig. 1A). The foreign body had entered the globe at the inferior limbus, extending through the anterior chamber, iris, and lens. Layered hemorrhage prevented viewing of the posterior segment. Prolapsed uveal tissue was also noted to be present at the wound site. Examination of the right eye was unremarkable.

Law enforcement officers provided the second of the pair of Taser darts for examination and surgical planning (Fig. 1B). The dart was found to be composed of 2 segments—a sharp harpoon-like barb that typically engages a subject to create a circuit for electricity to flow and a cylindrical metal support that connects the barb to the wire and provides a surface for handling. The availability of this other dart meant that the closed circuit necessary for deployment of the electrical shock between them was never complete; thus, no electrical injury occurred. The wire attached to the imbedded dart had been cut at the injury scene.

An orbital CT scan was performed to delineate the full extent of the patient's injuries (Fig. 2A–B). Sagittal and axial views demonstrated that the dart remained fully intact with the barb in place with its tip adjacent to the posterior aspect of the globe.

The patient was taken to the operating room emergently for exploration and removal of the dart. The features from the available dart provided by law enforcement were used to predict the orientation of the imbedded dart's tip since it was not visible through the injury. A careful attempt was made to remove the embedded dart without success. Unlike a regular fishhook that can be rotated to remove from tissue, the Taser tip had a sharp acute angle that did not easily permit withdrawal once engaged.

Conjunctival peritomy and posterior dissection were performed, and it was discovered that the dart had perforated the posterior sclera. The barbed end of the dart was hooked tightly to the outside of the sclera in the retrobulbar space. The entrapped dart compressed the globe through the perforation sites and had displaced the uveal tissue outward. A full-thickness posterior scleral incision around the exit point was performed to relieve the obstruction. The dart, along with the significant amount of uveal and retinal tissue, was removed though the anterior wound site. The remaining corneoscleral wounds were closed as posteriorly as possible with sutures. The patient did not recover any vision in the eye. In the immediate follow-up, enucleation was advised, but the patient refused. However, he recently decided to proceed, and enucleation was performed for his blind, painful eye.

DISCUSSION

The Taser was created in 1974 to provide law enforcement with an intermediary between a stun and handgun when attempting to control subjects. The Taser shoots 2 barbed-tip projectiles at 18.3 m/second.[1] When the 2 ends engage the subject, a closed circuit is created with the attached conduction wires and the Taser. Repetitive pulses of 50,000 volts are delivered that tetanize skeletal muscle, causing fatigue and pain.[1,2]

Ophthalmic injury by Tasers from either the initial impact of the barbed-tips or secondary to the electrical shot has been documented.[1–3] These Taser tips have caused damage to the cornea, lens, iris, and retina, while electrical shocks have resulted in cataracts. In these cases, however, the patients have ultimately been left with purposeful vision. Our case demonstrates complete visual loss from a perforating injury that included extensive uveal prolapse at the anterior entrance point and destruction of the posterior pole at the exit site. The ultimate outcome was a blind, painful eye that was enucleated. This case illustrates the extensive damage that can potentially occur from a direct Taser injury and the need to further investigate the full nature of such a trauma with appropriate imaging and surgical intervention.

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