trauma theory, external trauma or surgery causes disruption of the bone integrity, and epidermal cells are sown into the bone.[6] In our case, this could be a possible way because the patient was a coal miner, which could have caused trauma to the jaw. Higher prevalence of intraosseous epidermal inclusion cysts in male patients may be due to their higher probability to get involved in traumatic incidents compared with females. The second theory could be ectopic localization of the epidermal cells.[7] Another theory is that an epidermal cyst present in the soft tissues erodes into the underlying bone.[8]

These benign lesions grow slowly and do not cause any complaints for a long time. They are difficult to be recognized clinically. They may cause swelling, pain, and tenderness in the affected area.[9] Odontogenic keratocyst, unicystic ameloblastoma, aneurysmal bone cyst, and traumatic bone cyst should be considered in the differential diagnosis.[3]

Intraosseous epidermal cysts present radiologically as well-circumscribed, unilocular osteolytic lesions with sclerotic margins.[2] Sometimes, soft-tissue swelling may be present. A well-defined scalloped border may be present. This is thought to be the result of accelerated growth. This finding may also be present in the odontogenic keratocysts of the mandibular ramus. Odontogenic keratocysts tend to spread along the medullary space, causing only slight expansion until considerable size is reached.[10] In our case, the cyst expanded in all directions. Magnetic resonance imaging has varying signal intensity for T1-weighted images (high signal intensity is associated with hemorrhagic material in the cyst). These cysts are consistently hyperintense on T2-weighted images. High signal intensity can be seen on T1-WI from hemorrhagic or proteinaceous fluid. Contrast enhancement may be seen when the lesion is superinfected.

Intraosseous epidermal inclusion cysts are macroscopically intraosseous cysts with the diameter of several centimeters filled with white material. A connective tissue lining between the cyst and the surrounding bone may be present. Diagnosis can be verified with intraoperative frozen-section examination. Microscopically, the cyst is lined with stratified squamous cell epithelium and filled with keratinized cellular debris. There are no skin adnexa as in dermoid cysts. Odontogenic keratocysts have a keratinizing epithelial lining with distinctly palisaded cell layer in contrast to the epidermal inclusion cysts, which have epidermis and keratin layers.[10] Secondary granulomatous inflammation may be seen in ruptured cysts. Superinfection and bleeding have been described. Cholesterol granulomas, extensive bone destruction, and new periosteal bone formation may rarely be seen.[2,7] The treatment is curettage of the cyst lining and grafting if needed. No recurrence is seen with total excision. These cysts were reported to show malignant transformation.[11] Follow-up after surgical excision is necessary to detect any recurrence.

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Sinonasal Myxoma in an Infant

Ioannis A. Iatrou, DDS, MD, PhD, Nadia Theologie-Lygidakis, DDS, PhD,* Minas D. Leventis, DDS, PhD,* Catherine Michail-Strantzia, DDS, PhD†*

Abstract: Myxomas of the maxillofacial region are neoplastic entities of mesenchymal origin most often associated with odontogenic origin; sinonasal myxoma is rare, located in the nasolabial region and originating from the sinonasal tract.

The aim of the current study was to report a well-documented case of sinonasal myxoma in a 12-month-old boy, initially presenting with obliteration of his left nasolacrimal duct. A soft-tissue mass of the nasobuccal groove, firmly attached to the underlying bone, was revealed. After biopsy where benign fibroblastic elements were found, the tumor was removed surgically in wide margins, whereas great care was taken to reconstruct the involved adjacent anatomic structures and preserve facial aesthetics. Histopathologic findings were compatible with an extragnathic, nonodontogenic sinonasal myxoma originating from the nasolacrimal duct. The clinical significance of the case presented was its rather rare location and origin. Three and a half years postoperatively, functional and aesthetic results were satisfactory with no sign of recurrence. To the authors' knowledge, this is the second youngest reported case in the literature.

Key Words: Myxoma, sinonasal myxoma, children, surgical treatment

Myxomas are rare, slowly growing, benign neoplasms of mesenchymal origin showing several times locally aggressive behavior.[1] They usually develop in the muscles of the extremities, and when they appear in the face, they are mostly considered as having odontogenic origin, associated with unerupted teeth and arising from the mesenchymal portion of the tooth germ.[2] In recent studies, they have been found to represent 11.6% to 15.7% of all odontogenic

From the *University Department of Oral and Maxillofacial Surgery, Dental School of Athens; and †Department of Histopathology, Children's Hospital "P. & A. Kyriakou," Athens, Greece.
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Address correspondence and reprint requests to Ioannis A. Iatrou, 40 Asklipiou str., 11471 Athens, Greece; E-mail: iiatrou@dent.uoa.gr
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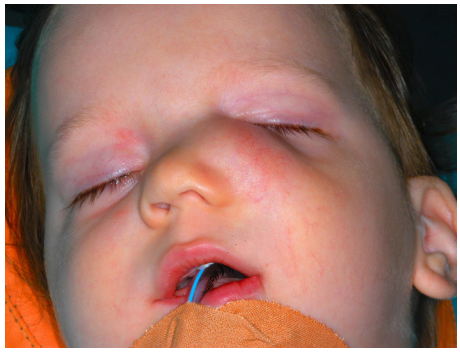

FIGURE 1. Appearance of the 12-month-old boy immediately before biopsy.

tumors.[3] A specific entity among myxomas, the sinonasal myxoma, is a nonodontogenic tumor arising either from or within the sinonasal tract. The condition is very rare and has been reported only a few times in the literature.[4,5]

The aim of this study was to present a well-documented case of a sinonasal myxoma in a 12-month-old child and discuss its characteristics and treatment.

CLINICAL REPORT

A 12-month-old white boy was referred to the Department of Oral & Maxillofacial Surgery at the Children's Hospital, with a history of swelling of the left nasobuccal groove over the last month. Clinical examination revealed an ill-defined, firmly attached, nontender mass, measuring 3 × 4 cm. The overlying skin was intact but thin and stretched by the lesion. The left eye remained partially closed because of the tumor's pressure to the lower eyelid. The nose was deviated toward the nonaffected right side (Fig. 1). The infant had been previously examined by ophthalmologists and had undergone exploration and catheterization of the left nasolacrimal duct (NLD) because of its obliteration, treatment that resulted to only temporary amelioration of the symptoms. There was no history of nasal or dental problems, although a recent ophthalmological consultation confirmed normal vision but also displacement of the ipsilateral globe.

In radiologic examination with computed tomography and magnetic resonance imaging (MRI), a relatively well-defined, oval soft-tissue mass was identified, surrounded by a thin bony layer, having intermediate homogenous appearance with its center at the

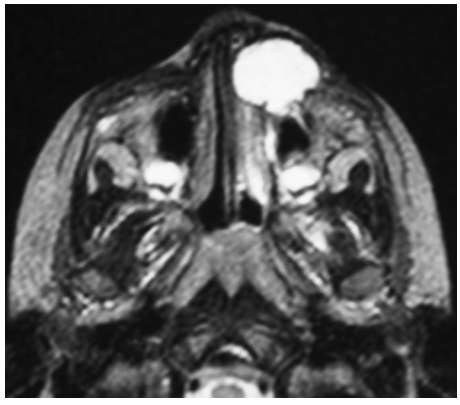

FIGURE 2. Preoperative MRI.

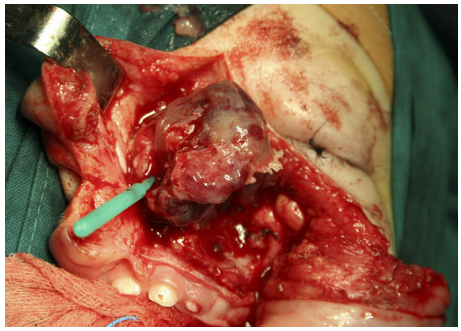

FIGURE 3. Intraoperative image.

NLD. The mass extended from the anterior aspect of the maxilla to the ipsilateral orbit, protruding to the left nasal cavity and the anterior ethmoid air cells (Fig. 2).

After biopsy and fine-needle aspiration under general anesthesia via an extraoral approach, histologic findings were compatible with a benign fibroblastic lesion. The patient was then scheduled for surgical removal of the tumor that was performed through a hemifacial flap via a Weber-Ferguson incision with Lynch extension,[3] leading to an extensive resection of the mass, including an island of intact skin (Fig. 3). The tumor was involving the left frontal process of the maxilla (the anterior and the superior wall), the left nasal bone, and the exterior part of the left middle concha, enveloping the NLD and eroding part of the suborbital rim.

The surgical specimen (3.2 × 3 × 1 cm) containing a tumor that was gelatinous in consistency was totally removed in macroscopically clear margins. In frozen-section biopsies performed during operation time, nonpositive margins were verified, and elements of a fibroblastic tumor were found. A part of the mucosa of the middle concha was preserved and sutured to restore the inner surface of the left nasal cavity. The lacrimal canaliculi were cannulated with silastic tubes (diameter 0.2 cm) for securing the function of the NLD that was removed together with the tumor. A tube of 0.5 cm in diameter was placed in the affected nasal cavity. The flap was repositioned and sutured in layers, with special care to facial aesthetics. Tubes of the lacrimal canaliculi were maintained for 60 days, and the one in the nasal cavity for 10 days. There were no postoperative complications, and a regular 3 monthly follow-up was scheduled.

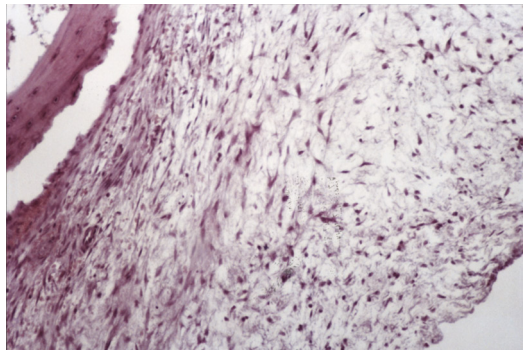

FIGURE 4. Histologic appearance of the specimen indicative of a poorly cellular nonodontogenic myxoma. The lesion is composed of widely separated stellate or spindle mesenchymal cells in a loose and myxomatous stroma and extended to a shell of woven bone. Hematoxylin-eosin stain, original magnification ×200.

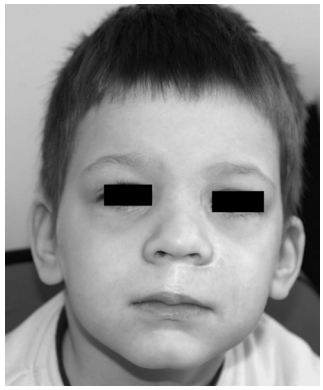

FIGURE 5. The patient 3 years postoperatively.

In histopathologic examination, the specimen consisted of a dense net of mainly spindle and secondarily stellate cells with hyperchromatic nuclei, few mitoses, and basophilic cytoplasm embedded in a loose myxomatous stroma with numerous blood vessels. The mesenchymal cells demonstrated strong and diffuse positivity in vimentin, laminin, and focal smooth muscle actin antibodies. CD31, CD34, S100, and desmin were negative; odontogenic islands of epithelium were not found (KerAE1/AE3 and epithelial membrane antigen were negative). The diagnosis of an extragnathic, nonodontogenic sinonasal myxoma was established (Fig. 4).

Three and a half years postoperatively, the functional and aesthetic results were satisfactory with no sign of recurrence (Fig. 5).

DISCUSSION

Myxomas of the face are mesenchymal tumors that may have osteogenic or odontogenic origin.[5,6] Nonodontogenic myxomas appearing in the nasolabial region are defined as sinonasal myxomas. These tumors are extremely rare, as very few well-documented cases have been reported in the literature, almost exclusively in children. To the authors' knowledge, the present case of a 12-month-old boy is the second youngest reported in the literature, the first being an 11-month-old,[7] reported in 1987 and rereported in 1993.[8]

As several tumors and tumor-like lesions present as midfacial deformities, myxomas should be differentiated from benign conditions such as fibromas or neurofibromas or from fibrous histiocytomas and malignant tumors such as rhabdomyosarcomas, fibrosarcomas, mucoid liposarcomas, and neurogenic sarcomas.[5]

Heffner[5] in 1993 was the first to underline the existing difference between myxomas of osteogenic or odontogenic origin, based on the anatomic observation of their epicenter. Following the present case where the initial finding was an obliteration of the NLD and the epicenter of the tumor was in the same area, the NLD should be added as a possible site of development of the nonodontogenic sinonasal myxomas.

To establish diagnosis of sinonasal myxomas, both computed tomography and MRI should be performed because the tumor affects both hard and soft tissues. Diagnosis can be confirmed only with fine-needle aspiration and biopsy.[2]

The histologic characteristics of the present case were similar to those previously reported[9]: not encapsulated tumor having spindle and stellate cells with hyperchromatic nuclei, few mitoses, and basophilic cytoplasm embedded in a mucoid intercellular stroma with numerous blood vessels.[2] The observed rather dense cellular network of the present case may be indicative of potential recurrence risk because dense cellularity has been found to be compatible with aggressiveness and higher risk of recurrence.[5]

The treatment of choice of the sinonasal myxoma has been reported to be the surgical excision in extended clear margins because of the local aggressiveness of the tumor,[1,6,9] as well as its potential for continuing growth, bone-destroying capacity, and recurrence rate.[5,10] Intraoperative frozen sections have been indicated to confirm healthy margins.[4] The previously described approach was applied in the present case, where a wide exposure of the tumor was achieved via a hemifacial flap, followed, because of the localization of the tumor, by an inevitable destruction and subsequent surgical removal of aesthetic anatomic structures of the middle face. Immediate reconstruction was performed with special attention to the rehabilitation of the affected nasal cavity and NLD as well as the facial aesthetics. Clinical and radiographic follow-up will continue for long because of the reported local aggressiveness.

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A Unique Presentation of Epignathus

Janna Joethy, MRCS, Yong Chen Por, FAMS,† Yeow Vincent, FAMS†*

Abstract: Palatal clefts in conjunction with space-occupying lesions of the oral or nasal cavities are of interest because they may represent a developmental etiology of palatal clefts. Epignathus is a rare

From the *Singapore General Hospital; and †KK Women's and Children's Hospital, Singapore.
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Address correspondence and reprint requests to Janna Joethy, MRCS, Singapore General Hospital, Outram Rd, Singapore 169608; E-mail: janajoethy@hotmail.com
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