Subconjunctival "Ring" Recurrence of Basal Cell Carcinoma of the Globe

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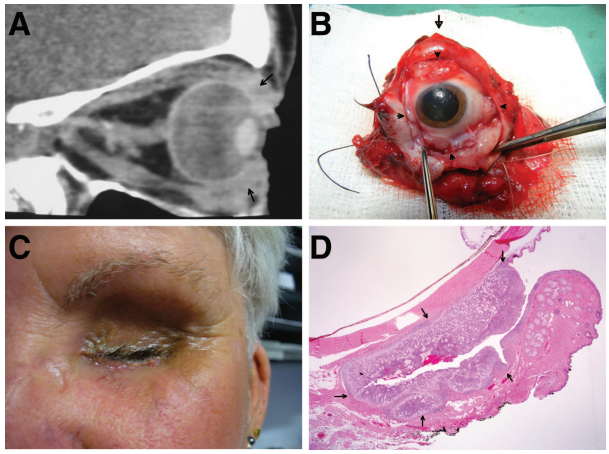
Abstract: **Basal cell carcinoma is the most common indication for orbital exenteration. The recurrence rate of BCC removed with microscopically controlled histology sections is up to 6%. The authors describe the recurrence of a lower eyelid BCC resected with microscopic control that did not manifest itself until 15 years later as a subconjunctival lesion, encircling the globe, and without apparent skin involvement. BCC can present in any manner following surgery, and therefore, judicious follow-up is necessary even after microscopically controlled resection.**

Basal cell carcinoma is the most common malignant eyelid cancer and the main cause for orbital exenteration.[1] Risk factors include chronic ultraviolet exposure, light skin pigmentation, and increasing age. It is a slowly growing cancer that rarely metastasizes. Surgical excision with microscopic control of the margin is the preferred method of treatment.[2] After excision, recurrence of a lesion is uncommon. The largest report in the literature quotes a rate of 5.36%, and of these recurrences, 60.3% were of the medial canthus. Further, after the first reoperation, the figure continues to rise to 14.7% and 40%, respectively, for each subsequent reoperation and recurrence.[3] Risk factors in this analysis were sclerosing type of basal cell carcinoma (BCC), multiple surgeries, involvement of the medial canthus, and those with deep invasion.

We present the unusual case of a BCC, where skin involvement was not present, but the lesion spread through the subconjunctival plane. Ring melanoma classically refers to the circumferential spread of a ciliary body or iris tumor. We coin the term "ring" BCC in this case to demonstrate the possible atypical presentation of a tumor that can easily be mistaken for lymphoma, amyloid deposits, simple chemosis, or retinal buckle. This lesion had no apparent skin involvement, or extension thereof, and therefore, the diagnosis was initially elusive.

CASE REPORT

A 56-year-old woman presented with a 6-month history of an irritated and painful left eye, associated with discharge and epiphora. On examination, unusual grey and salmon patches were raised circumferentially 360° around the left globe reminiscent of an encircling retinal buckle. There was some injection and chemosis with the lesions. CT showed a circumferential lesion 360° around the globe and deep regional extension in the inferior orbit (Fig. A). Her ocular history was significant for a lower eyelid resection of BCC with histologically controlled margins 15 years ago at the age of 41. A Hughes procedure reconstruction was performed at that time without event. The patient had no history of retinal surgery, lymphoma, or other known systemic disease.



A, CT shows tumor invasion in the orbit; arrows pointing to the tumor. **B,** Exenterated eye, long arrow on top pointing to the 12 o'clock position, open arrow heads marking an encircling process. **C,** Two weeks postexenteration with partial upper eyelid sparing. **D,** Histologic preparation. Arrows point to deep invasion of the basal cell carcinoma. Note that the sclera above the tumor and the skin below are uninvolved.

An incisional biopsy determined the lesion to be BCC. Surgical options were discussed with the patient, and she elected to undergo a limited exenteration. A partial eyelid sparing exenteration was performed to preserve as much of the normal remaining eyelid for reconstruction. The excised globe shows 360° involvement of the tumor mimicking an encircling retinal band (Fig. B). The cavity was closed primarily without defect (Fig. C). Pathology shows the characteristic nest of BCC nodular and infiltrative with deep invasion without eyelid involvement (Fig. D).

DISCUSSION

This case is unusual in that its presentation is atypical for most BCCs. The young age of the patient, subconjunctival spread in a circumferential manner, and lack of skin involvement at time of presentation all make for a difficult diagnosis. The lack of follow-up in the 15 years since the first surgery makes it difficult to assess the mechanism of initial spread. The literature would implicate the medial canthus as the most likely area. In this case, however, the most likely site is incomplete excision at the margins of previous resection. The relative geographic isolation of the patient also contributed to the difficulty of follow-up.

The case also demonstrates the shortcomings of frozen section analysis, especially when the tumor has spread posteriorly or toward the medial canthus. It is documented that microscopic sections (frozen and Mohs) are unable to assess tumor extension beyond conjunctiva and fatty tissue.[4] It has been suggested that surgeons can be timid in taking sections that approach the lacrimal apparatus. Further, sections taken from superficial eyelid margins are often clear, whereas posterior margins can be ragged or more difficult to obtain. This case emphasizes the need for judicious follow-up, even in cases of tumors with clear margins on paraffin sections and those with slow growth patterns such as BCC.

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Finally, exenteration in these cases demonstrates the need to individually tailor the surgery to the patient's pathology without simply categorizing the surgery in total, subtotal, or extended exenteration. In this case, the lesion was carefully demarcated to preserve the patient's upper eyelid for reconstruction.

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Spontaneous Dural Cavernous Fistula in Infancy

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Abstract: **A 4-month-old boy presented with the sudden onset of proptosis and dilated episcleral veins. CT revealed an enlarged superior ophthalmic vein and diffusely thickened extraocular muscles. MRI and angiography confirmed a dural middle meningeal—cavernous sinus fistula. Embolization was performed leading to successful resolution of the patient's signs and symptoms. Spontaneous atraumatic arteriovenous cavernous fistulae in infancy are extremely rare, but should be included in the differential diagnosis of infants with orbital congestion. Appropriate treatment leads to prevention of amblyopia, glaucoma, and anisometropia.**

Dural sinus fistula is an exceedingly rare diagnosis in childhood, particularly without associated trauma. This sparingly reported entity has not been described in the ophthalmic literature. Herein, we report the case of a 4-month-old boy with a dural sinus fistula and provide a relevant review of the disease for practicing ophthalmologists and ophthalmic plastic surgeons.

CASE REPORT

An otherwise healthy 4-month-old Hispanic boy presented to our oculoplastic service with a 10-day history of proptosis and redness of the right eye. His mother described the onset of symptoms as gradual, with a slowly progressive course. Prior to referral, he was treated by his pediatrician with Polytrim eye drops without clinical change. There was no history of fever, rash, weight loss trauma, child abuse, foreign travel, or change in behavior. Medical history was significant for bronchiolitis, for which he used albuterol, and exposure to tuberculosis in the early postnatal period. The patient's purified protein derivative test at 1 week of age was negative, and he had been treated prophylactically with oral isoniazid for 4 months. Family history was noncontributory; in particular, there was no history of thyroid disease.

FIG. 1. **A,** Preembolization view shows relative proptosis, dilated episcleral vessels, and conjunctival injection of the right eye. **B,** Complete resolution of orbital congestion 2 months after embolization.

Examination revealed symmetric visual acuity without ocular preference. Three millimeters of relative proptosis associated with dilated, prominent episcleral vasculature was noted in the right eye (Fig. 1). The eye was nontender, but displayed moderate resistance to retropulsion. Extraocular motility was full bilaterally, and the patient was orthophoric at near without correction. The pupils were normal without afferent pupillary defect. The intraocular pressure, as measured by Tonopen, was 21 mm Hg OD and 12 mm Hg OS. The penlight and dilated funduscopic examination was otherwise normal bilaterally.

CT of the orbits revealed diffuse tendon-sparing thickening of all extraocular muscles in the right eye. There was extra- and intraconal fat stranding without evidence of a discrete retrobulbar mass, abscess, or sinusitis. The superior ophthalmic vein was dilated.

The differential diagnosis included orbital cellulitis, thyroid eye disease, idiopathic orbital inflammation, and a vascular anomaly. The patient was admitted and received 36 hours of intravenous ampicillin/sulbactam (Unasyn) without clinical change. Laboratory evaluation revealed a normal complete blood count. Thyroid function tests were unremarkable. In light of the lack of response to systemic antibiotics and the orbital congestion, the patient underwent MRI and angiography, which revealed a right dural–cavernous sinus fistula (Fig. 2). At this point, the patient underwent a complete physical examination; no signs of Ehlers-Danlos Syndrome type IV, which include thin, translucent skin and extensive bruising, were noted. On the following day, the patient was successfully treated with neuroradiologic n-butyl cyanoacrylate (NBCA) glue embolization of the fistula. With the microcatheter in position, a continuous column injection of 0.4 ml radioopaque NBCA in a 50% NBCA-ethiodal combination with Tantalum powder was injected for occlusion of the fistula (Fig. 2). The patient continues to have a normal ophthalmic examination with 6 months of follow-up. The patient's extraocular motility was full, with no evidence of sixth nerve palsy, both before and after the procedure. The patient's postoperative intraocular pressure was normal.

DISCUSSION

Arteriovenous fistulas are extremely infrequent in infants. Further, the atraumatic variant is exceedingly rare. In 1981, Pang et al. reported the case of a 7-week-old infant who developed a nontraumatic fistula of the external carotid artery and the cavernous sinus. The patient underwent therapeutic

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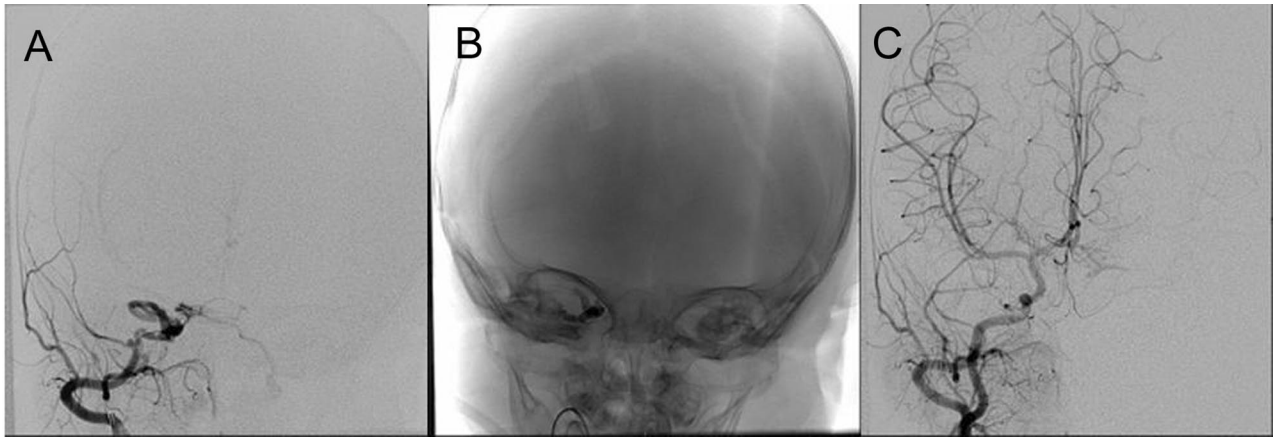

FIG. 2. **A,** Dural sinus fistula preembolization. **B,** With the microcatheter in position, a continuous column injection of 0.4 ml radiopaque NBCA in a 50% NBCA-ethiodal combination with Tantalum powder was injected for occlusion of the fistula. **C,** Immediately after embolization.

embolization in the hopes of preventing persistent proptosis, intraocular hypertension, and anisometropic amblyopia. The embolization was noted to be successful clinically and angiographically.[1] To the best of our knowledge, only 2 additional cases have been described, none of these in the ophthalmic literature. In 2004, Rai et al. reported the case of an 11-month-old girl who presented with symptoms of orbital venous congestion. An enlarged superior ophthalmic vein and cavernous sinus were seen on CT, and a direct carotid-cavernous fistula with intra- and extracranial venous drainage was confirmed on catheter angiography. The fistula was treated with coil embolization, leading to resolution of the infant's symptoms and a return of normal intraocular pressure.[2] The same year, Albayram et al. reported a similar case in a 3-month-old boy who was treated successfully with embolization using n-butyl 2-cyanoacrylate.[3]

The case of this 4-month-old boy is remarkable in terms of its rarity, dramatic clinical presentation and radiographic findings, and complete resolution of signs and symptoms with embolization treatment. Further, increasing awareness of arteriovenous fistula in infancy to the ophthalmic community is necessary because ophthalmologists, in particular oculoplastic subspecialists, will likely be the primary referral source in this patient subset. Although rare, atraumatic arteriovenous fistula in infancy should be suspected in the setting of sudden, unilateral proptosis associated with signs of orbital congestion. Proptosis and corkscrew epibulbar vessels are the most commonly seen clinical manifestations.[4] Radiographically, hallmarks include a dilated superior ophthalmic vein, extraocular muscle enlargement, and enlargement of the cavernous sinus. Although these features are reported to be seen in 80%, 65%, and 35% of patients with carotid—cavernous fistulas, all 3 features were demonstrated in this case.[5]

Cavernous fistulas are generally divided by flow characteristics. Typically, the high-flow fistulas are associated with trauma or ruptured aneurysms and create a direct connection between the internal carotid artery and the cavernous sinus. These are classified as Barrow type A fistulas.[4] The low-flow fistula are usually spontaneous and can be classified by their arterial supply: Barrow type B, meningeal branches of the internal carotid artery; Barrow type C, meningeal branches of the external carotid artery; and Barrow type D, meningeal branches of both the external and internal carotid arteries.[4] Although Barrow types B and D fistulas are thought to arise from spontaneous thrombosis, type C fistulas may be secondary to minor trauma or valsalva. This fits with anecdotal descriptions of this type of fistula being more common in younger patients, and could explain why it may be possible in infancy, where the patient may be predisposed after an episode of crying. Approximately 20% to 50% of types B to D fistulas are thought to resolve spontaneously, and clinicians can therefore consider expectant therapy in the setting of a stable ophthalmic examination. Of the 4 case reports (including the present case) documenting spontaneous fistulas in infancy, all were treated subacutely via interventional radiology. We suggest that the consideration of preventing amblyopia in the infant population— which is not as accurately examined—may, in part, account for this more aggressive approach. Additionally, as in our patient, fistulas originating from the external carotid artery typically have only one feeder vessel, making them more amenable to successful embolization.[4,5] Dural sinus fistulas can lead to an increase in the axial length of the eye, potentially resulting in anisometropic amblyopia. Although there is sparse literature in this subset of cavernous fistula patients, ophthalmologists should attempt to make an early diagnosis and consider intervention with embolization in an effort to thwart the potential sequelae of amblyopia, secondary glaucoma from elevated episcleral venous pressure, anisometropia, and corneal decompensation from persistent proptosis.

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Metaplastic Bone Formation in the Orbit

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Abstract: **Osseous or cartilaginous metaplasia in the orbit are both rare phenomena. The authors describe a unique case of combined primary orbital osseous and cartilaginous metaplasia presenting with orbital pain and ocular dysmotility and discuss the radiologic, histopathologic, and clinical features. This entity should be considered in the differential diagnosis of an orbital mass showing similar radiographic features of calcification.**

Metaplastic bone and cartilage formation in the orbit are rare, with only one reported case of heterotopic orbital ossification in the literature.[1] As with any orbital mass, depending on the location, such metaplasia may present with significant orbital pain

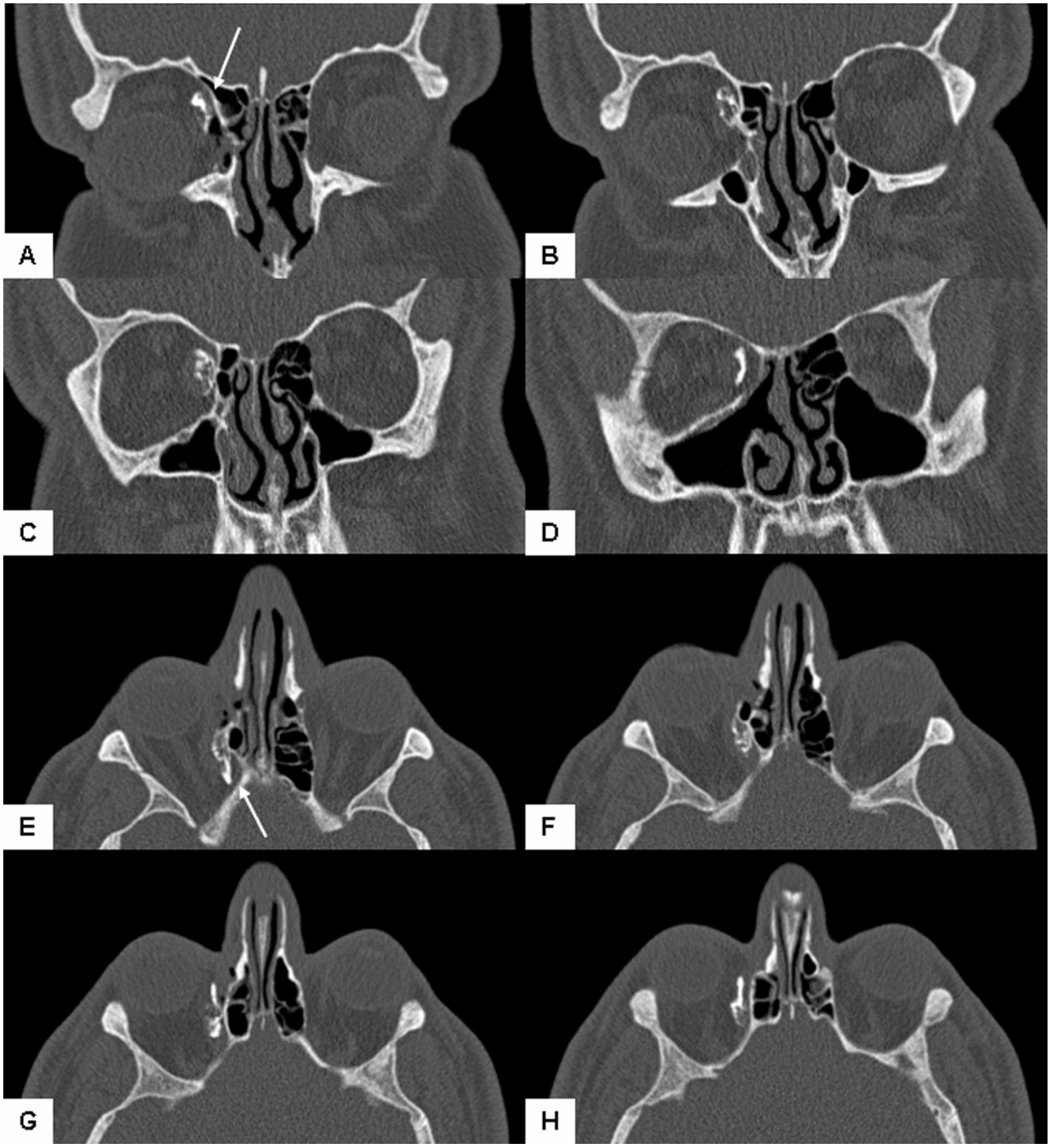

FIG. 1. Coronal **(A–D)** and axial **(E–H)** CT views show the medial orbital mass.

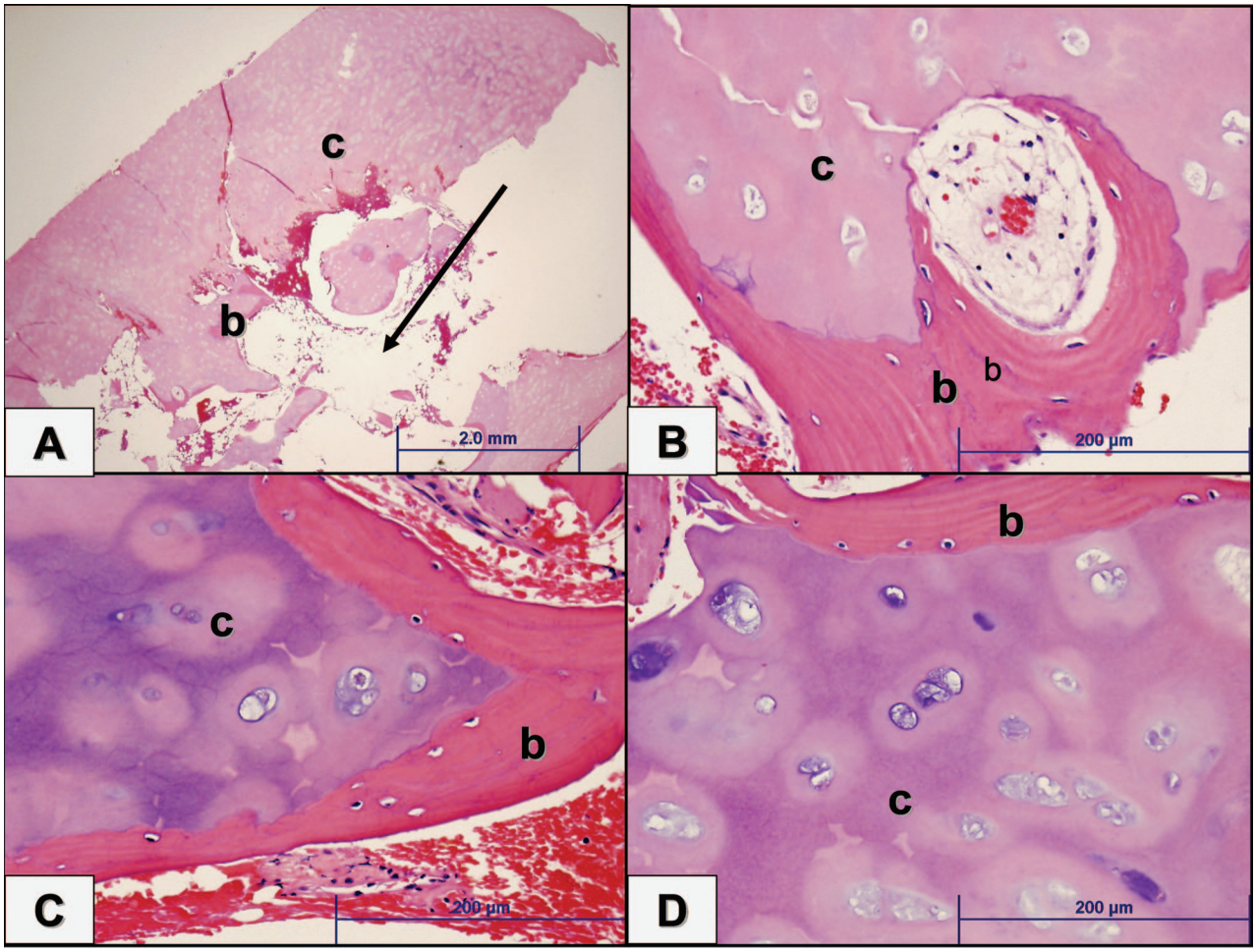

FIG. 2. Histopathologic findings of the orbital lesion. **A,** Low power view of the lesion demonstrating mostly cartilage (c) with a center of bone (b) and fibrous adipose tissue with vessels (arrow). **B,** Higher power view of the fibrovascular center surrounded by bone (b) and cartilage (c). **C** and **D,** Closeup view of the bone trabecula (b) and cartilaginous cells (c). There are few features of degeneration of cartilage, such as cloning of nuclei, but the cells are mature and without atypia.

and ocular dysmotility. The clinical presentation and the radiologic and histopathologic features of a patient with combined bone and cartilaginous metaplasia of the orbit is discussed.

A 30-year-old obese white man presented with a 4-month history of right eye pain, exacerbated by eye movement, which troubled him throughout the day and frequently awakened him at night. He had undergone a combined transconjunctival and endonasal endoscopic posterior orbital and optic canal decompression on the right side 13 years earlier for posttraumatic optic neuropathy, right medial rectus resection and lateral rectus recession 10 years ago for exotropia, and a transconjunctival medial orbitotomy for exploration and drainage of 1 ml orbital abscess 6 years ago. Ophthalmic examination was remarkable for 2 mm of right relative exophthalmos, 2 mm of right relative hypoglobus, normal fifth nerve function, no light perception OD (unchanged from his accident 13 years earlier) with a total relative afferent pupillary defect, variable right exodeviation with −½ abduction and −3 adduction OD, marked pain on eye movement, and right optic nerve pallor.

CT revealed postsurgical changes in the right maxillary sinus, right ethmoid sinuses, and posterior orbit, with a partially calcified mass in the right medial orbit abutting the medial rectus muscle (Fig. 1).

The patient underwent right transconjunctival orbital exploration with removal of a dense, partially calcified mass intimately associated with the midportion of the medial rectus muscle but also adherent to local adipose. Postoperatively, the patient enjoyed resolution of his preoperative pain and regained a normal range of horizontal movement.

Microscopically, the specimen was found to be consistent with metaplastic bone and cartilage (Fig. 2).

DISCUSSION

Metaplasia is the irreversible replacement of one mature, differentiated cell type with another secondary to a presumed stimulus.[2] Properly, metaplasia is distinguished from heteroplasia, which is the abnormal growth of mature cytologic

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elements in an abnormal location without a stimulus.[2] Many, however, use the terms interchangeably.

Bone formation through metaplasia in mesenchymal cells or in dystrophic tissues with or without prior calcification has been abundantly documented with common examples, including myositis ossificans, pulmonary ossifications, ossification in Ghon lesions (calcification seen in pulmonary parenchyma and hilar nodes resulting from tuberculosis), and in aortic atheromatous plaques.[3] There are numerous reports of heterotopic ossification in other areas of the body, including but not limited to Achilles tendon, intestinal mesentery, auricles, penis, and burn scars.[4] Metaplastic bone formation has also been associated with both benign and malignant tumors, including pituitary adenoma, benign parotid tumors, rectal and pulmonary adenocarcinoma, and basal cell carcinoma.[5]

Shankar et al.[1] reported a single case of "heterotopic ossification" of the orbit and hypothesized that the ossification may have originated from an inflammatory pseudotumor, thus more accurately metaplastic rather than heterotopic ossification. In our case, the mature bone and cartilage with central fibrovascular infiltration, but without bone marrow elements, most closely represent articular bone.

Local tissue injury is a known cause of metaplastic ossification and is likely the stimulus in our patient given his history of orbital trauma, infection, and previous orbital surgeries. Prior imaging of this patient over a decade earlier precludes the possibility of choristoma.

This case thus represents the first report of combined cartilage and bone metaplasia in the orbit and the first documentation of metaplasia involving an extraocular muscle, a tissue well known to be distinct from other skeletal muscle.[6] Although there are no known examples of malignant degeneration of metaplastic or heterotopic ossification, metaplasia may occur secondary to local malignancy. Therefore, excisional biopsy of suspected metaplastic masses arising without other obvious stimulus is probably warranted.

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Compressive Effects of Intravascular Papillary Endothelial Hyperplasia

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Abstract: **A 45-year-old woman presented with an 8-month history of a firm mass over the right superonasal orbital rim, followed by an episode of hemorrhage and anesthesia 5 months later. Clinical history and CT were suggestive of a venous malformation with phleboliths. Excision biopsy was performed. Histopathology revealed intravascular papillary endothelial hyperplasia with fibrotic-calcific changes. Hypoesthesia in the supraorbital region persisted postoperatively. Although a benign lesion, intravascular papillary endothelial hyperplasia should be considered a differential diagnosis of a vascular lesion with compressive effects.**

Intravascular papillary endothelial hyperplasia (IPEH) is a benign condition characterized by papillary endothelial projections within a vessel lumen, in association with a thrombus.[1] This lesion occurs most commonly in the head and on the extremities; only 11 cases have been reported in the ocular adnexa (7) and orbit (4).[2,3] Because of their benign nature, these lesions have not been reported to cause any structural or functional deficit aside from one report of an orbital lesion, which invaded the temporal fossa by eroding the lateral wall of orbit.[4] Our case highlights, to the best of the our knowledge, a previously unreported compressive effect of IPEH of the eyelid.

CASE REPORT

A 45-year-old woman presented with a hard mass in the right upper nasal quadrant of the orbit of 5 months' duration. Systemic history was unremarkable. On examination, vision was 6/6 OU, with no relative afferent pupillary defect. Extraocular movements were full. An 8-mm hard mass was palpable at the right superior orbital rim, adjacent to the supraorbital notch. There was hypoesthesia in the right supraorbital nerve distribution. Anterior segment and fundus evaluation were unremarkable. Hertel exophthalmometry measured 17 mm OU. CT showed an ill-defined area of hypodensity, with 2 central hyperdense masses (Fig. A). Presumed diagnosis was a vascular lesion with 2 phleboliths. She presented 10 weeks later, while awaiting excision biopsy, with hemorrhage within the lesion leading to bruising over the superior orbital rim and a further decrease in sensation. An anterior orbitotomy revealed a bluish vascular lesion containing 2 hard, white nodules (Fig. B). At 1-week follow-up, she had made an uncomplicated recovery with persistent anesthesia in the distribution of the supraorbital nerve.

Grossly, the excised specimens were irregular, nodular, and grayish masses measuring 7 mm × 4 mm × 4 mm, with cystic changes on cut section. The lesions were localized within a dilated vein and had a peripheral rim of compressed fibrous tissue and central calcification (Fig. C). Histopathology revealed endothelial lined papillary structures, with a small amount of hemosiderin pigment in both the specimens (Fig. D). There was no evidence of endothelial atypia, mitotic activity, or inflammatory reaction. CD31 immunostain, suggesting proliferation of endothelial cells, was positive in the specimen. Thus, the final diagnosis was

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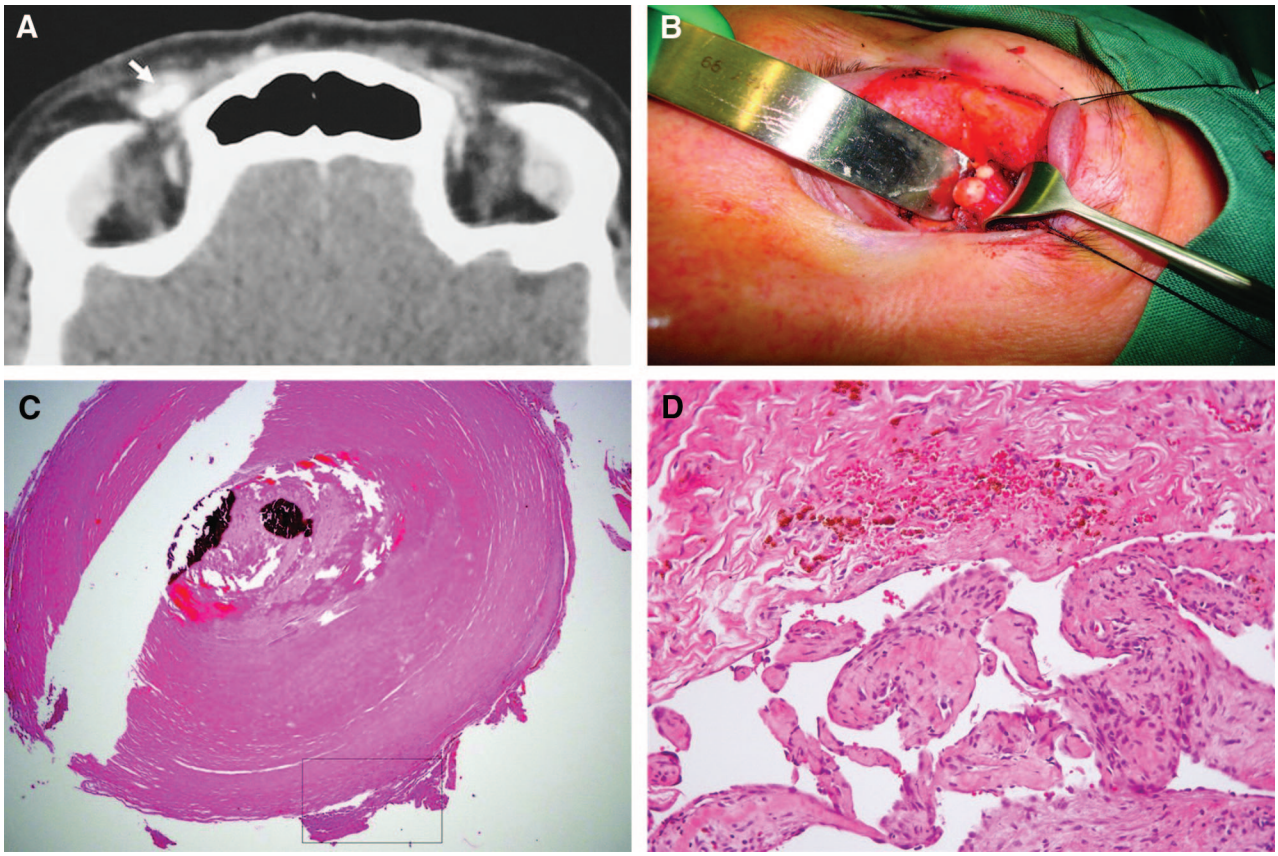

A, Axial CT of the orbit shows ill-defined areas of hypodensity at the superior orbital rim with 2 areas of calcification. **B,** Intraoperative photograph shows a vascular, cystic lesion with 2 calcified nodules. **C,** Histopathology shows a fibrotic and calcified intravenous nodule with a pseudocapsule indicating preexisting venous wall and IPEH compressed to a side (hamatoxylin-eosin, ×10). **D,** Finger-like papillary structures with normal endothelial lining and no inflammatory/ mitotic activity typical of Intravascular papillary endothelial hyperplasia (hematoxylin-eosin, ×40).

a pure type of IPEH, not associated with any preexisting vascular lesion as pyogenic granuloma, lymphangioma, varix, or hemangioma.

DISCUSSION

The first description of IPEH was by Masson[1] in 1973, who called it "vegetant intravascular hemangioendothelioma." He believed endothelial proliferation to be the primary process, followed by thrombus formation, whereas most others report thrombus to be the inciting event for this endothelial papillary formation.[5] This lesion can occur at any age and is usually found in the soft tissues of the head and the extremities. Depending on the underlying pathology, it generally presents as a firm to cystic swelling, present for some time but with a history of recent increase in size, although, spontaneous lesions have been reported. It is also possible that in our case, there was an underlying small venous malformation or calcified varix, not apparent on histopathology, which led to the formation of a thrombus with calcification, followed by papillary endothelial formation.

There have been occasional reports of Masson hemangioma or IPEH involving the eyelids or orbit. Font et al.[6] reported a series of 5 IPEH cases in 1983, which affected the orbit in 3, and eyebrow and eyelid in 1 case each. Werner et al. described a further 5 cases in eyelids in 1997.[2,3] However, none of these reports had any evidence of compression or erosion of surrounding structures. In 1981, Weber and Babel[4] reported a single case of IPEH of the orbit, which had infiltrated in the temporal fossa by destroying the lateral wall of the orbit. Our patient presented with anesthesia in the distribution of the supraorbital nerve, which persisted even after surgical removal of the mass. This is most likely a consequence of the location of the lesion, abutting the supraorbital nerve at the supraorbital notch.

Discrete areas of calcification within the centre of the lesion, as seen in our patient, have not, to the best of our knowledge, been reported previously. Shields et al.[3] described a case of IPEH in a thrombosed varix, where CT showed a few, ill-defined hyperdense areas, suggestive of calcification. Our case possibly represents an advanced stage of organizing thrombus.

Hence, this case highlights that, although a benign lesion, IPEH may lead to compressive effects on adjacent structures, such as nerves, and may demonstrate calcification on imaging.

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Management of Facial Disfigurement in Orbitotemporal Neurofibromatosis

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Abstract: **A 19-year-old woman with type I neurofibromatosis requested reconstruction of her severe facial disfigurement caused by the plexiform neurofibroma of the right upper eyelid. Previously, she had glaucoma surgery for buphthalmus and enucleation with dermis fat transplantation. She was unable to wear prosthesis in the last year. Transverse palpebral resection of the lesion, lateral canthal reconstruction, and frontal sling ptosis surgery were performed at the same session. Postoperatively, the cosmetic appearance of the patient was markedly improved. No complication or progression was observed during 2 years follow-up. We believe that an individual-based surgical plan may give acceptable results in these challenging cases.**

Plexiform neurofibroma is the most common benign peripheral nerve tumor in the eyelid and orbit and is considered to be a characteristic lesion of neurofibromatosis type I.[1] Surgical excision is generally complicated with excessive bleeding and may result in poor cosmesis. Repeated debulking may be necessary.[2] We report the surgical reconstruction of a case with severe disfigurement of the right eyelid due to plexiform neurofibroma.

CASE REPORT

A 19-year-old woman presented to the outpatient clinic with megaloblepharon on the right side causing facial asymmetry. She was known to have neurofibromatosis type I with characteristic café-au-lait spots. In the newborn period, she was diagnosed with buphthalmus OD and underwent 2 surgeries in her first year of life. At the age of 5 years, she underwent enucleation and placement of a mersilene mesh-wrapped implant. The implant extruded 1 year after implantation. It was subsequently removed, and dermis fat graft implantation was performed with prosthesis insertion. On presentation, the right upper eyelid was floppy and distended to the extent that bulky eyelid skin covered the right lower eyelid by 1 cm (Fig. 1). Lateral canthal displacement and oculopalpebral diastasis were observed. She had no levator function, no skin crease, and had an artificial eye that was leaning on the eyelid. The socket had no movement. MRI revealed a defect in the right orbital wall and lesser wing of the sphenoid bone.

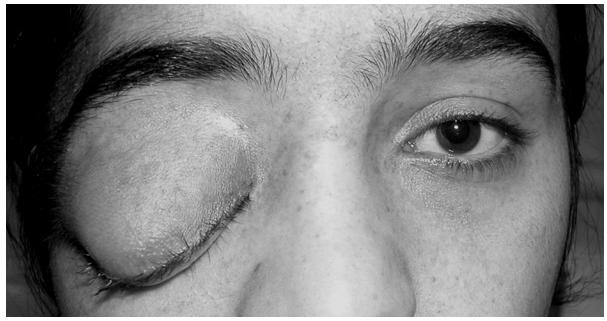

FIG. 1. Preoperative appearance of the patient showing the bulky right upper eyelid extending 1 cm below the lower eyelid margin.

Reconstructive eyelid surgery was planned (Fig. 2). With the contralateral eyelid serving as a model, the intended excision area was marked on the skin. Transverse palpebral resection of the upper eyelid and lateral canthal attachment of both upper and lower tarsal plates to the periosteum were performed. This was followed by a frontal sling operation using an expanded poli-tetra-fluoro-ethylene material (Ptose up, FCI Ophthalmics, Marshfield Hills, MA, U.S.A.).

Macroscopically, the excised tissue appeared as spindle-shaped whitish tissue. Pathology revealed expanded tortuous peripheral nerve sheaths with loose stroma that stained with S-100, and a diagnosis of plexiform neurofibroma was made (Fig. 3).

Two weeks after surgery, the patient was able to insert her old prosthesis. Her eyelid crease was 7 mm, and interpalpebral distance was 7 mm. She had 7-mm hypoglobus on the right side. Her cosmetic appearance was markedly improved (Fig. 4) and remained stable during the 2-year follow-up period.

DISCUSSION

Plexiform neurofibromas diffusely infiltrate the tissues resulting in very vascular with wide open capillaries that bleed copiously during surgery. Surgical principles in the management of orbito-craniofacial neurofibromatosis have been described in many reports.[2–4] Since each patient has different clinical manifestations, the surgical approach and the extent of resection depend on the severity of the orbital soft-tissue and bony involvement, and the visual potential.

In orbitotemporal neurofibromatosis, Lee et al. recommended partial excision in debulking of neurofibromas. As the lateral part of the upper eyelid is typically more infiltrated, a full-thickness pentagonal wedge excision can be performed in this area.[2] Other authors recommend supratarsal, transverse or transverse-oblique, full-thickness wedge resection of the affected upper eyelid, with immediate reconstruction of the lateral canthus and levator apparatus resulting in functional and aesthetic benefit despite the progressive and debilitating nature of the disease.[4] In the present case, we preferred transverse palpebral resection. Because of the massive bulk in the eyelid, we believed that partial resection would not suffice and might result in conjunctival prolapse. Our patient also had a defect in the posterolateral orbital bone and had previous enucleation and dermis fat transplantation. Because the socket was not protruding 14 years after surgery, and the complaint of the patient was the disfigurement of the eyelid, we did not consider reconstruction of the orbital bone.

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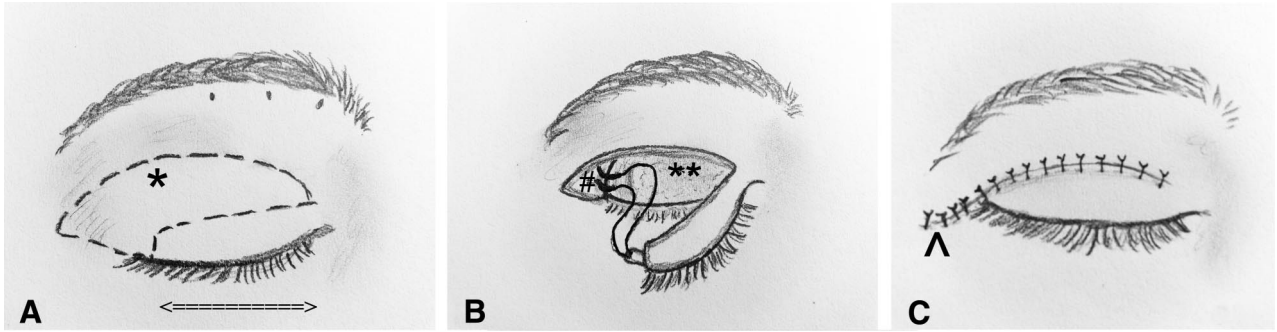

FIG. 2. The drawing of the transverse radical excision and canthopexy. **A,** The intended excision area was marked (*). Her fellow eye served as a model. The width of the normal upper eyelid was measured (<==>). The lower margin was marked according to the distance of skin crease from the eyelid margin in normal eye. Three dots marked the medial, mid, and lateral points of the normal eyebrow. The distance from the skin crease to these 3 dots was noted from the fellow eye to mark the upper margin of the intended excision. **B,** Following the transverse radical excision of the upper eyelid bulk, the lateral orbital rim (#) was exposed, and anopthalmic socket (**) was observable. **C,** The lateral tarsus was fixated to the periosteum, and lateral canthopexy (▲) was performed. Next, facial sling procedure was performed, and upper eyelid skin was closed.

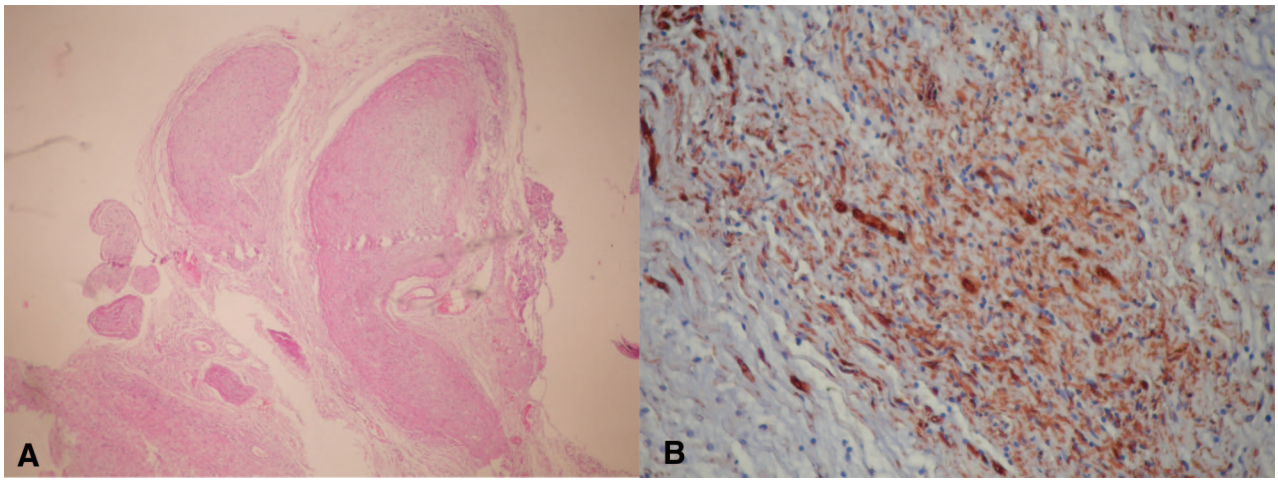

FIG. 3. Pathologic examination revealed **(A)** expanded tortuous peripheral nerve sheaths with loose stroma (hematoxylin-eosin, ×100), and **(B)** the peripheral nerve sheaths stained with S-100 (S-100, ×400).

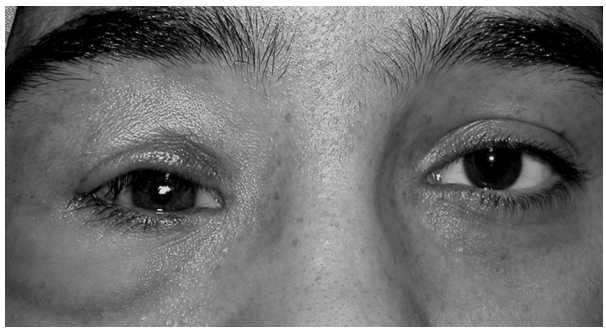

FIG. 4. At 2 months after surgery, marked improvement in clinical appearance was observed.

Ptosis surgery in patients with neurofibromatosis type 1 is challenging. Because of the heavy infiltration, it is often difficult to identify the levator aponeurosis and judge the correct amount of levator resection. Despite this, it was reported that all of the patients, even those with complete ptosis preoperatively, achieved a satisfactory elevation of the upper eyelid above the pupillary axis.[2] Given the extensive palpebral involvement resulting in floppy eyelid with no levator function, and patient already wearing an ocular prosthesis, our choice in ptosis surgery was frontalis sling. With this method, we achieved a satisfactory elevation of the upper eyelid, and no related problems were observed during the 2 years of follow-up. To our knowledge, this is the only report where alloplastic material was used in frontalis sling to correct the ptosis in a case with neurofibromatosis.

In conclusion, our case with plexiform neurofibroma was corrected with transverse palpebral resection, lateral canthal fixation, and a frontalis sling. Postoperatively, the patient's appearance and social interaction improved markedly. We believe that surgery tailored according to the clinical characteristics of the individual patient can achieve acceptable outcomes even in challenging orbitotemporal neurofibromatosis cases with severe facial disfigurement.

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IgA Orbital Plasmacytoma in Multiple Myeloma

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Abstract: **The authors report a case of orbital plasmacytoma in a 48-year-old man with known multiple myeloma. He presented with proptosis, diplopia, and decreased vision of the left eye for several weeks. He had been previously treated for IgA λ multiple myeloma with chemotherapy, radiation, and autologous stem cell transplant. After a left orbitotomy, flow cytometry revealed a tumor rich in plasma cells expressing CD138 with equivocal λ light chain expression. The patient underwent orbital radiation, with improvement of vision and disc edema OS. The patient is currently undergoing salvage chemotherapy for relapse of multiple myeloma. This is the third reported case of IgA myeloma involving the orbit.**

Plasmacytomas are tumors of plasma cell origin that can occur either primarily or secondarily.[1] Primary plasmacytoma could involve either the bone (solitary plasmacytoma of bone) or soft tissues (solitary extramedullary plasmacytoma). Primary plasmacytomas account for 10% of all plasma cell disorders. Multiple myeloma, which accounts for the other 90% of plasma cell disorders, can lead to secondary plasmacytomas.[2] Both solitary extramedullary plasmacytomas and secondary plasmacytomas involving the orbit are rare. Approximately 60 cases of secondary orbital plasmacytomas are found in literature.

CASE REPORT

A 48-year-old man had been diagnosed 4 years previously with IgA λ multiple myeloma. The diagnosis had been made after the patient experienced a fall and was found to have a pathologic fracture of the right femur and signs of abnormal kidney function. After he had undergone chemotherapy and radiation of the pathologic fracture site, disease progression was confirmed by bone marrow biopsy, and an autologous stem cell transplant was performed. A bone survey 1 year later confirmed further bony involvement in the left femur. Four years after the original diagnosis, he presented to the orbit clinic with binocular diplopia, pain with extraocular movements, and decreased vision of the left eye of several weeks' duration. Best-corrected visual acuity was 20/20 OD and 20/30 OS. There was significant dyschromatopsia and an afferent pupillary defect OS. The patient also had 6-mm proptosis OS (Fig. 1). Further examination revealed 14 prism diopters of exotropia and 12 prism diopters of right hypertropia in primary gaze, with limited abduction and supraduction. Intraocular pressures were 21 mm Hg OU. Fundus examination revealed pink and sharp optic nerves without any disc edema or vascular abnormalities.

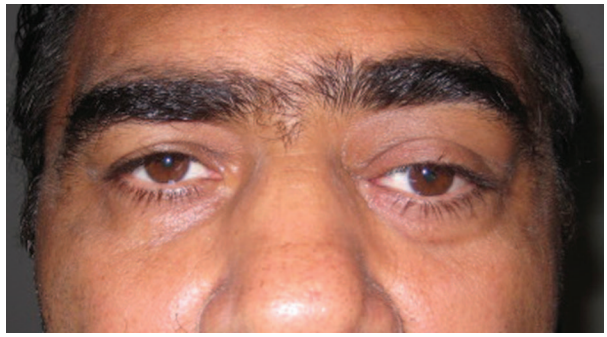

FIG. 1. The left eye is proptotic and has been displaced inferotemporally.

Orbital MRI revealed a homogeneously T^1 hypointense and T^2 hyperintense mass (Fig. 2) measuring 3.0 × 3.0 cm transverse × 2.1 cm craniocaudad. The mass was centered at the superolateral left orbit and extended through the lateral wall of the left orbit in the left infratemporal fossa. The patient underwent a left orbitotomy with biopsy. Flow cytometry revealed plasma cells expressing bcl-2 and CD138 with equivocal λ light chain expression not expressing CD19, CD20, or CD45. Before starting radiation treatment, the patient suffered further loss of vision to hand motions only OS with optic nerve edema. Orbital CT (Fig. 3) documented a significant increase in the mass size to 4.4 × 5.5 cm transverse × 4.2 cm cranio-

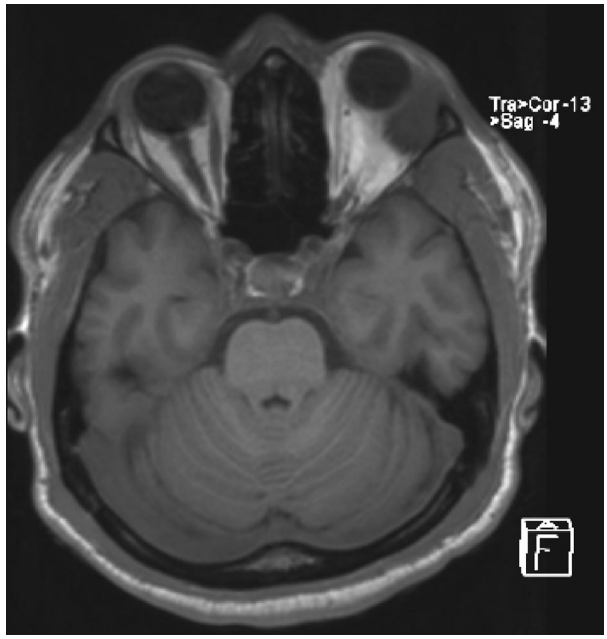

FIG. 2. Orbital MRI shows T_1-weighted image with a large left orbital mass that is hypointense on the left.

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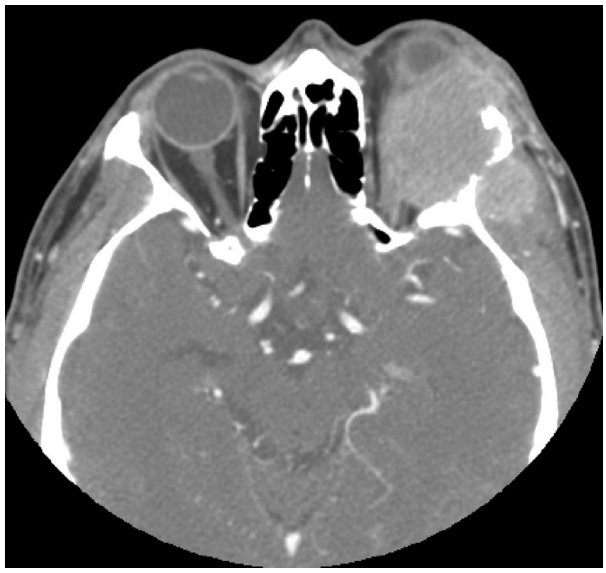

FIG. 3. Axial image of orbital CT with contrast reveals a large left orbital mass, extending in the temporal fossa and causing bone remodeling.

caudad with extension in the left orbit, temporal fossa, frontal sinus, and left frontal epidural region. The patient underwent orbital radiation of 40 Gy over a period of 5 weeks. Vision was dramatically improved to 20/30 OS with improvement in disc edema and only residual inferior arcuate visual field defect. The mass decreased to 1.8 × 0.4 cm transverse × 1.5 cm craniocaudad. The patient is currently undergoing salvage chemotherapy treatment.

DISCUSSION

Multiple myeloma is only rarely seen in the orbit. The differential diagnosis of an orbital mass in myeloma patients should include lymphoproliferative lesions such as lymphoma and plasmacytoma. These patients most commonly present with exophthalmos, as seen in a review of 30 cases of orbital involvement in multiple myeloma by Rodman and Font in 1972. Other signs include chemosis, conjunctival hemorrhage, diplopia, and elevated intraocular pressure.[3] Myeloma may also present as orbital cellulitis as a result of immunosuppressed state or from sinus invasion.[4] Other atypical presentations include paraproteinemia causing diffuse orbital muscle swelling or necrobiotic xanthogranuloma.[5]

IgG is more commonly associated with multiple myeloma and has more frequent orbital involvement than IgA. Specific chromosomal abnormalities can be associated with multiple myeloma, some of which indicate a worse prognosis. For example, deletion of 13q, deletion of 17p, gain of 1q, and translocation t(4;14) are associated with higher risk of death.[6] In our patient, chromosome analysis was normal, and he has survived 4 years of multiple myeloma.

MRI in our patient revealed a hypointense signal on T_1-weighted images and a hyperintense signal on T_2. These findings are characteristic of orbital plasmacytomas, which are usually avascular.[7] CT may show an enhancing soft tissue mass with bony expansion and destruction.[8]

Multiple myeloma can result in severe morbidity and mortality. Soft-tissue involvement often marks the terminal phase of this disease. With orbital involvement, median survival is approximately 20 to 30 months, and death often results from infection or renal failure.[4,9] The orbital surgeon should be cautioned that acute renal failure may develop with the use of anesthetics during surgery or contrast material for imaging. Most orbital lesions are treated with systemic chemotherapy and radiation with good response. On initiation of radiation, our patient, who is the third reported case of IgA multiple myeloma involving the orbit, responded well with dramatic shrinkage of tumor and improvement of vision.

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Recurrent *Rhodococcus gordoniae* Eyelid Infection After Combined Septoplasty and Blepharoplasty

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***Abstract:* *Rhodococcus* species are rare human pathogens. Most cases of human infection have involved *Rhodococcus equi*, a natural zoonotic pathogen causing pneumonia in young foals. The human population at risk mainly includes immunocompromised patients. Infection of immunocompetent hosts has been rare, with less than 30 reported cases in the literature. The authors report a first case of recurrent *Rhodococcus gordoniae* granulomatous lower eyelid infection that occurred after an immunocompetent 44-year-old woman underwent a combined intranasal and lower blepharoplasty procedure. A literature search revealed only 10**

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reported cases of non-*R. equi* rhodococcal infections, most involving traumatic inoculation; 5 of these cases involved the eye and ocular adnexa. To the author's knowledge, this is the first case report of a non-*R. equi* rhodococcal infection of the eyelids. Identification of *Rhodococcus* by most laboratories is difficult, and in this case, 2 of 3 cultures were misidentified prior to definitive testing at a specialized laboratory. Multidrug resistance is also a concern, and the treatment course could be long and difficult.

A 44-year-old healthy woman was referred to the University of Tennessee's Hamilton Eye Institute Oculoplastic Service 3 months after undergoing a combined septoplasty with turbinate reduction and bilateral upper and lower eyelid blepharoplasty. She developed multiple nodules in both lower eyelids several weeks postoperatively, which responded modestly to multiple triamcinolone injections. On presentation to us, she had a 1 × 1-cm firm, nonerythematous, painless nodule in the right lower eyelid surrounded by mild soft-tissue swelling (Fig., top). Excision revealed purulent material with a surrounding fibrous capsule. Histology revealed fibroadipose tissue with chronic granulomatous inflammation. Cultures grew "*Rhodococcus* species not *Equi*." Infectious disease recommended erythromycin and rifampin therapy. The patient had a slow response to therapy and subsequently developed multiple recurrent nodules of both lower eyelids over the next several months (Fig., bottom), which grew *Nocardia* species and *Corynebacterium* species. Because of the varied culture results, samples were sent for definitive 16s rRNA analysis; the results confirmed *Rhodococcus gordoniae*. The patient successfully responded to a combination of vancomycin and erythromycin induction and was maintained on trimethoprim/sulfamethoxazole, minocycline, and azithromycin for 6 months. She had no recurrence at her last visit 7 months after antibiotics began.

DISCUSSION

Rhodococcus spp. are facultative intracellular Gram-positive coccobacillus of the suprageneric taxon nocardioform actinomycetes. They exhibit pleomorphism growing as cocci, short rods, or branching filaments. They are ubiquitous in the environment and primarily cause zoonotic infections but uncommonly can infect humans. *Rhodococcus* species are widely distributed in soil and have been found in bovine, porcine, and equine fecal flora. Most cases of human infection involve *Rhodococcus equi*, a natural pathogen causing pneumonia in young foals. *Rhodococcus* infection has a variable clinical presentation; most common is a granulomatous inflammatory reaction with abscess formation.[1] Immunocompromised patients are at risk, especially those with AIDS. Only 30 cases have been reported in immunocompetent hosts. Reports of nonequi rhodococcal infections are rare but most are secondary to traumatic inoculation. Cutaneous infections, endophthalmitis, and keratitis after trauma or laser in situ keratomileusis have been reported.[2–6]

Based on in vitro studies, *Rhodococcus* species are sensitive to tetracyclines, aminoglycosides, macrolides, trimethoprim/sulfamethoxazole, rifampin, and vancomycin.[7] Multidrug resistance has been reported in the literature. This makes the prospect of these organisms developing increasing virulence (through plasmid exchange, etc.) daunting, given their ubiquitous nature and common exposure to standard antibiotic regimens through veterinary applications and the likelihood of the development of multidrug resistance.

R. equi infected patients frequently have a history of exposure to farm animals, soil, or both[8]; however, there are not sufficient data in the literature to support this correlation in human infections with non-equi rhodococci. Although our patient had no direct exposure to livestock, her husband owns farmland, and he may have been a source of exposure.

An evaluation of human nasal flora described by Rasmussen et al.[9] determined that *Rhodococcus* species comprised a significant portion of the native nasal flora in 4 of 10 human subjects evaluated. Although nasal cultures were not performed, it is possible our patient was colonized with *R. gordoniae* with subsequent inoculation of her eyelids during the combined septoplasty/blepharoplasty procedure.

Another consideration in our patient is whether the administration of multiple triamcinolone injections created local immunosuppression, allowing a less-restrictive environment for the growth of the involved organism. In fact, the first reported case of *Rhodococcus* infection in humans was in a patient immunosuppressed with systemic corticosteroids for the treatment of chronic hepatitis. The possible role of steroids is further supported by a 1978 study by Haburchak et al. that demonstrated the development of granulomas in steroid-treated guinea pigs inoculated with *Rhodococcus* spp.[10]

It is important to note the confusion in laboratory assessment of these organisms with related bacteria. In our case, the organism was identified as *Rhodococcus* spp., *Corynebacteria*, and *Nocardia*, respectively, on 3 separate cultures. The clinician should be aware of the potential laboratory pitfalls in the identification of *Rhodococcus* spp., as its diphtherioid morphology may prompt discarding the organism as a contaminant or misidentifying it as a related organism such as *Mycobacteria, Corynebacteria*, or *Nocardia*. A high index of suspicion is required to request definitive testing for the aerobic actinomycetes and the genus *Rhodococcus* in particular.

We present the first case report of *R. gordoniae* infection of the eyelids. The clinician should be aware of its clinical findings and potential pitfalls in the identification of *Rhodococcus species*.

Top, patient at the time of presentation to the oculoplastic service, 3 months after combined septoplasty with turbinate reduction and bilateral upper and lower eyelid blepharoplasty. Bottom, 1 month after incision and drainage of the nodule.

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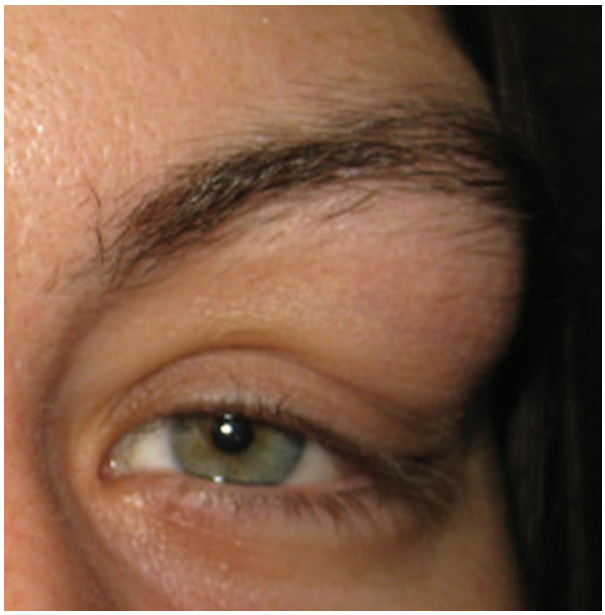

FIG. 1. The left orbit contains a large mass.

Myxofibrosarcoma of the Orbit: A Clinicopathologic Case Report

Qing Zhang, M.D., Ted H. Wojno, M.D.*, Barry M. Yaffe, M.D.†, and Hans E. Grossniklaus, M.D., M.B.A.*‡*

Abstract: **A 27-year-old woman developed a rapidly progressive left orbital tumor that extended in the cranial fossa. MRI revealed a heterogenous enhancing lesion confined to the left frontal bone and superior orbit. An incisional biopsy was performed, and histopathologic examination of the specimen showed findings diagnostic of a high-grade myxofibrosarcoma. Complete excision with postoperative adjuvant radiation therapy and chemotherapy was performed, and the patient had no evidence of tumor recurrence within 6 months' follow-up. Myxofibrosarcoma is a fibroblast-derived soft tissue neoplasm with up to a 60% local recurrence rate, and metastasis may be associated with intermediate to high-grade tumors.**

Myxofibrosarcoma (MFS), formerly known as a myxoid or a myxoid variant of malignant fibrous histiocytoma, was first proposed in 1977[1] and is recognized by the World Health Organization classification as a category of fibrous tumor.[2] Although MFS is a malignant mesenchymal tumor of soft tissue that commonly involves limb and limb girdle, there has been only 1 case described of MFS occurring in the orbit.[3] We report an additional case of MFS of the orbit and briefly review the clinical, radiologic, and histopathologic characteristics of this tumor.

CASE REPORT

A 27-year-old woman presented with a 6-week history of a painless lump in her left upper eyelid and left eyebrow area. She had mild diplopia and no history of trauma or surgery. Her best corrected vision was 20/25 OD and 20/20 OS. Slit lamp examination was unremarkable. A 3 × 3-cm firm mass was palpable in the superotemporal orbital region (Fig. 1). MRI demonstrated a mass involving the frontal bone at the lateral superior aspect of the orbit, extending superiorly in the frontal bone, and impinging on but not clearly invading the dura (Fig. 2).

A biopsy of the lesion was performed, and histopathologic examination showed a tumor composed of loosely coherent sheets of cells, including elongated spindle-shaped cells with fusiform nuclei and prominent nucleoli and round cells with round nuclei. Some of these cells contained abundant eosinophilic cytoplasm (Fig. 3A). Curvilinear blood vessels

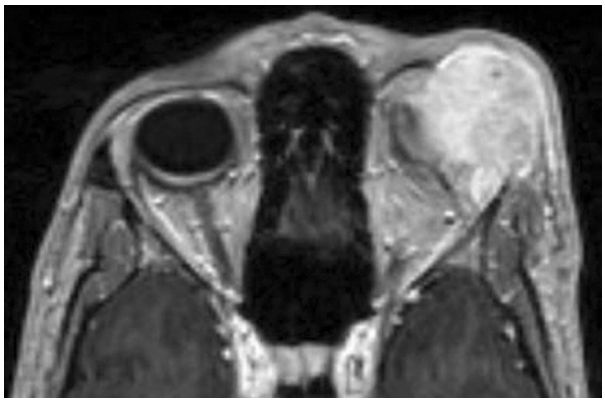

FIG. 2. MRI demonstrates a mass involving the frontal bone at the lateral superior aspect of the orbit, extending superiorly in the frontal bone.

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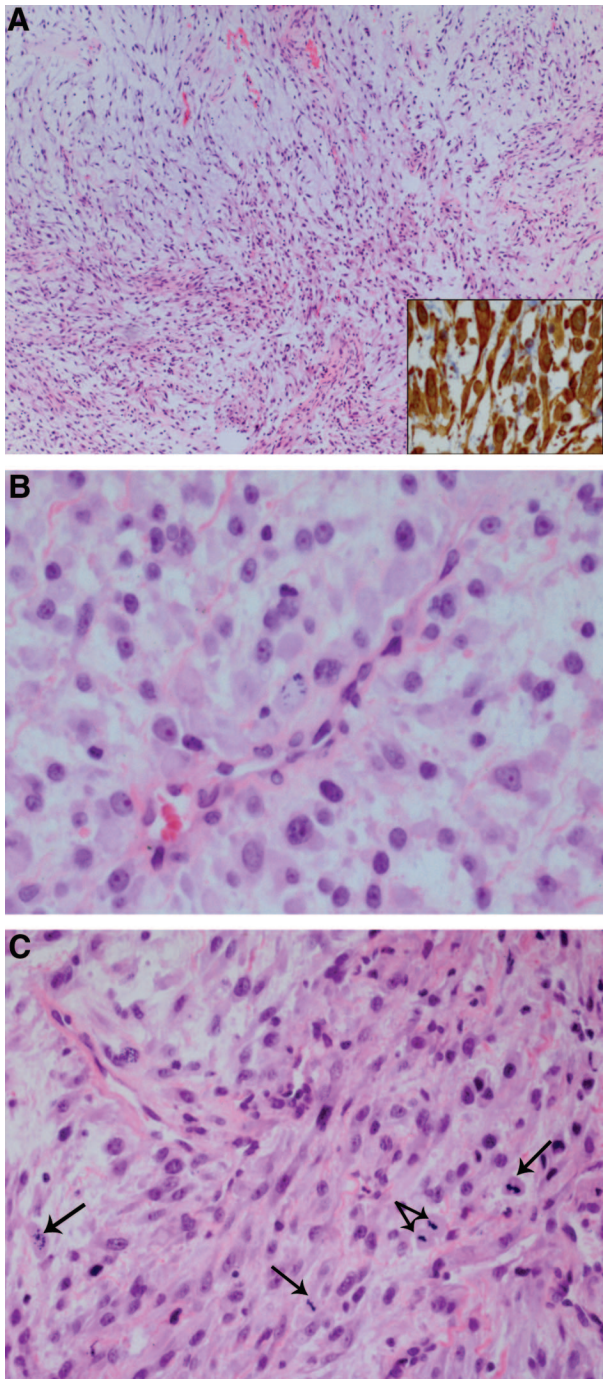

FIG. 3. **A,** The tumor contains loosely coherent sheets of spindle-shaped or stellate cells within a myxoid stroma. Those cells have fusiform nuclei and prominent nucleoli (hematoxylin-eosin, ×25). Inset, the tumor stained positive with vimentin (×100). **B,** Curvilinear blood vessel can be seen within the lesion (hematoxylin-eosin, ×100). **C,** Several mitotic figures are present in the tumor (arrows), including occasional bizarre mitotic figures (hematoxylin-eosin, ×100).

were present in some areas (Fig. 3B). There were greater than 20 mitotic figures per high power field in the tumor, and occasional bizarre mitotic figures were present (Fig. 3C). Immunohistochemical stains were strongly positive for vimentin (Fig. 3A, inset), for Ki67 in 20% of nuclei, and negative for desmin, myogenin, smooth muscle actin, and S100 in the tumor. The findings were consistent with high-grade MFS.

The patient underwent a left fronto-orbital craniotomy for tumor resection, resection of a small portion of underlying dura, and immediate plastic reconstruction. Intraoperatively, the tumor had a relatively well-defined capsule and was found to extend in the lateral orbital region. The bone in the frontal orbital area including the rim was substantially destroyed by the tumor. The histologic findings were as described above. Electron microscopic examination of the tumor showed scattered neoplastic cells, ranging from spindle shaped with round to oval nuclei, prominent nucleoli, and heterochromatin to stellate with irregular nuclei. These cells contained prominent intracytoplasmic rough endoplasmic reticulum, intracytoplasmic membrane, bounded vacuoles, and Golgi apparati.

The patient completed 38 fractions of adjuvant radiation treatment to a total of 66.6 Gy using the intensity-modulated radiotherapy technique, which was followed by 4 cycles of adjuvant chemotherapy, each consisting of doxorubicin 75 mg/m^2 continuous infusion over 3 days and ifosfamide 2 g/m^2 four times a day for 4 days, with mesna uroprotection. Treatment was well tolerated, and the patient had no physical or radiologic evidence of tumor recurrence with 6 months of follow-up.

DISCUSSION

MFS is considered a distinct and definable entity that predominantly occurs in the lower and upper limbs. Underrecognition and hence underdiagnosis may be a reason for the rare occurrence of this neoplasm in the orbit. Clinically, MFS often presents as a gradually enlarging painless mass. Although MFS is one of the most common soft-tissue sarcomas in elderly patients in the sixth to eighth decades, cases have been reported in patients younger than 20 years of age.[4] The present case occurred in a 27-year-old woman.

Histopathologic findings in MFS include a multinodular proliferation of spindle-shaped or stellate fibroblasts within a myxoid stroma containing curvilinear blood vessels. The tumor cells are hyperchromatic and display moderate to marked pleomorphism.[5,6] Immunohistochemical staining of MFS shows diffuse strong positivity for vimentin, consistent with fibroblastic differentiation, and negative staining for cytokeratins, desmin, human melanoma black 45, and S100.

Ultrastructural findings include prominent rough endoplasmic reticulum, well-developed Golgi apparati, and a moderate number of mitochondria. The differential diagnosis of MFS includes pleomorphic rhabdomyosarcoma, myxoid liposarcoma, melanoma, myoepithelial carcinoma, and metastatic carcinoma. These entities were excluded in our case because of histologic, immunohistochemical, and ultrastructural features.

There is some controversy regarding the grading of this tumor, because there are both 3- and 4-tiered grading systems. One scheme classifies MFS as grades I–IV, whereas another system classifies MFS as low, intermediate, and high grade (Table).[1,5,6] In comparison with the previous report of MFS of the orbit described by Wang et al.,[3] our case is relatively higher grade according to the degree of tumor cellularity, cellular atypia, and mitotic figures.

Complete excision with tumor-free margins is the recommended treatment for MFS. The tumor response to therapy may be monitored with MRI. When negative margins cannot be assessed, a combination of postoperative radiation therapy and systemic chemotherapy are recommended, especially for high-grade MFS.[5,6]

The local recurrence rate of MFS is about 60%, and recurrent lesions often progress to higher grades with increased

Myxofibrosarcoma grading system

	Low grade	Intermediate grade	High grade
Cellularity	Hypocellular architecture with abundant myxoid matrix	More cellular with discernable myxoid matrix	Hypercelluar architecture with small myxoid areas
Celluar atypia	Mild nucelar pleomorphism and hyperchromasia	Moderate nuclear atypia	Prominent cellular pleomorphism and nuclear atypia
Mitotic activity	Occasional mitoses	Higher mitotic rate	Conspicuous mitotic figures with bizarre nuclear features

metastatic potential.[1,6] Lung and bone are the most common sites of metastasis, although spread to regional lymph nodes may occur.[5,6] Because of a high recurrence rate and risk of metastasis, patients with orbital MFS should be treated aggressively and followed closely.

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Conservative Treatment of Adenoid Cystic Carcinoma With Plaque Radiotherapy: A Case Report

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Abstract: **Adenoid cystic carcinoma is a rare but aggressive malignant tumor of the lacrimal gland. Mortality rates have been found to be quite high (>50%). This has led to the traditional method of treatment with radical surgery including orbital exenteration and bone resection, followed by external beam radiotherapy to the orbital bed. Despite this aggressive treatment strategy, mortality rates still remain high with only a few reported cases with long-term survival. Recently, several cases of successful treatment of adenoid cystic carcinoma with plaque radiotherapy have been documented. These may show promise of a conservative treatment strategy in selected cases. The authors present a functionally monocular patient with a well-circumscribed adenoid cystic carcinoma that was treated with local excision of the tumor with reverse plaque brachytherapy "boost" to the lacrimal tumor bed followed by a lower dose of radiotherapy to the orbit. The patient is alive and without local recurrence at nearly 4 years.**

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Adenoid cystic carcinoma is a rare but aggressive malignant tumor of the lacrimal gland epithelium. It is the most common primary malignant tumor of the lacrimal gland but accounts for only 4% to 5% of lacrimal gland lesions.[3,4] Adenoid cystic carcinoma is known for its slow growth and frequent recurrences, possibly due to its early and diffuse infiltration and tendency for perineural invasion.[5] Aggressive treatment is typically performed, including orbital exenteration, resection of neighboring bone, and usually adjuvant radiation therapy to eradicate potential microscopic residual cancer cells.[6] Despite these measures, mortality rates remain greater than 50% at 5 years and survival rates at 10 years are dismal at around 20%.[7,8] Death is commonly due to intracranial spread and pulmonary metastasis.

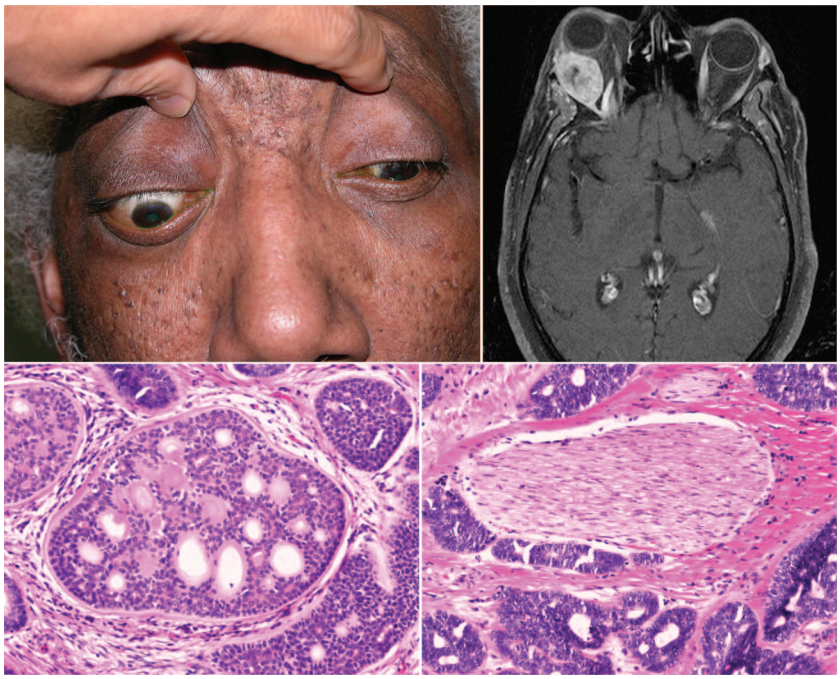
Although long-term follow-up is not yet available, plaque radiotherapy has been proposed in certain cases of adenoid cystic carcinoma to preserve sight and function. Several cases of successful treatment have been reported to date.[1,2] This treatment has allowed these patients to avoid disfiguring surgery and to preserve the eye. Plaque radiotherapy "boost" allows delivery of a high radiation dose directly to the tumor or tumor bed while sparing the radiosensitive globe. In our case, this was followed by a lower external radiation dose to the entire orbit to eradicate potential residual microscopic cancer cells. This mode of treatment is an attractive alternative in the young or functionally monocular patient.

CASE REPORT

Our patient is a 59-year-old African American male who presented in August 2005 with a 3-year history of progressive proptosis of the right eye with intermittent sharp pain in the surrounding region. MRI of the orbits was obtained and revealed a circumscribed 3.5 × 3 × 2 cm enhancing soft tissue mass within the superolateral right orbit displacing the optic nerve, lateral and superior rectus muscles. No bony erosion or defacement was noted.

The patient underwent right lateral orbitotomy with local tumor excision. Pathologic examination of the resected mass revealed a diagnosis of adenoid cystic carcinoma of the lacrimal gland of a combined cribriform and tubular subtype with a very minor solid component. Margins involving the lateral orbital wall were positive for malignancy, and potential residual microscopic tumor remained at the lateral rectus muscle. Workup revealed no evidence of metastatic disease. This is representative the tumor's locally advanced nature and incomplete tumor excision. The tumor was classified as stage T4N0M0 (Fig.).

The patient has a history of open-angle glaucoma and previous ischemic optic neuropathy of the left eye leaving him



Top left, Our patient with right unilateral proptosis and downward displacement. **Top right**, MRI showing enhancing right orbital mass in the area of the lacrimal gland. **Bottom left**, Adenoid cystic carcinoma, cribriform subtype shown in this area. **Bottom right**, Perineural invasion.

with only count fingers vision in the left eye and, therefore, functionally monocular. The patient was offered orbital exenteration of the right orbit for residual tumor followed by adjuvant radiation therapy, but he refused. The patient stated that he would rather let the cancer grow back gradually and be able to see for a period of time and die from cancer than undergo surgery that would leave him without meaningful vision. At the time, visual acuity of the right eye was 20/100.

After discussion with the patient and with the VA tumor board, it was decided that to attempt to minimize ocular and visual side effects of radiation therapy a combination of localized and external beam radiotherapy would be performed. The patient underwent local plaque radiotherapy with I-125 seeds followed by external beam radiation therapy to the orbit and globe. A single "reverse" scleral plaque was sutured to the superotemporal sclera. The plaque radiotherapy dose was 1542 cGy at 75.2 cGy/hour for 20.5 hours to the lateral rectus muscle and 1356 cGy to the superolateral orbit for 29.5 hours after being readjusted. The plaque was removed 29.5 hours after insertion. The external radiation therapy to the orbit began 4 weeks after the plaque was removed, and the dose delivered was 5040 cGy at 180 cGy daily fraction in 28 fractions over 37 elapsed days. Both of these treatments were well tolerated. The patient has had yearly MRI scans of the brain and orbits and has had no sign of orbital or intracranial recurrence or metastasis.

Subsequently, the patient has undergone fascia lata suspension of the right upper eyelid for ptosis, has persistent problems with nonhealing corneal epithelial defects related to exposure and xerophthalmia, and has developed dense cataract formation causing a significant decline in visual acuity. The patient is currently scheduled to undergo cataract extraction, which could potentially improve his visual acuity.

DISCUSSION

Plaque radiotherapy is used frequently in the treatment of certain intraocular malignancies but evidence of its use in treating orbital malignancies, specifically adenoid cystic carcinoma is sparse. This is in part due to its recurrent and lethal nature. Orbital exenteration with or without external beam radiotherapy, and radical cranio-orbital resection have been the standard of care despite a lack of improvement in survival rates.

Intracarotid administration of chemotherapeutic agents, in addition to orbital exenteration and subsequent external beam radiation to the orbit (55–60 Gy), has recently been advocated in advanced cases and may show promise of improved survival.[9,10]

Improvements in the quality and accessibility of imaging studies have resulted in the discovery of many adenoid cystic carcinomas at an earlier stage. These may be more amenable to local resection. This, coupled with the tumor's lack of improvement in prognosis with radical disfiguring surgery, has led to an interest in more conservative localized treatments. Also it has been suggested that children with adenoid cystic carcinoma have a better prognosis.[11] Therefore in certain cases, such as a child with good vision or a monocular patient, the argument for radical surgery is difficult to justify.

There have been only a few published cases of the use of plaque radiotherapy following tumor excision in the treatment of orbital adenoid cystic carcinomas.[1,12] Of these, most cases having microscopic residual tumor following resection showed no recurrence after plaque radiotherapy with limited follow-up. Shields et al.[2] published a series of 4 patients with adenoid cystic carcinoma treated with local excision and Iodine-125 radiotherapy for microscopic residual tumor. One of the 4 ultimately needed orbital exenteration for recurrence of the tumor outside of the field of brachytherapy. The other 3 were tumor free at 1, 3, and 6 years. None of the patients in this series developed any ocular side effects associated with external beam radiotherapy.

Our case illustrates the therapeutic dilemma facing the physician and patient when given the option of radical surgery with poor chance of a cure, versus the unproven conservative

treatment of a frequently lethal malignancy. In consultation with a radiation oncologist, we decided to limit the side effects of radiation to the radiosensitive globe using localized plaque brachytherapy and lower dosages of external beam radiotherapy. Our design of brachytherapy plaque was similar to that used by Shields et al.[2] Our patient appears to be without recurrence at nearly 4 years but has experienced dense cataract formation, restriction of ocular motility, and xerophthalmia attributable to the external beam radiotherapy.

Radical surgery, external beam radiotherapy, and chemotherapy are likely to remain necessary in the treatment of advanced and metastatic or recurrent adenoid cystic carcinoma. Evidence is increasing supporting the use of plaque radiotherapy in the treatment of orbital adenoid cystic carcinoma. Longer follow-up and more evidence of successful treatment of adenoid cystic carcinoma with plaque radiotherapy in selected cases must be gathered to better evaluate this form of conservative treatment.

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Extensive Juvenile Xanthogranuloma Involving the Orbit, Sinuses, Brain, and Subtemporal Fossa in a Newborn

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Abstract: **A 6-week-old-boy presented with a 3-week history of right axial proptosis. Vision, motility, anterior segment, and fundus examinations were normal in both eyes. Imaging revealed a multicystic right orbital lesion with extensive involvement of the infratemporal fossa and paranasal sinuses with intracranial extension. Systemic workup was negative, and he showed no functional deficits. Histopathology revealed a tumor rich in histiocytes, and immunohistochemistry indicated a juvenile xanthogranuloma. He did well with observation, and the tumor partially involuted after 18 months of follow-up.**

Histiocytic orbital tumors are unusual in newborns.[1–3] The most common are Langerhans cell histiocytoses (LCHs), which include eosinophilic granuloma, Hand-Schuller-Christian disease, and Letterer-Siwe disease. Immunohistochemistry and electron microscopy suggest that the less common non-LCHs are derived from dermal dendrocytes. Non-LCHs are classified into 3 major groups: predominately affecting skin (juvenile xanthogranuloma, JXG), affecting skin with additional major systemic involvement (Erdheim Chester, Rosai-Dorfman), and mostly extracutaneous.[4,5] We report a puzzling case of an extensive orbital, subtemporal fossa, sinus, and intracranial tumor that after extensive workup and pathologic evaluation was diagnosed as benign JXG.

CASE REPORT

A 6-week-old-boy presented with a 3-week history of right-sided proptosis without a history of pain, redness, or strabismus. He was eating well and thriving. Visual acuity, alignment, ductions, pupils, and anterior and posterior segment examinations were all within normal limits OU. Four millimeters of right-sided proptosis was noted on Hertel measurement, with increased resistance to retropulsion. MRI showed a large right orbital mass extending in the ethmoid and maxillary sinuses, with involvement of the subtemporal fossa and middle cranial fossa (Fig. 1). Complete systemic workup and neurologic examination was negative. A complete blood count was normal. Bone marrow biopsy revealed 9% to 10% blasts, thought to represent a normal cluster of stem cells. Polymerase chain reaction assays did not indicate the presence of T- or B-cell clonal proliferation, and marrow chromosomes were normal. A fluid-filled cyst was biopsied via a right inferior orbitotomy. Histopathology revealed an infiltrate of histiocytes, lymphocytes, and eosinophils. A definitive pathologic diagnosis was difficult to obtain, and a repeat biopsy was performed on the maxillary sinus component of the mass. Two outside pathologic consultations were obtained. The first consultant's diagnosis was granulocytic sarcoma. After negative oncology workup, a second outside pathology consultation indicated JXG. Since the oncology workup was negative and the patient did not show any signs of progression, the diagnosis of JXG was made.

Immunohistochemical analysis was positive for CD68 and XIIIa and negative for CD1a, S100 protein, CD99, myelo-

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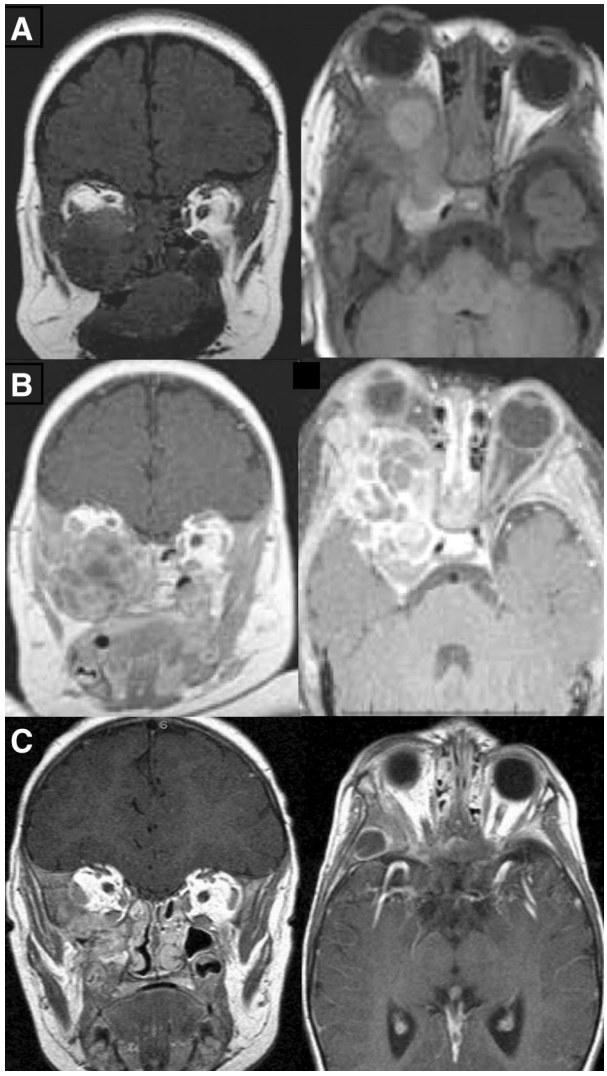

FIG. 1. T1-weighted MRI, coronal and axial cuts at **(A)** age 2 months without gadolinium, **(B)** at 5 months with gadolinium, and **(C)** at 18 months with gadolinium, shows regression of a heterogeneously enhancing right orbital mass with involvement of the maxillary and ethmoid sinuses, pterygopalatine fossa, infratemporal fossa, middle cranial fossa, and right cavernous sinus.

peroxidase, muramidase, CD34, TdT, desmin, myogenin, neuroblastoma antigen, and keratin (Fig. 2). Clonality studies were negative for B- or T-cell gene rearrangement by polymerase chain reaction. Because the child had no ocular or systemic deficits, treatment was not advised. Neuroimaging shows partial spontaneous involution over the course of 18 months (Fig. 1). The child continues to do well with 18 months of follow-up.

DISCUSSION

JXG affects infants and young children. Patients usually present with yellow-red skin nodules that regress spontaneously. Iris infiltration can cause hyphema and glaucoma. JXG pathology is notable for Touton giant cells. Immunohistochemistry shows positive staining for XIIIa, CD68, CD 163, fascin, and CD14 and negative staining for CD1A, GFAP, desmin, keratin, CD99, and S100.[6] Patients with systemic JXG (~4% of cases) may not have skin lesions, and histopathology often fails to demonstrate Touton giant cells. No consensus for treatment has been reached for orbital JXG. Options include observation, radiation, excision/debulking, and steroids.

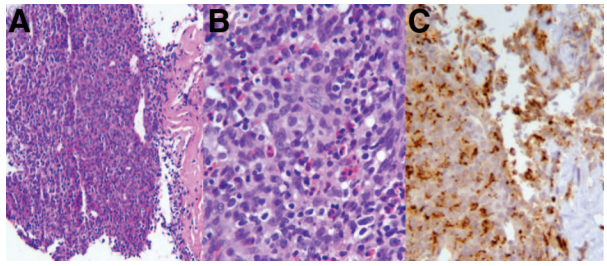

FIG. 2. Biopsy specimens showing histiocytes, small lymphocytes, and eosinophils from the **(A)** orbital and **(B)** maxillary sinus components of the mass (hematoxylin-eosin, ×40), and **(C)** positive CD68 staining of the orbital mass.

This is an unusual case of a non-LCH of the orbit, subtemporal fossa, maxillary sinus, middle cranial fossa, and cavernous sinus and appears to represent a rare cystic orbital JXG without cutaneous involvement. The differential diagnosis included venous vascular malformation, granulocytic sarcoma, and non-LCH. The diagnosis was made based on pathology studies and consultations, characteristic immunohistochemical staining, and systemic workup.

The patient continues to do well with more than 18 months of follow-up without treatment. Imaging studies show regression of the lesion, and visual and systemic development have been normal.

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Primary Adenoid Cystic Carcinoma of the Eyelid

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Abstract: **Adenoid cystic carcinoma is a rare epithelial malignancy, which tends to grow slowly. It is an intractable**

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neoplasm due to its ability to invade perineural spaces. A 59-year-old female presented with a gradually increasing mass in the right lower eyelid. An excisional biopsy with wide margins revealed a diagnosis of primary adenoid cystic carcinoma of eyelid skin with perineural invasion. Although a rare neoplasm, primary adenoid cystic carcinoma of eyelid skin should be included in the differential diagnosis of eyelid tumors.

Primary adenoid cystic carcinoma (ACC) is one of the rarest tumors of skin.[1] In the eyelid, it can arise from the adnexal glands, the palpebral lobe of the lacrimal gland, the accessory lacrimal glands in conjunctiva, or from ectopic lacrimal gland tissue.[2] We present a rare case of primary ACC arising from the lower eyelid skin.

CASE REPORT

A 59-year-old female presented with a swelling on the right lower eyelid since 1 year. There was gradual increase in the size of swelling since the past 6 months. There was no other significant history.

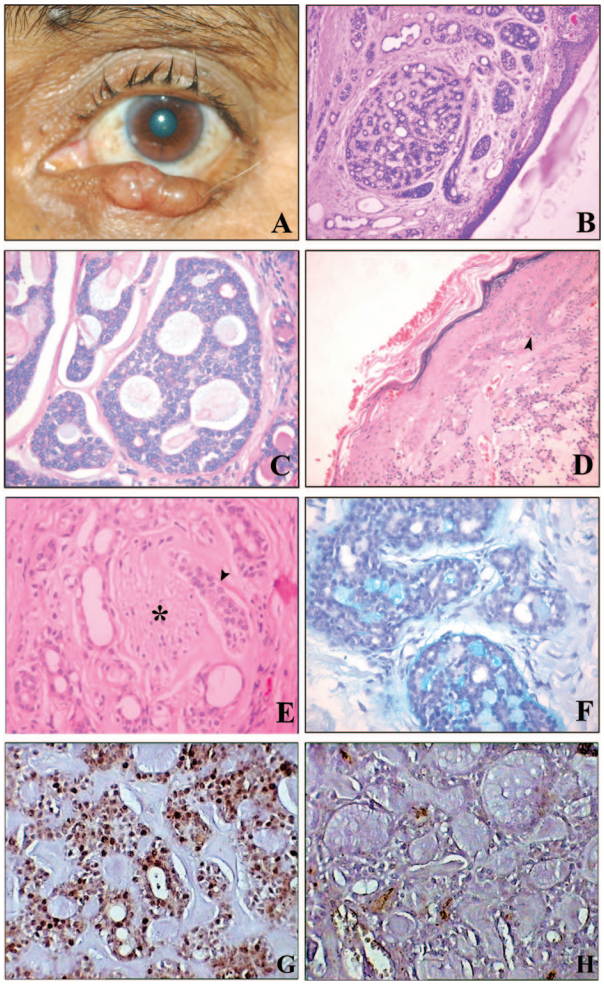
On examination, best-corrected visual acuity OU was 20/20. Anterior segment examination of the right eye revealed a nodular growth involving the central third of the lower eyelid. It was firm in consistency with well-defined margins measuring about 11 mm × 5 mm (Fig. A). It involved the eyelid margin with superficial vascularization, but there was no afferent conjunctival invasion. The rest of the anterior and posterior segment examination of the right eye was normal. Left eye examination was normal. There was no preauricular or submandibular lymphadenopathy. Systemic examination was normal. A provisional diagnosis of meibomian gland carcinoma was made. The patient underwent full-thickness excision biopsy of the mass along with 5-mm margins.

Histopathologic examination showed tumor tissue formed by epithelial and myoepithelial cells, with formation of pseudocysts and duct-like structures arranged in cribriform pattern (Fig. B and C). The neoplastic process seemed to originate from adnexal glands of skin. There was no continuity to conjunctiva; however, there were areas of epidermal infiltration (Fig. D) and perineural invasion (Fig. E). The pseudocysts contained eosinophilic material stained by Alcian blue (Fig. F). Immunohistochemical study was positive for epithelial membrane antigen (Fig. G) and negative for carcinoembryonic antigen (Fig. H) and S-100. The above findings were suggestive of primary ACC with perineural invasion. The surgical margins were free from tumor cells.

Thorough systemic evaluation with complete hemogram, liver, and kidney function tests were normal. CT scan of the head and neck revealed no lacrimal gland and no major or minor salivary gland masses. CT scan of the chest and abdomen revealed no abnormality. The patient underwent lower eyelid reconstruction. The patient is on close follow-up, without evidence of disease recurrence for 1 year of follow-up.

DISCUSSION

Primary ACC of the skin presents as a slow-growing cutaneous nodule without ulceration. It presents in elderly people with slight female preponderance. In a review of 37 cases of primary cutaneous ACC, the average age of presentation was 58.1 years and 23 (62%) were females.[3] The most common site of involvement is scalp (32%) followed by face, nose, chest, back, and abdomen.[3] It rarely occurs in eyelid and can clinically mimic meibomian gland carcinoma[2] or chalazion.[1,4]



A, Clinical photograph of the patient showing a nodular growth, firm in consistency involving the central third of the lower eyelid margin measuring about 11 mm × 5 mm with superficial vascularization. **B,** Hematoxylin-eosin staining with low power view (×10) showing pseudocysts and duct-like structures arranged in cribriform pattern formed by epithelial and myoepithelial cells. **C,** High power view (×40) showing cribriform pattern along with pseudocysts. **D,** Infiltration of tumor cells in epidermis (arrow head). **E,** High power view (×40) showing perineural invasion (asterisk) by tumor cells (arrow head). **F,** Alcian blue stain showing positivity to eosinophilic material in the pseudocystic spaces. **G,** Immunohistochemical examination showing positivity to epithelial membrane antigen (EMA) in the tumor cells. **H,** Immunostaining with carcinoembryonic antigen (CEA) was negative in tumor cells.

In review of literature on eyelid tumors, 5 cases of ACC are reported, 2 arising from upper eyelid and 2 from lower eyelid. In 2 such reports, the tumor was originating primarily from skin,[2,5] in one report the tumor was originating from accessory lacrimal glands,[4] and in the other 2 reports, no definite conclusion about the origin of tumor was offered.[1,6]

The most common differential diagnosis includes adenoid basal cell carcinoma.[2] Adenoid basal cell carcinoma and ACC both have an infiltrative growth pattern and tendency to invade perivascular and perineural spaces. However, ACC presents with lack of peripheral palisading of the nuclei and occasional presence of central apoptotic or necrotic cells.

Moreover, ACC pseudocysts stains positively to Alcian blue and periodic acid-Schiff.[2] The tumor also shows positive immunostaining to epithelial membrane antigen and amylase and negative immunostaining to S-100, carcinoembryonic antigen, and vimentin.[7]

Perineural invasion is considered an indicator of poor prognosis because of inherent risks of local recurrence and spread to skull base. Perineural invasion is a common feature of ACC arising from lacrimal gland with nearly all patients developing recurrence or metastasis with or without treatment. However, in all 5 case reports of ACC of the eyelid, none showed evidence of perineural invasion, which was one of the distinguishing feature of our case. The recurrence rate of cutaneous ACC is 51%[3]; however, none of the reported eyelid ACC recurred or developed metastasis. We have kept this patient on close follow-up to look for early signs of recurrence due to the invasive nature of the tumor.

The mainstay of treatment consists of local excision with wide margins. Radiotherapy and chemotherapy may be used when complete excision is not possible or for locally aggressive and potentially metastasizing carcinoma. Long-term follow-up is necessary because perineural extension may be discontinuous and can lead to false-negative margins with a higher local recurrence rate.

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Endoscopic Removal of Nasoglabellar Dermoid Cysts

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Abstract: **Dermoid cysts are common tumors resulting from entrapped ectodermal elements at fetal suture lines. Management is conceptually straightforward, with surgical excision of the mass in its entirety without rupture. The conspicuous location and potential scarring from direct excision can be objectionable, particularly in children. The authors describe 2 cases using a hidden hairline incision and an endoscopic approach to remove dermoid cysts in the nasoglabellar region.**

Dermoid cysts result from entrapped ectodermal elements at fetal suture lines, are lined by squamous epithelium, and contain adnexal structures. These cysts usually arise at the frontozygomatic suture near the lateral canthus but occasionally arise from the frontonasal suture in the nasoglabellar region.[1]

Management of dermoid cysts requires complete surgical excision without rupture. However, ruptures occur not uncommonly, and incomplete removal may promote recurrence.

The conspicuous location of these lesions, particularly in the nasoglabellar region, makes direct excision with resultant scarring undesirable. Several approaches have been described to minimize visible incision lines by using brow and eyelid crease incisions.[2–4] We describe an endoscopic approach, which allows for a small, well-hidden incision in the hairline, while providing adequate access to the nasoglabellar region.

Case 1. A 6-month-old female presented for evaluation of 2 forehead masses that were noted shortly after birth. Past medical and surgical histories were unremarkable, and her ocular examination was within normal limits. External examination revealed a soft, immobile mass measuring 1 × 1.5 cm that was located medially near the radix of the nose, and a smaller, glabellar mass laterally (Fig. 1). CT scanning showed no attachment to the frontal sinus.

Case 2. A 19-month-old male presented with a mass on his forehead that had recently increased in size. Past medical and surgical histories were unremarkable, and his ocular examination was within normal limits. External examination revealed a soft, nontender 2 × 2-cm mass that was located in the nasoglabellar region. CT scanning revealed a subcutaneous low-density nodule in the frontal scalp without evidence of bony erosion (Fig. 2).

Description of Procedure. All cases were performed in the operating room under general anesthesia. The forehead and scalp were injected with local anesthetic (0.5% lidocaine with 1:100,000 epinephrine). A 2-cm incision was placed posterior to the hairline for entry of surgical instruments and the endoscope.

Subperiosteal blunt dissection was maintained to the arcus marginalis and superior orbital rim, medial to the supraorbital notch. Using endoscopic scissors, periosteum was opened from the underside of the lesion. Digital counter pressure from the overlying external skin over the lesion was used to help identify and dissect the mass. A combination of elevation, sharp, and blunt dissection with endoscopic peri-

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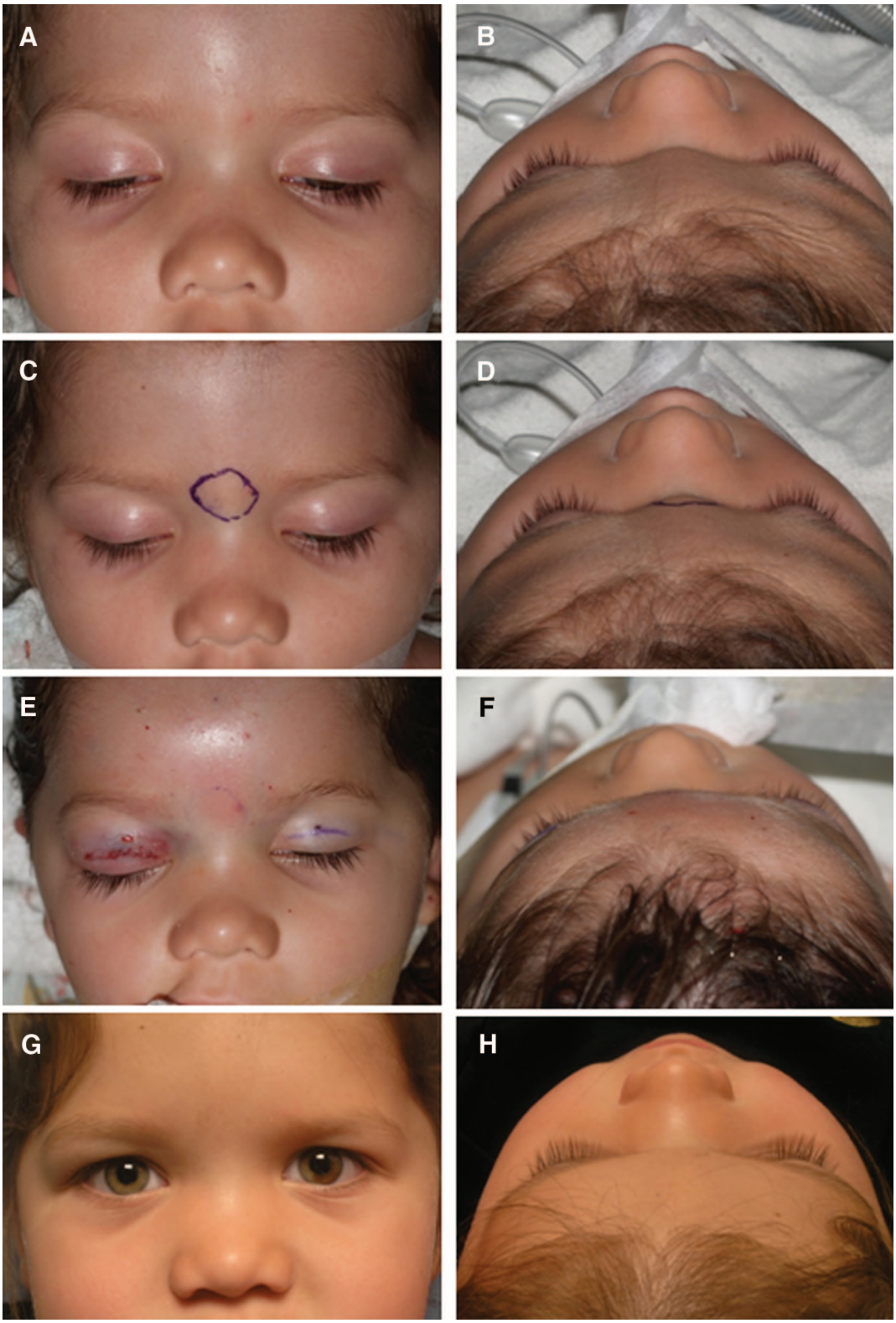

FIG. 1. Case 1. Preoperative (**A–D**), immediately postoperative (**E, F**), and 3-month postoperative images (**G, H**) of a 6-year-old female with a nasoglabellar dermoid cyst, which was removed with a combined endoscopic-assisted and eyelid crease approach. No recurrences were noted.

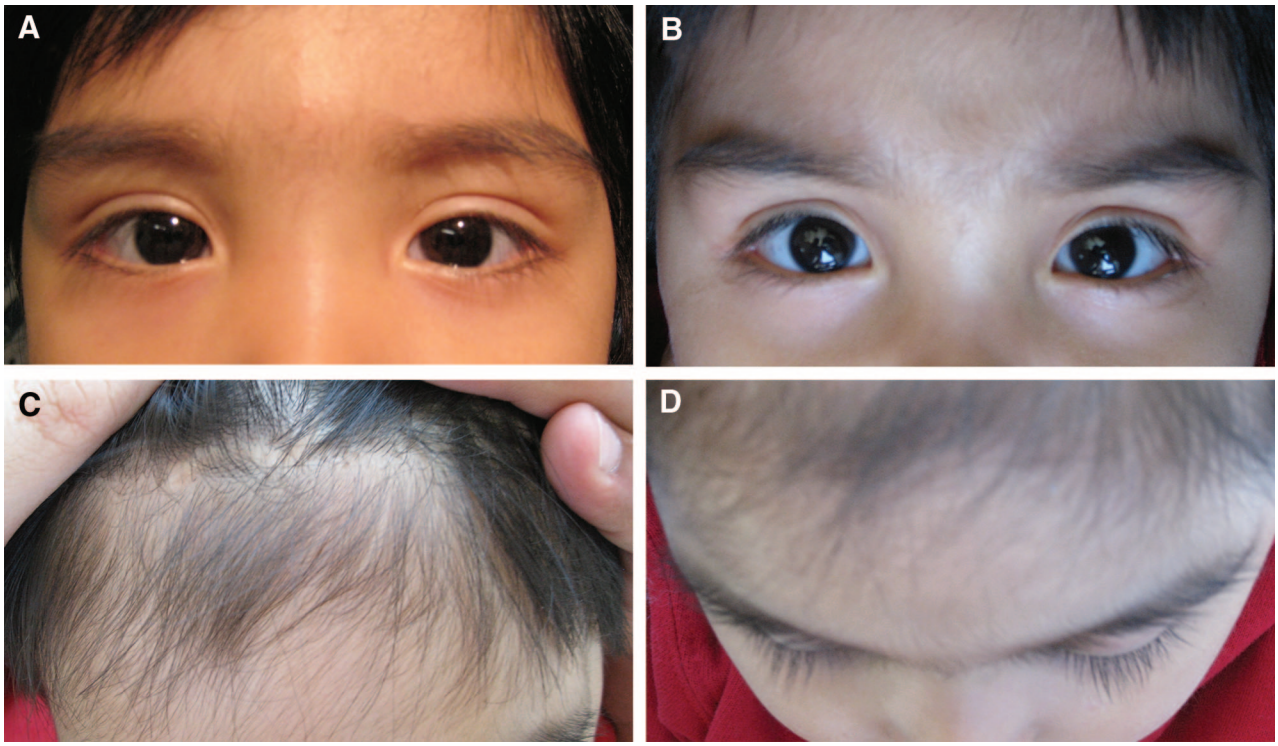

FIG. 2. Case 2. Preoperative (**A**) and 11-month postoperative images (**B–D**) of a 19-month-old male with a nasoglabellar dermoid cyst, which was removed with an endoscopic-assisted approach. No recurrences were noted with good cosmesis.

osteal elevators and scissors were used to free and excise the cyst in its entirety (Fig. 3). In the first case, an additional right upper eyelid crease incision was made for bimanual manipulation of the cyst.

DISCUSSION

Important considerations in the management of periorbital dermoid cysts include accurate diagnosis, complete

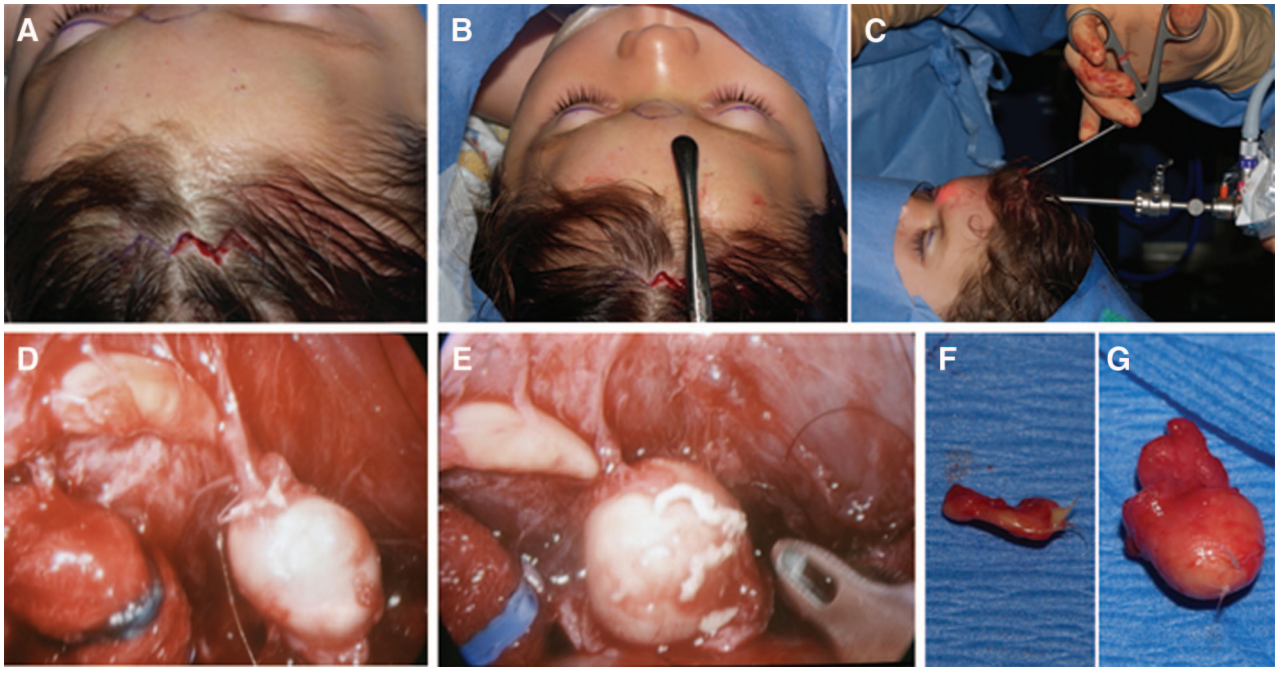

FIG. 3. Case 1. (**A–C**) A 2-cm incision was placed slightly posterior to the hairline to allow for entry of surgical instruments and the endoscope. (**D, E**) The dermoid cyst as visualized through the endoscope. (**F, G**) A combination of elevation, sharp, and blunt dissection with endoscopic periosteal elevators and scissors were used to free and excise the cyst in its entirety.

removal, and minimization of postoperative scarring, given the conspicuous location of these lesions. Imaging is an important component in the evaluation of midline nasal lesions, particularly if an endoscopic approach is considered, as the differential diagnosis is broad and intracranial extension may be present.

Traditional surgical approaches conceal the incision near the eyebrow and yield good results.[4] Nevertheless, this approach may be suboptimal for those patients with a dermoid cyst in a nasoglabellar location and for individuals with a tendency to develop keloids or hyperpigmented scars. Also, additional incisions may be necessary for those patients with multiple lesions in geographically distinct locations.

An upper eyelid crease approach has also been described with good success and excellent cosmesis.[2,3,5] Cozzi et al.[5] reported superior scar quality with this technique in a study comparing 26 patients who underwent a transbrow incision with 8 patients who underwent an upper eyelid crease incision for angular dermoid cyst excision. Access to lesions in the central brow or nasoglabellar area, however, may be limited with this approach.

Endoscopic techniques continue to evolve, and its described uses continue to expand. This approach optimizes aesthetic outcome by concealing the small port scar to a less noticeable location on the scalp.[6,7] This technique also allows for adequate access to the nasoglabellar and mid-forehead regions, areas not readily accessible via the eyelid crease. This was an important consideration in our first case. Disadvantages include the need for new instrumentation, limited space for surgical manipulation, and potentially longer operative times (1.5 to 2 hours in our cases). Potential complications include a visible scar in the setting of future baldness and nerve/vessel injury. In our cases, no functional or aesthetic complications were noted.

An endoscope-assisted approach may be a viable alternative technique to access periorbital dermoid cysts in the nasoglabellar area, with good cosmesis, adequate exposure, and low rates of significant sequelae.[6,7] It may be particularly useful for those patients with multiple, geographically separate lesions, and may be combined with a traditional superior eyelid crease if necessary.

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