sheets of oval to spindle myoepithelial cells, was present in all 5 of the previously described cases.1-4 The first case noted areas of central calcification in an irregularly bordered, partially encapsulated tumor with numerous clear cells and sparse ductal structures associated with pleomorphic adenoma.3 The case reported by Chan et al.6 showed a low-grade encapsulated tumor. Focal necrosis, cellular atypia, and enlarged nucleoli were noted by Wiwatwongwana et al.7 in a T2N0M0 tumor. Singh et al.1 described frequent mitotic figures in the more solid areas of the tumor and only occasional mitotic figures in the ductal epithelial portions.

All prior cases displayed histologic evidence of disease at the surgical margin, but lacked lymphovascular invasion. Three of the reported cases, as well as the described case, displayed myoepithelial anaplasia. Wiwatwongwana et al.7 described the only tumor to show necrosis, while Singh et al.1 described the only tumor to exhibit perineural invasion. The described case is the first report of a tumor displaying both necrosis and perineural invasion.

The authors herein describe what may be the first reported case of a carcinoma ex pleomorphic adenoma of the lacrimal gland with epithelial–myoepithelial carcinoma type that presented with rapid visual loss from compressive optic neuropathy. A multidisciplinary team should be recruited to properly diagnose and treat such patients, with complete surgical excision attempted when feasible with postoperative radiotherapy.

REFERENCES

Arteriovenous Malformation of the Eyelid: Surgical Management and Histologic Study

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Abstract: A raised erythematous eyelid lesion that appeared in a 31-year-old man was diagnosed as an arteriovenous malformation (AVM), with confirmatory Doppler ultrasound demonstrating high arterial flow. Surgical excision, aided by electrocautery for extensive hemorrhage, resulted in an acceptable cosmetic result. Histopathology of the excised lesion showed collapsed capillary channels lined by endothelium. AVM is rarely encountered in the eyelid.

A healthy 31-year-old man noted a gradually enlarging, painless red mass of the right upper eyelid for 3 months. There was no history of trauma or hemorrhage. The lesion swelled acutely with exercise. Initially diagnosed as a hordeolum, warm compresses and a topical antibiotic-steroid ointment were prescribed without improvement. There was a raised, soft, erythematous, cutaneous lesion that measured 6 mm × 8 mm near the right upper eyelid margin (Fig. 1). The mass was surrounded by a network of engorged, nonpulsatile vascular channels that collapsed with digital compression on the dome-shaped portion and briskly refilled with the release of pressure. The eyelid margin had normal contour, showing no ulceration, pigmentation, or madarosis. There was no exophthalmos. Applanation tensions were 14 mm Hg bilaterally without increased excursions of the tonometer on the involved side. No other ocular abnormalities were present, and no similar cutaneous lesions were noted elsewhere on the body or reported in his family. Timolol gel was applied to the lesion without resulting regression.

Orbital MRI with contrast showed a lesion with abnormal signal intensity that was predominantly hyperintense on long TR sequences with eyelid enhancement (Fig. 2). There were no associated flow voids, orbital involvement, or other orbital abnormalities. Doppler ultrasound demonstrated a soft tissue structure with exuberant arterial flow and venous shunting, suggestive of an arteriovenous malformation (AVM); (Fig. 3). According to the Schöbinger scale of superficial AVM, the lesion was designated grade II (expanse lesion with dilated draining veins). The patient declined recommendations of angiography with possible embolization and sclerotherapy.

Surgical excision involved injection of the eyelid with 2% lidocaine with 1:100,000 epinephrine. The upper eyelid crease was incised along its middle third, and a subcutaneous dissection plane was created, extending to the base of the lesion. Large venous vessels were observed coursing within the orbicularis oculi, and copious bleeding was controlled by bipolar cautery. Skin and muscle were dissected bluntly away from the lesion. The spherical malformation collapsed following coagulation of the arterial feeders, and the lesion was excised. The skin incision was closed with a running subcuticular 6-0 prolene...

FIG. 1. A and B, Raised erythematous lesion near eyelid margin with feeder vessels. C and D, Postoperative result with slight eyelid retraction.
FIG. 2  A, Coronal MRI T1-weighted image shows minimal intensity in upper eyelid region. B, Marked hyperintensity appears on T2-weighted imaging. C, Sagittal T1-weighted postcontrast imaging shows intense enhancement of the lesion.

FIG. 3  A and B, Doppler ultrasound shows high arterial blood flow.

FIG. 4  A and B, Blood-free vascular channels following digital pressure and cautery of feeder vessels (hematoxylin-eosin). C, CD31 immunostain highlights endothelial linings. D, Smooth muscle actin immunostain is positive in capillary walls. E, FLI1 immunostain identifies endothelial nuclei (A, original magnification ×200; B–E, original magnification ×400).

suture. One week postoperatively, a zone of skin ulceration and crusting was noted and treatment with warm compresses and antibiotic-steroid ointment was instituted. Slow healing of the wound occurred over 2 weeks with granulation tissue closing the defect, resulting in slight eyelid retraction (Fig. 1C,D).

The excised lesion was a tan-gray, rubbery mass that measured 0.3 × 0.2 × 0.1 mm. Histopathology showed a cluster of capillary-sized vessels with endothelium that stained positively with CD34, CD31, and FLI1 immunostains. Some vessels stained positively with smooth muscle actin (Fig. 4). Immunostain for herpesvirus 8 was negative, ruling out Kaposi’s sarcoma.

DISCUSSION

AVMs are congenital or acquired vascular anomalies that occur uncommonly within the orbit. Arteries and veins are present with or without a network of intervening capillary-sized vessels that may be primary in origin or reflect “secondary” capillary hyperplasia. This nidus of capillaries may assume tumor-like configurations, as in the current case, and is histologically indistinguishable from capillary hemangioma unless large arteries or veins are included in the excised tissue. Pure preseptal eyelid involvement is rare and—to the authors’ knowledge—only 13 reports are present in
the literature,2–8 with none containing histologic illustration of the nidus. Nearly all palpebral AVMs involve the upper eyelid and are pulsatile. The reasons for absent pulsations in the current case and case 3 in the Hayes report4 are unclear but may relate to an undetected thrombotic occlusion. Previous trauma is etiologic for some AVMs,5,8 although most are presumed to have a congenital origin. Vascular syndromes such as Wyburn-Mason, Sturge-Weber, and hereditary hemorrhagic telangiectasia should be ruled out.

Clinical diagnosis is aided by pulsations, bruits, and warmth of the lesion. Doppler ultrasound demonstrates high arterial flow, and digital subtraction angiography may disclose supply by such vessels as the ophthalmic artery or superficial temporal artery. MRI studies often show hypointense arterial flow voids but may be inconclusive for very small lesions.

Treatment is challenging and involves combinations of sclerosing injections,9 embolization techniques, and surgery.4–10 While marked intraoperative hemorrhage may be avoided by prior embolization, the latter technique may result in retinal ischemia and blindness.7 The current case illustrates a successful direct surgical approach.

REFERENCES


Abstract: Orbital arteriovenous malformations are rare congenital vascular anomalies that can cause eyelid distortion, proptosis, diplopia, and vision loss. The current treatment paradigm involves endovascular embolization followed by resection, with delayed reconstruction. The authors report 2 young adult patients with orbital arteriovenous malformations, who underwent embolization followed days to months later by resection with immediate soft-tissue reconstruction. For both patients, the immediate outcome provided good functionality and cosmosis. The authors conclude that immediate reconstruction after embolization-resection of orbital arteriovenous malformations may facilitate early return of form and function for patients, improve quality of life, and potentially reduce the need for further reconstructive procedures.

CASE REPORTS

Case 1

Presentation. A 21-year-old man presented for evaluation with a left orbital AVM involving the left upper eyelid, left brow, superior orbit, and scalp (Fig. 1A). He had undergone multiple laser treatments and resections/reconstructions of his forehead and brow AVMs, and 18 months prior to presentation, he had undergone embolization of his orbital AVM with 70% obliteration of the nidus. His visual acuity was normal, although the severity of the ptosis precluded use of the eye. MRI/A showed preseptal AVM in left upper eyelid with anterior involvement of the superomedial orbit and posterior intraconal extension (Fig. 1B, C). Feeder vessels included left facial, superficial temporal, and ophthalmic arteries. The goal of treatment was to restore use of his left eye.

Treatment Course. Patient underwent sequential superselective angiographies with embolizations (4:1 Ethiodol:n-BCA glue) and sclerotherapy (bleomycin or dehydrated alcohol mixed with Ethiodol) of left external carotid feeder vessels and distal left ophthalmic artery and intralesional sclerotherapy of the superior and medial aspects of the AVM, leading to extensive and prolonged period of inflammation (Fig. 1D). Once the key feeders were addressed and the inflammation somewhat subsided, the patient underwent anterior orbitotomy through a eyelid crease incision for en bloc excision of the eyelid and anterior orbital AVM, with immediate left upper eyelid reconstruction. Intraoperatively, there was a multitude of small interconnected vessels within and just posterior to the orbital septum, with an extensive network of collaterals with the marginal and peripheral arcades. The collaterals were mostly cauterized with bipolar cautery. Larger (dilated diseased) vessels were identified and ligated with multiple silk sutures both superomedially and centrally just deep to the superior orbital rim. Most of the dilated AVM vessels had limited blood flow secondary to the prior embolization and sclerosing treatment, although very few were fully embolized. Tissue cutting utilized both the monopolar unit and scissors, the latter most useful for cutting through glued vasculature. Arteriovenous malformation debulking was guided by intraoperative stereotactic navigation and left upper...