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CASE REPORT

Craniofacial/Pediatric

Arteriovenous Malformation of the Upper Eyelid: A Case Report

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Summary: Arteriovenous malformations (AVMs) are rare lesions formed by direct connection of arteries and veins bypassing capillary beds. AVM locations can vary; however, extracranial AVMs remain the least common. The vast majority of these malformations are congenital and can be triggered by hormonal changes. A 49-year-old woman presented with a progressive right-sided upper eyelid swelling over the past 12 years. The patient had mechanical ptosis and mild impairment of the right visual field. The swelling was compressible with a thrill, and bruits were heard. The patient was diagnosed with AVM using cerebral angiography. Preoperative endovascular embolization was not possible; however, surgical excision was successfully done with no complications. AVMs and fistulas are high-flow vascular lesions that usually occur during childhood and progress with time. Only a few cases of face and eyelid AVMs have been reported in the literature. Diagnosis of these anomalies is mostly made based on clinical presentation and radiographic imaging such as angiography. Treatment options remain controversial, and management should be individualized for each patient. Endovascular embolization followed by surgical excision is advisable in many cases. Due to its rare entity, information regarding the best management options for AVMs is limited. Reducing the risk of bleeding and achieving total resection is the goal of treatment. Long-term follow-up is required in these patients because recurrent cases of AVMs have been reported.
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Vascular anomalies are a group of lesions that originate from blood vessels and lymphatic tissue. They constitute the most common congenital anomalies in infants and children.[1] Arteriovenous malformations (AVMs) are considered rare vascular lesions formed by the direct interconnection between arteries and veins bypassing capillary beds.[1] Most of these lesions are located intracranially as reported in the literature, while extracranial arteriovenous malformations remain less common.[2] These malformations persist or progress with time and are usually triggered by hormonal changes or trauma.[3,4] Management options include embolization, electro thrombosis, surgical resection, or a combination of 2 of these methods. Complete surgical excision is often not achievable due to AVMs' diffusive nature, which involves different tissues, and their location in crucial anatomical areas.[5,6]

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CASE REPORT

A 49-year-old woman presented with a painless swelling on her upper right eyelid, which had gradually progressed since she was 15 years old. The swelling was initially small, with no change in size until 12 years previously. On examination, the swelling was covering most of the right eyelid but mainly extended to the lateral side toward the right forehead and hairline. It was causing mechanical ptosis covering the upper pupillary region (Fig. 1). By palpation, the swelling was compressible with a thrill. Moreover, bruits were heard upon auscultation. Extraocular muscles were intact in both eyes; however, the right upper visual field is mildly compromised due to ptosis. The patient was sent for diagnostic

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Fig. 1. Preoperative presentation showing right upper eyelid swelling.

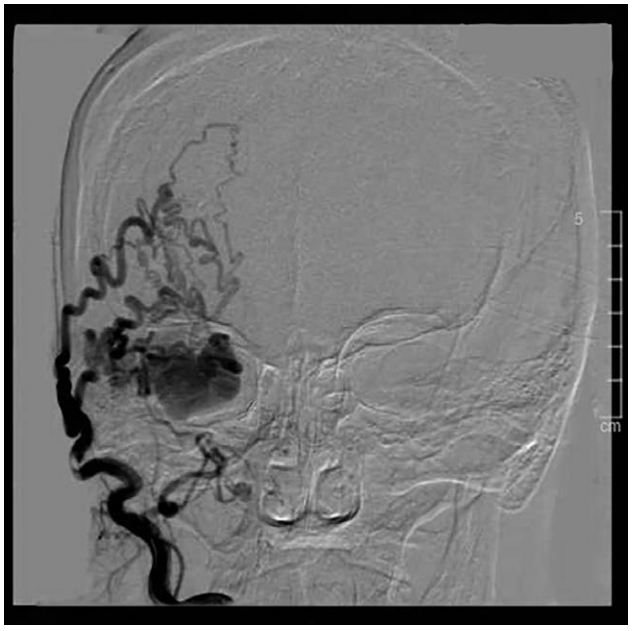

Fig. 2. Selective catheterization of the right external carotid artery showing prominent enlarged arteries coursing anteriorly. These arteries are likely the frontal branch of the superficial temporal and the transverse facial arteries. An early filing large ectatic vein, suggestive of shunting, is seen projecting over the right upper eyelid, likely a superior palpebral vein. The site of shunting is at the medial aspect of the orbit.

cerebral angiography as well as computed tomography (CT) angiogram and the diagnosis of a right facial arteriovenous fistula was made (Fig. 2). Endovascular

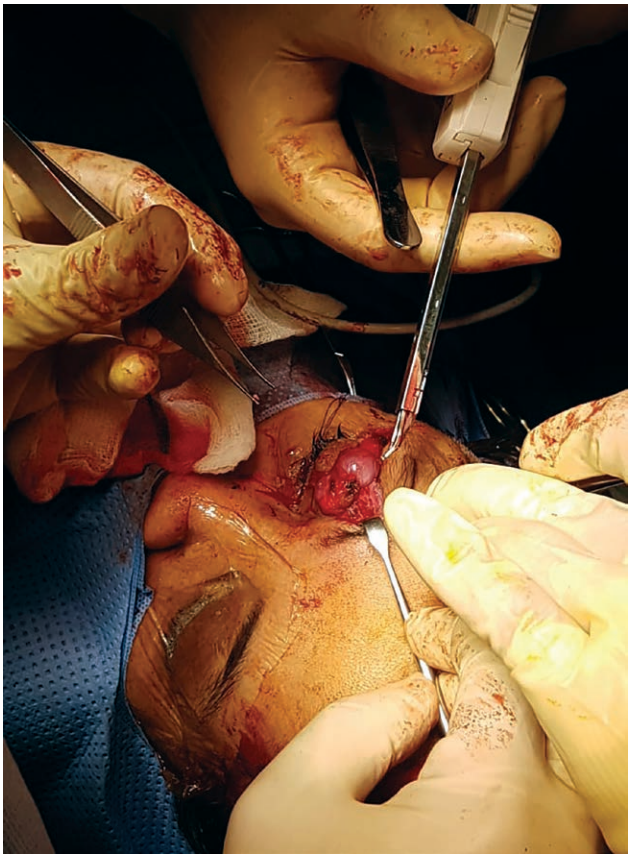

Fig. 3. Upper blepharoplasty incision (debulking of the intraorbital mass, extension with clipping of the feeding vessels, and removal of excess skin).

embolization followed by surgical resection was planned. Unfortunately, embolization was not possible due to very tortuous feeding arteries. During surgery, a bicoronal incision was made, followed by dissection of the subperiosteal midline and supramuscular to the temporal region. Clipping of the temporal feeding vessels was achieved. Moreover, a right upper blepharoplasty incision was made, with excision of the skin and intramuscular dissection. Separation of orbicularis muscle followed by dissection of the pouch with clipping of the major feeding vessels was achieved (Fig. 3). The advantages of performing simultaneous blepharoplasty incision are the ability to visualize and excise the entire lesion, correct the mechanical ptosis, and achieve a pleasant aesthetic result. The excised lesion was sent for histopathological confirmation. Dilated vascular spaces consistent with arteriovenous malformation were reported.

The postoperative course showed no complications, and CT angiogram showed no shunting. The patient was stable, with normal eye movements and intact facial nerve. The patient was followed up for 1 year and showed excellent results with no recurrence or other complaints (Fig. 4).

DISCUSSION

In 1982 Mulliken and Glowacki introduced a biological classification differentiating between haemangiomas and

Fig. 4. Follow-up of the patient showing no recurrence.

vascular malformations.[7,8] Haemangiomas are neoplastic lesions that demonstrate endothelial hyperplasia; they present at birth and usually involute with time. Conversely, vascular malformations lack this cellular proliferation. Vascular malformations are further classified based on the blood flow into high flow lesions (AVMs and Fistulas) and low flow lesions (Venous and lymphatic malformations). Differentiating between the 2 types is important for proper intervention.[9] AVMs mostly present at birth with a slight female predominance (1:1.5). These lesions usually become evident during childhood but can present during the second, third, or even the fifth decade of life.[10] Vascular malformations of the head and neck are rare lesions. Several cases have reported unusual locations, including the face, lips, and eyelids.[10–12] Our patient presented with a face AVM specifically located on the right upper eyelid. To our knowledge, only a few cases of eyelid AVMs have been reported to date.[13] Diagnosis of these lesions is primarily made based on the patient's history and clinical presentation. AVMs mostly present as painful pulsating swelling, and a bruit is usually heard over the lesion. Patients with AVMs may have a spontaneous hemorrhage or bleeding after dental procedures as their primary presentation.[5] Radiographic evaluation, including Doppler ultrasonography, CT, and magnetic resonance imaging, is helpful to identify the exact location and for the assessment of the flow dynamics. However, catheter angiography remains the gold standard test.[11]

Surgical excision, endovascular embolization, laser therapy, and combination therapy are different approaches to treating AVMs. According to the literature, treatment should be individualized for each patient. Cases treated with endovascular embolization followed by surgical excision showed the highest success rates since embolization is used to minimize hemorrhage during resection.[11,12] However, the embolization method carries its risks. A previous study has reported a patient with eyelid AVM who had developed retinal artery occlusion after being treated with embolization.[14] Another patient refused to undergo embolization and was successfully treated with surgical excision only.[13] As in our case, endovascular embolization of the feeding vessels before surgery was not possible as discussed earlier. However, the following surgical excision was successful with minimal bleeding. Total surgical excision remains the goal of treatment in head and neck AVMs. Reports emphasize on the importance of complete resection, as incomplete embolization without resection can lead to recurrence of the lesion through recruitment of new feeding arteries. Nonetheless, total excision gives better cosmetic results.[6,12] Because these lesions have a tendency to recur, follow up is recommended for all patients with AVMs.[10,12]

CONCLUSIONS

Arteriovenous malformations of the face are rare congenital lesions that progress with time. Proper history, clinical examination, and different imaging modalities are required for an accurate diagnosis. Treatment should be individualized according to the patient's needs and the complexity of the lesion. Total excision remains the goal of treatment. A preoperative endovascular embolization, if possible, can be used to minimize the bleeding and achieve better outcomes. Patients with AVMs have a risk of recurrence. As a result, follow-up is advisable for these patients.

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PATIENT CONSENT

The patient provided written consent for the use of her image.

REFERENCES

1. Raghu M, De R, Higgins N, et al. Spontaneous arteriovenous malformation of the external auditory meatus. *J Laryngol Otol.* 2004;118:912–913.
2. Burrow PE, Mulliken JB, Fellows KE, et al. Childhood hemangiomas and vascular malformations: angiographic differentiation. *AJR.* 1983;141:483–488.
3. Bhat VS, Aroor R, Satheesh Kumar Bhandary B, et al. Traumatic arteriovenous malformation of cheek: A case report and review of literature. *Int J Otorhinolaryngol Clin.* 2013;5(3):173–177.
4. Legiehn GM, Heran MK. A Step-by-step practical approach to imaging diagnosis and interventional radiologic therapy in vascular malformations. *Semin Intervent Radiol.* 2010;27:209–231.
5. de Carvalho MF, Kaline de Oliveira N, Duailibi E, et al. Extensive arteriovenous malformation in the face of a pediatric patient – Case report. *Rev Port Estomatol Med Dent e Cir Maxilofac.* 2014;55:250–255.
6. Gupta AK, Iyengar SN, Sharma A. Scalp arteriovenous malformation: a case report with review. *Rom Neurosurg.* 2018;32:151–155.
7. Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. *Plast Reconstr Surg.* 1982;69:412–422.
8. Ethunandan M, Mellor TK. Haemangiomas and vascular malformations of the maxillofacial region—a review. *Br J Oral Maxillofac Surg.* 2006;44:263–272.

9. Kitagawa A, Yamamoto T, Matsunaga N, et al. Polidocanol sclerotherapy combined with transarterial embolization using n-butyl cyanoacrylate for extracranial arteriovenous malformations. *Cardiovasc Intervent Radiol.* 2018;41:856–866.
10. Karankot G, Arutla R, Rajan R, et al. Multifocal arteriovenous malformation of face: a rare entity. *J Indian Acad Oral Med Radiol.* 2020;32:77.
11. Shobeirian F, Sanei Taheri M, Yeganeh R, et al. Huge arteriovenous malformation of upper lip- A case report. *Iran J Otorhinolaryngol.* 2020;32:49–52.
12. Molavi B, Tafti MF, Sinaei F, et al. A rare extracranial arteriovenous malformation of the face: a case report. *Rev Port Estomatol Med Dent e Cir Maxilofac.* 2019;60:13–17.
13. Lo C, Petris CK, Haberman I, et al. Arteriovenous malformation of the eyelid: surgical management and histologic study. *Ophthalmic Plast Reconstr Surg.* 2017;33(3S suppl 1):S138–S140.
14. Shaver J. Eyelid arteriovenous malformation treated with embolization leading to a branch retinal artery occlusion. *Optometry.* 2011;82:744–750.