An Unexpected Presentation of Orbital Cavernous Venous Malformation

Miles Menuck OMS-IV, Steven Jones MD
Corewell Health Farmington Hills Diagnostic Radiology

Introduction:
Orbital cavernous venous malformation (OCVM) – also known as orbital cavernous hemangioma or orbital cavernoma – is the most common orbital vascular tumor, accounting for 5-9% of all orbital tumors. This lesion is typically found in the intraconal space, and is composed of dilated vascular spaces without cellular proliferation[1,2]. CT shows a round or ovoid mass with variable enhancement, while MRI shows a T1 isointense, T2 hyperintense lesions with progressive enhancement [3]. Here we present a case of a cavernous hemangioma seen in an atypical location; within the nasolacrimal duct.

Case Presentation:
The patient is a 55 year old male with no significant past medical history who presented to otolaryngology with a complaint of a right medial orbital lesion. Initial imaging with a maxillofacial CT with IV contrast (Figure 1) showed a 1.4cm oval-shaped lesion centered in the nasolacrimal duct with bony expansion but no erosion and heterogeneous arterial enhancement. Subsequent MRI with and without IV contrast (Figure 2) showed a T1 hypointense, T2 hyperintense lesion with avid postcontrast enhancement. The lesion was excised by otolaryngology and oculoplastics, and the pathology report confirmed orbital cavernous venous malformation.

Discussion:
While OCVM is a relatively common entity, the location seen in this case was unexpected. One case series found that only approximately 14% of OCVMs were seen in the extraconal space, and none of the cases in this report involved the nasolacrimal duct [4]. Despite the atypical location, our patient went on to have successful excision of the lesion with resolution of his symptoms. Surgical resection is the treatment of choice for OCVMs, with recurrence reported only rarely, and likely due to either incomplete resection or presence of a separate, smaller lesion[2]. For patients who do not wish to undergo surgery or in whom the lesion location make surgery difficult, an alternative treatment option is sclerotherapy with pingyangmycin, which can reduce lesion volume and thus symptoms and has been found to be safe in at least one case series [5].

Conclusion:
OCVM is a relatively common lesion with excellent prognosis following surgical resection. Familiarity with this entity can help lead to prompt and accurate diagnosis, and knowledge of less common locations can prevent diagnostic confusion and unnecessary biopsies or additional imaging.

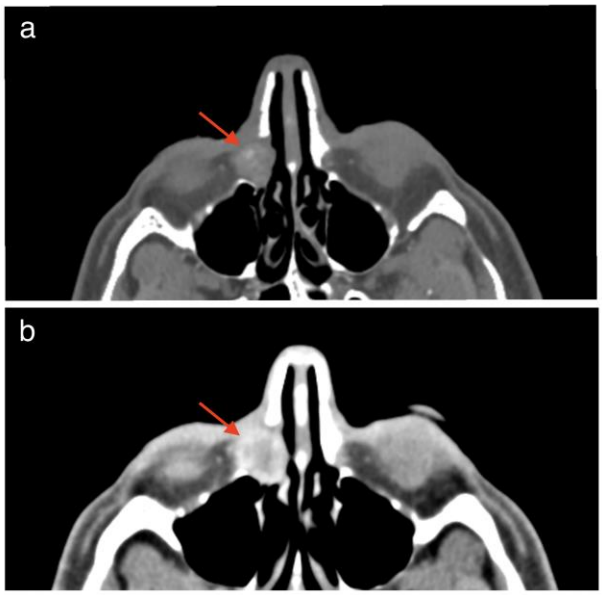

Figure 1: Maxillofacial CT in arterial (a) and delayed (b) phases showing the lesion centered in the right nasolacrimal duct with heterogenous arterial and homogenous peripheral delayed enhancement.

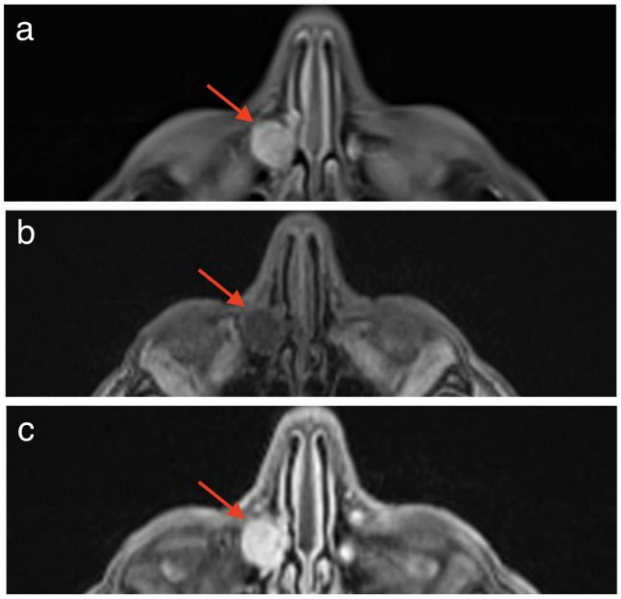

Figure 2: T2 (a), T1 precontrast (b) and T1 postcontrast (c) images show the same lesion with homogenous T2 hyperintensity and T1 hypointensity with avid, relatively homogenous postcontrast enhancement.

Citations:

1. Bilaniuk LT. ORBITAL VASCULAR LESIONS: Role of Imaging. *Radiol Clin North Am*. 1999;37(1):169-183. doi:10.1016/S0033-8389(05)70085-3

2. Calandriello L, Grimaldi G, Petrone G, et al. Cavernous venous malformation (cavernous hemangioma) of the orbit: Current concepts and a review of the literature. *Surv Ophthalmol*. 2017;62(4):393-403. doi:10.1016/j.survophthal.2017.01.004

3. Tanaka A, Mihara F, Yoshiura T, et al. Differentiation of Cavernous Hemangioma from Schwannoma of the Orbit: A Dynamic MRI Study. *Am J Roentgenol*. 2004;183(6):1799-1804. doi:10.2214/ajr.183.6.01831799

4. McNab AA, Selva ,Dinesh, Hardy ,Thomas G., and O'Donnell B. The Anatomical Location and Laterality of Orbital Cavernous Haemangiomas. *Orbit*. 2014;33(5):359-362. doi:10.3109/01676830.2014.915329

5. Yue H, Qian J, Elner VM, et al. Treatment of orbital vascular malformations with intralesional injection of pingyangmycin. *Br J Ophthalmol*. 2013;97(6):739-745. doi:10.1136/bjophthalmol-2012-302900