



Multi-compartment skull base orbital cavernous venous malformation: A rare presentation of a common orbital mass

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ARTICLE INFO

Keywords:

Multicompartmental cavernous venous malformation
Endoscopic
Transorbital
Endonasal
Multi-disciplinary
Cavernous hemangioma
Cavernoma
Trigeminal nerve tumor

ABSTRACT

Purpose: We present a unique case of an orbital intraconal cavernous venous malformation that extended along the trigeminal nerve to the pterygopalatine and middle cranial fossa. Our aim is to describe an atypical presentation of this common orbital vascular mass.

Observations: A 57-year-old female presented with right eye proptosis. Orbital magnetic resonance imaging demonstrated a lobulated contrast-enhancing mass involving the right intraconal orbital space, pterygopalatine fossa, and right middle cranial fossa, radiographically presumed to be a schwannoma. Intraoperative and histopathologic evaluation confirmed a cavernous venous malformation that extended along the trigeminal nerve. The mass, including its attachments to the cranial nerves and dura, was successfully removed via a combined transorbital and endoscopic endonasal approach. The patient recovered well with 20/20 vision, full extraocular movements, and resolution of proptosis.

Conclusions: This a rare presentation of an orbital cavernous venous malformation not previously described. Cavernous venous malformations typically present as ovoid well-circumscribed lesions; however, they can also extend outside the orbit along the path of cranial nerves, as was observed in this case. These types of lesions should be included in the differential diagnosis of masses arising from or extending along cranial nerves, even when involving the orbit.

1. Introduction

Cavernous venous malformations (CVMs) are the most common primary orbital lesions in adults with an incidence of 4–9% among orbital neoplasms.^{1,2} They typically present with progressive axial proptosis and preferentially involve the intraconal orbital space.³ Orbital CVMs are slow-growing ovoid masses that differ from those found elsewhere in the body in that they are well-circumscribed with a distinct fibrous pseudocapsule and rarely undergoes spontaneous hemorrhage.⁴ We report a rare presentation of an orbital CVM, that extended along the maxillary division of the trigeminal nerve between the orbit and middle cranial fossa, as well as a multidisciplinary surgical approach to this complex bilobed mass.

2. Case report

A healthy 57-year-old female presented to the ophthalmology clinic with right eye proptosis. Visual acuity was 20/25 in both eyes with normal pupils and confrontation visual fields and –1 restriction to downgaze in the right eye. Hertel exophthalmometry demonstrated 3 mm of proptosis and resistance to retropulsion. Optical coherence tomography of the optic nerve and Humphrey visual field 24-2 showed no evidence of optic nerve compression. Computed tomography (CT) and magnetic resonance imaging (MRI) demonstrated a lobulated contrast-enhancing intraconal orbital mass abutting the optic nerve in the orbital apex and extending through the inferior orbital fissure, pterygopalatine fossa and middle cranial fossa next to the carotid artery and extradural cavernous sinus (Fig. 1A–D). The lesion was originally

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<https://doi.org/10.1016/j.ajoc.2021.101020>

Received 24 July 2020; Received in revised form 9 November 2020; Accepted 14 January 2021

Available online 21 January 2021

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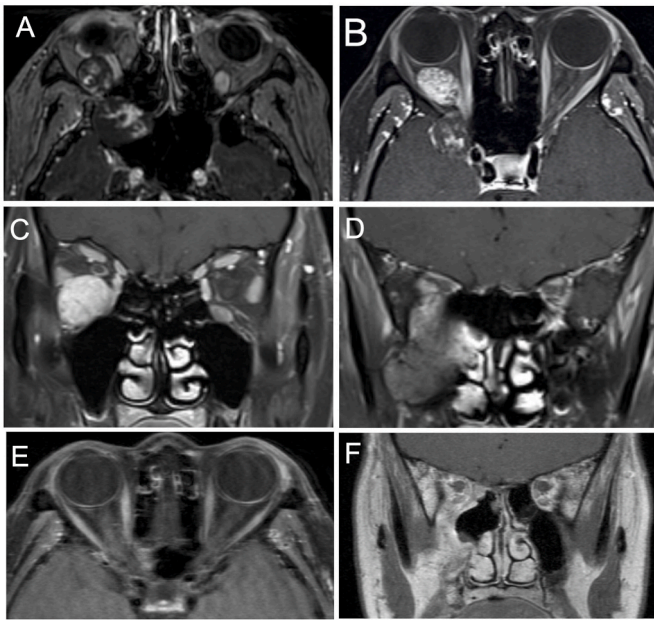


Fig. 1. (A–F): Magnetic resonance image (MRI) of orbits showing a multi-compartment mass involving the right orbit, pterygopalatine fossa, ethmoid and sphenoid sinuses and right middle cranial fossa. Axial T1 sequences with fat suppression post-gadolinium: (A) Demonstrates a bilobed right intraconal heterogeneously enhancing mass adjacent to the optic nerve (B) Extending along the middle cranial fossa to the anterior cavernous sinus. Coronal T1 sequences with fat suppression post-gadolinium: (C) Demonstrates a well-circumscribed heterogeneously enhancing intraconal mass (D) Extending through the infraorbital fissure to the pterygopalatine and middle cranial fossa. Post-surgical axial and coronal T1 sequences with fat suppression post-gadolinium of orbits: (E–F) Demonstrating gross total resection and post-surgical change.

presumed to be a schwannoma, radiographically, due to its involvement of the maxillary division of the trigeminal nerve, extension through the inferior orbital fissure and heterogenous enhancement pattern. However, a multi-compartment CVM was also considered in the differential due to the avid enhancement of the mass within the orbit. Multidisciplinary endoscopic endonasal and transorbital skull base resection was performed due to the multi-compartment nature of the mass.

The oculofacial plastic surgery team performed a transconjunctival orbitotomy to excise the orbital portion of the mass. A vascular encapsulated mass was dissected from the lateral, medial and inferior rectus muscles as well as the optic nerve superiorly. The intraconal mass (Fig. 2A) extended to the infratemporal orbital apex and was transected in the infraorbital fissure as it entered the pterygopalatine fossa. The bone along the infraorbital fissure and posterior wall of the maxillary sinus was removed to dissect the mass along the path of the zygomatic nerve to the maxillary nerve in the pterygopalatine fossa. Subsequently, an extended endonasal transpterygoid approach, through the right nostril, was performed by the rhinology team to continue the pterygopalatine dissection. The oculoplastics team, via the transconjunctival approach, worked simultaneously with the neurosurgical team, via the endonasal approach, to dissect the superolateral aspect of the pterygopalatine fossa mass. The mass was notably adherent to the maxillary nerve and dissected free by placing anterior traction on the infraorbital nerve while posteriorly dissecting the mass from the proximal maxillary nerve in the pterygopalatine fossa (Fig. 2B). The neurosurgery team dissected the adherent mass medially from the descending palatine nerve and posteriorly from the Vidian nerve. The pterygoid body above the Vidian canal was drilled to open the lateral recess of the sphenoid sinus and expose the maxillary nerve in the middle cranial fossa, the pituitary gland prominence, the optic nerve prominence at the paraclinoidal space, as well as the superior orbital fissure and orbital apex.

The greater wing of the sphenoid bone of the middle cranial fossa was drilled away leaving behind the maxillary strut and providing the necessary access to bluntly dissect the lesion away from the dura of the middle cranial fossa. The pterygopalatine fossa and middle cranial fossa portions of the lesion were excised in one piece through the nose and was pathologically consistent with a CVM (Fig. 2C–D). The patient recovered well with 20/20 vision, full extraocular movements, and resolution of proptosis. Post-operative orbital MRI showed gross total resection of the orbital and skull base mass (Fig. 1E–F).

3. Discussion

We report a rare presentation of a bilobed complex CVM extending between the cavernous sinus, pterygopalatine fossa and intraconal orbit. Due to extension along the maxillary division of the trigeminal nerve, the lesion was initially thought to be a schwannoma. Trigeminal schwannomas arise from Schwann cells that surround the cranial nerves, are the second most common intracranial mass and can extend into the orbit. Whereas, orbital CVMs tend to be well encapsulated and do not typically follow the course of cranial nerves or extend outside the orbit. A prior study described five cases of intracranial CVMs that followed the path of the trigeminal nerve.⁵ However, this is the first report to describe a CVM that extended between the intraconal orbital space, the pterygopalatine fossa and the middle cranial fossa along the trigeminal nerve.

Indications for surgery included new onset proptosis and eyelid swelling due to a large skull base mass abutting the optic nerve and cavernous sinus. Observation was an option but deferred by the patient due to proptosis and the risk of vision loss and headaches with mass progression. The risks of surgical excision included scarring, loss of vision, bleeding from the internal maxillary artery, cerebrospinal fluid leaks and meningitis associated with dural involvement in the middle cranial fossa. Given the complex presentation, the mass was excised via a multi-disciplinary, minimally invasive approach with ophthalmology, otolaryngology and neurosurgery. Advantages of the combined transorbital and endoscopic endonasal approach in this case are threefold. First, the transconjunctival approach avoids a noticeable incision and allows for direct access to the intraconal space for careful dissection of the mass from the optic nerve and delicate orbital apex structures. Second, the orbital exposure of the pterygopalatine fossa allowed for multi-portal and multi-angled access to the mass and obviated the need for a *trans*-septal approach, which would have required a posterior septectomy to facilitate additional angulation via the contralateral nostril. Instead, an ipsilateral extended endonasal transpterygoid approach was sufficient to access the mass, leaving the septum intact. While commonly performed for endoscopic skull base surgery, a posterior septectomy can increase the risk of epistaxis or injury to olfactory neuroepithelium and can prolong healing time relative to cases where the septum is preserved.⁶ Third, the transconjunctival approach provides excellent exposure to the lateral orbit and lateral infraorbital fissure which are very familiar territories for orbital surgeons. In contrast, traversing the lateral orbit endoscopically may result in unintended manipulation or injury to the optic nerve or inferior division of the oculomotor nerve, resulting in blindness or diplopia.⁷ If needed, the transorbital transconjunctival approach can be extended laterally and medially to access masses that extend to the temporal lobe, infratemporal fossa, lateral middle cranial fossa, cribriform plate, ethmoid and sphenoid sinuses, nasal cavity, medial orbit and central skull base.^{8,9} Combination endoscopic and transorbital skull base approaches have gained popularity in the last few years and have the advantage of increased working angles, improved ease of access, avoidance of crossing critical neurovascular structures and similar outcomes than more traditional open transcranial approaches.¹⁰ This case supports the utility of multi-disciplinary skull base surgery to successfully resect a multicompartiment skull base mass, particularly for tumors extending between the orbit and middle cranial fossa.

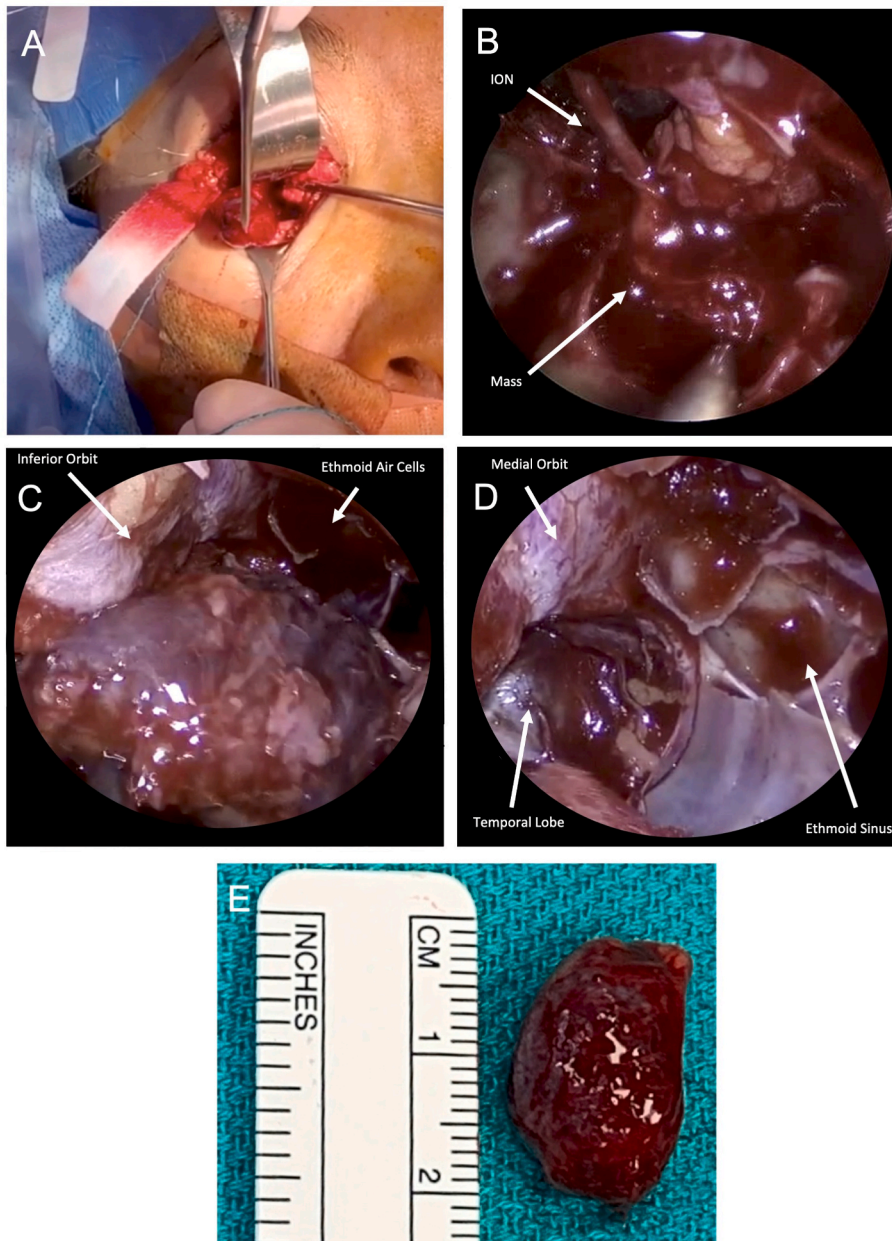


Fig. 2. (A–E): Intraoperative photos. (A) Resection of the orbital lobe of the mass via an inferior trans-conjunctival orbitotomy incision. (B) Transnasal endoscopic dissection of the mass in the pterygopalatine fossa (PPF). Here, the mass is wrapped around and adherent to the maxillary nerve in the PPF (large arrow), the maxillary nerve continues to travel within the inferior orbital floor (surgically removed in this photo) as the infraorbital nerve (ION, small arrow). (C) Resection of the mass from pterygopalatine fossa and middle cranial fossa via an extended endonasal transpterygoid endoscopic approach, the inferior orbit (large arrow) is noted above the mass and the ethmoid air cells (small arrow) are visible posterior and medial to the mass. (D) Middle cranial fossa after mass removal with adjacent medial lobe, temporal lobe, and ethmoid sinus visible. (E) Gross specimen of orbital portion of the bilobed mass.

4. Conclusions

Multicompartment cavernous venous malformations that involve the orbit and nerves of the skull base are rare. This case demonstrates that CVMs can extend through skull base fissures and foramina along cranial nerves. This case adds to the literature of complex CVMs and the utility of the transorbital approach to offer excellent orbital and skull base exposure that can be combined with endoscopic endonasal surgery to access the perinasal sinuses and middle cranial fossa for complex skull base masses.

Funding

None.

Author's contributions

ALK and PH significantly contributed to the care of the patient and

the surgical approach. ADA and CS contributed to the writing of the case report. All authors offered significant edits to the final manuscript.

Declaration of competing interest

None.

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