Orbital Cavernous Hemangioma with Atypical Symptoms and in an Atypical Location in a Young Adult

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Disclosures

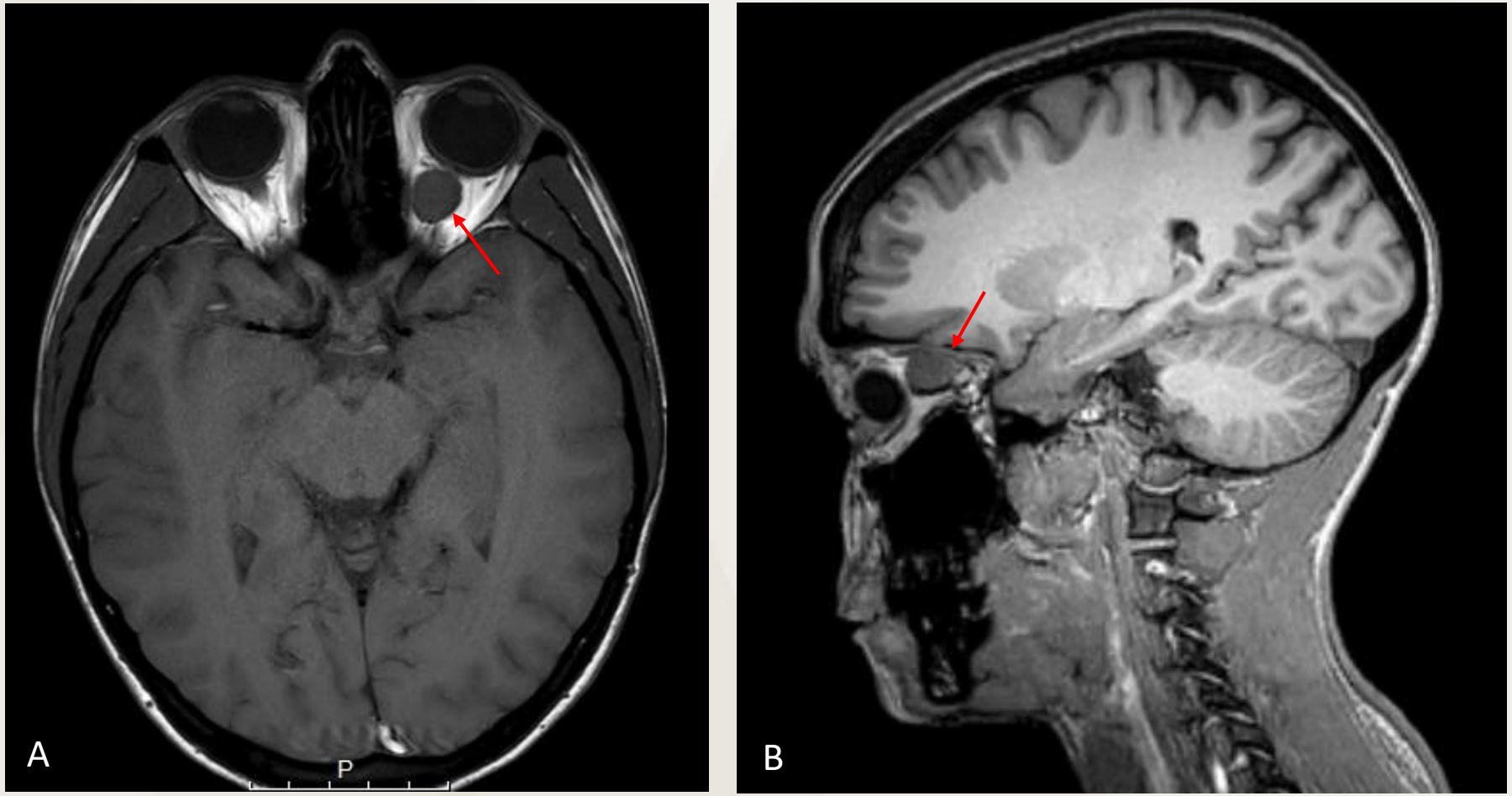
- None

Clinical Presentation

- A 30-year-old female with no significant past medical history presented with months of progressive blurry vision and periocular discomfort in the left eye. She also reported a feeling of “weakness” in the left eye, peripheral vision darkening, a gray spot in her central vision, ocular swelling, intermittent retro-orbital pain radiating to the ear, tinnitus, and photophobia.
- Examination revealed visual acuity of 20/15 in both eyes, a trace left relative afferent pupillary defect (left eye dilation of 0.5mm on swinging light test), and 2 mm of proptosis of the left eye. Intraocular pressures were 12 mmHg bilaterally. No periorbital swelling or erythema was noted.

Imaging Discussion

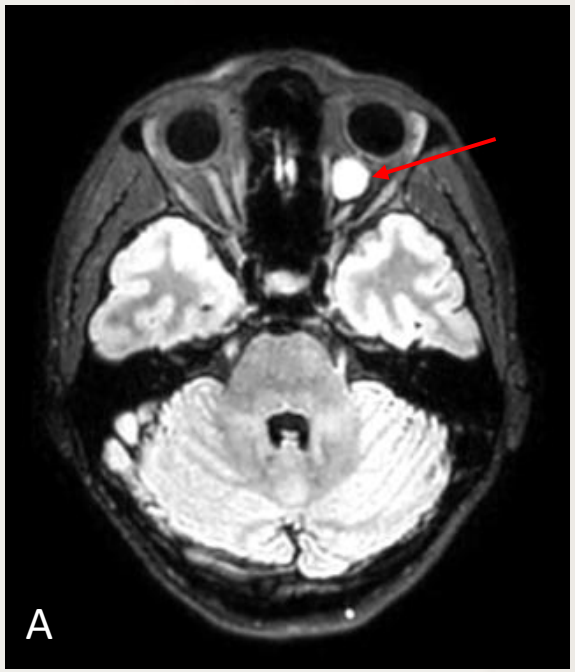
- MRI Orbits with and without IV contrast was performed, which demonstrated a well-circumscribed ovoid mass 16 x 14 x 12 mm in the left orbit
- The mass is hypointense on T1-weighted imaging

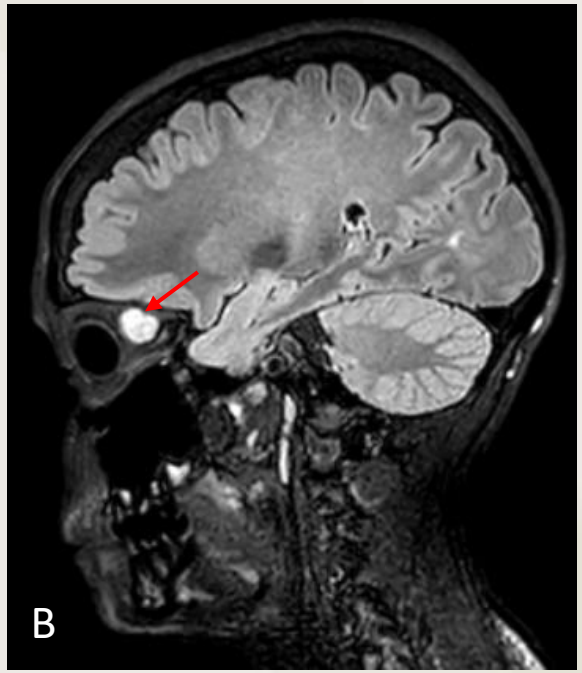


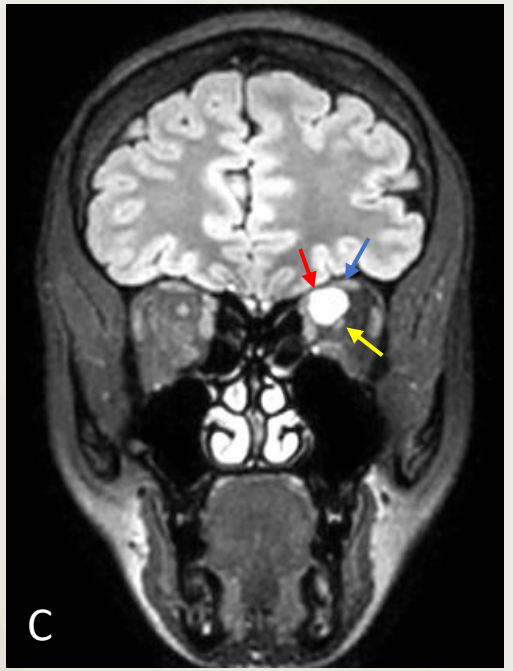
MRI Orbits with and without IV contrast demonstrating a well-circumscribed ovoid mass (arrows) in the left orbit. The mass is hypointense on T1-weighted imaging in the (A) axial and (B) sagittal planes.

Imaging Discussion

- The lesion is hyperintense on T2-weighted imaging
- Coronal views demonstrated that the mass occupies the superomedial intraconal space. There is local mass effect with leftward bowing of the optic nerve and slight lateral bowing of the superior rectus muscle, without encasement of either structure.







The mass (red arrow) is hyperintense on T2 FLAIR sequences, as seen in the (A) axial, (B) sagittal, and (C) coronal planes. Coronal images also demonstrate how the mass occupies the superomedial intraconal component and exerts mass effect on the optic nerve (yellow arrow) and superior rectus muscle (blue arrow).

Imaging Discussion

- Initial T1-weighted post-contrast imaging demonstrates focal, heterogenous nodular enhancement with adjacent tortuous vessels within the anterior portion of the lesion. There is then progressive fill-in on more delayed sequences, culminating in more uniform, homogenous enhancement on the final T1 post-contrast sequence.

(A) Initial heterogenous post-contrast enhancement of the mass on T1-weighted axial imaging, followed by progressive fill-in and homogenous uptake of contrast on final (B) axial and (C) sagittal T1 post-contrast images.

Imaging Discussion

- The combination of a well-circumscribed intraconal lesion that is T1 hypointense, T2 hyperintense, and demonstrates initial heterogenous contrast filling that progressively becomes more homogenous over time is classic for a benign, low-flow venous malformation: in this case, an **orbital cavernous hemangioma** [1, 2]
  - Lack of prominent arterial supply to the mass explains its slow filling with contrast
  - These imaging features distinguish orbital cavernous hemangiomas from other intra-ocular tumors and/or vascular lesions, which are often more irregular in appearance [3]
  - Orbital hemangiomas are the most common orbital mass in adults, with higher female prevalence
- However, the medial intraconal location of this lesion is relatively atypical, as most orbital cavernous hemangiomas present in the lateral intraconal space [3]
- Findings on imaging explain many aspects of this patient's clinical presentation, including her proptosis and ocular pain; blurry vision, reduced visual acuity, and relative afferent pupillary defect (from optic nerve compression); and ocular weakness (from superior rectus muscle compression)
- Other presenting symptoms, including her retro-orbital pain radiating to the ear and tinnitus, are fairly atypical for orbital cavernous hemangiomas.

Management

- Given the patient's preserved visual function and the benign nature of orbital cavernous hemangiomas, observation was recommended, with follow-up MRI scheduled for 4 months after initial presentation
- Repeat imaging demonstrated stability of the lesion, further confirming the diagnosis of an orbital cavernous hemangioma
- At this time, the patient was informed that she could choose between surgical removal or continued observation of the lesion. Given her ongoing symptoms, she opted for surgical management
  - Surgery is usually recommended for interval enlargement in lesion size, functional compromise, or cosmetic concerns (e.g. significant proptosis)
- The patient underwent left orbitotomy with mass removal 6 months after her initial presentation

Outcome

- After surgery, the patient's left eye pressure and pain improved
- However, two weeks post-op, she re-presented to the ED with left periorbital edema, erythema, and purulent discharge from her skin incision
- CT of the orbits with contrast demonstrated a large left intraconal abscess measuring 23 x 21 x 12 mm abutting the optic nerve, superior rectus muscle, superior oblique muscle, and medial rectus muscle
- The patient is now s/p abscess drainage, and is currently completing a course of antibiotics and clinically improving

CT of the orbits with contrast demonstrated a large left intraconal abscess measuring 23 x 21 x 12 mm, near the location of the patient's resected orbital cavernous hemangioma.

Take Home Points

- Orbital cavernous hemangiomas are benign, slow-flow venous malformations and represent the most common orbital mass in adults. They have a predilection for females in their 3rd-5th decade of life
- The typical presentation is painless, slowly progressive proptosis; acute pain, tinnitus, and photophobia seen in this patient are atypical and may prompt urgent evaluation
- MRI is the preferred imaging modality for diagnosis, and demonstrates a well-circumscribed lesion that is hypointense on T1, hyperintense on T2, and that demonstrates initially heterogeneous yet progressive contrast fill-in due to lack of prominent arterial supply
- 80% of lesions are intraconal. Superomedial mass location as seen in this patient is less common than the typical lateral intraconal position, and may influence surgical approach

Thank You!

Questions?

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