Published in final edited form as:

JAMA Ophthalmol. 2016 July 01; 134(7): 837–838. doi:10.1001/jamaophthalmol.2015.6106.

Proptosis in a Healthy Young Woman

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A girl in her midteens with an unremarkable medical history was referred for evaluation of proptosis. She denied pain but described a pressure sensation behind her left eye. Uncorrected visual acuity measured 20/80 OD and 20/60 OS, improving to 20/40 OD and 20/30 OS with pinhole testing. Pupils were round and reactive without an afferent pupillary defect. There was moderate restriction to elevation of the left eye. Intraocular pressure was 18 mm Hg OU by applanation tonometry. Hertel exophthalmometer measurements demonstrated 7 mm of proptosis OS. Anterior segment and ophthalmoscopic examination results were unremarkable.

Computed tomography of the orbits with and without contrast was performed (Figure). A homogeneously enhancing, well-demarcated, ovoid mass was visualized within the medial intraconal space, measuring $2.0 \times 2.2 \times 2.6$ cm. The lesion displaced the globe inferiorly and anteriorly. Smooth bony remodeling was present without erosive changes. Magnetic resonance imaging of the orbits corroborated these findings. Conventional angiography demonstrated a tortuous branch of the left ophthalmic artery supplying the medial aspect of the orbital mass.

Diagnosis

Intraorbital hemangiopericytoma

What to Do Next

C. Embolization followed by orbitotomy and biopsy

Discussion

The differential diagnosis of orbital tumors includes vascular disorders (cavernous hemangioma, hemangiopericytoma, venous vascular malformation), lacrimal gland tumors

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Conflict of Interest Disclosures: All authors have completed and submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest and none were reported.

Role of the Funder/Sponsor: The funders had no role in the design and conduct of the study; collection, management, analysis, and interpretation of the data; preparation, review, or approval of the manuscript; and decision to submit the manuscript for publication.

Additional Contributions: Amy Lin, MD, Department of Ophthalmology and Visual Sciences, Illinois Eye and Ear Infirmary, University of Illinois at Chicago, assisted with histopathologic images; she received no compensation.

(pleomorphic adenoma, adenoid cystic carcinoma, lacrimal gland lymphoma), optic nerve lesions (optic nerve glioma, optic nerve sheath meningioma, schwannoma, neurofibroma), infectious etiologies (orbital cellulitis), and occult neoplasms (metastasis, rhabdomyosarcoma, lymphoproliferative disorder). Thyroid eye disease and idiopathic orbital inflammation should also be considered, especially in young patients. Fine-needle aspiration biopsy is unnecessary if surgery is scheduled. Fluorescein angiography provides no information and observation is inappropriate for tumors with malignant potential.

Vascular imaging in this patient revealed a high-flow feeder vessel to the orbital mass. The neurosurgery service was consulted and diagnostic cerebral angiography followed by glue embolization was performed. The following day, the patient underwent uncomplicated orbitotomy and excision of the orbital mass. Based on histopathologic studies, the patient was diagnosed as having a hemangiopericytoma.

Hemangiopericytomas are uncommon mesenchymal neoplasias that account for fewer than 1% of all orbital tumors.¹⁻³ Patients are usually diagnosed in middle age (fourth or fifth decades) without sex or racial predilection.^{3,4} Hemangiopericytomas are slow-growing lesions with variable malignant potential and may be benign or capable of metastasis. Common clinical manifestations occur from the mass effect on adjacent orbital structures and include proptosis, strabismus, and restriction of extraocular motility.^{4,5} Pain, afferent pupillary defects, and visual acuity changes are less common.

Imaging studies may reveal a solid, well-circumscribed, lobulated mass with discrete borders.⁵⁻⁷ The lesions are typically isointense relative to gray matter on T1-weighted images and may show equal or lower signal intensity on T2-weighted images. Bony erosion and calcification are uncommon except in very aggressive lesions. Accurate diagnosis requires histopathologic identification.

Hemangiopericytomas are histologically characterized by the proliferation of pericytes of Zimmerman, contractile support cells that surround small vessels.^{2,4} Histopathologic sections reveal a dense, vascular, hypercellular tumor with spindle-shaped cells and a vascular branching pattern classically described as “staghorn vessels.” Prognostic factors for recurrence and survival include tumor size larger than 5 cm, metastatic disease at presentation, infiltrative margins, necrosis, high cellularity, nuclear pleomorphism, and a mitotic index with more than 4 mitoses per 10 high-power fields.⁸ Staining with CD34 and CD31 also helps to characterize the lesion.

Given the potential for hemangiopericytomas to recur and potentially metastasize, the preferred initial treatment is complete excision of the tumor.^{5,9} Hemangiopericytomas are resistant to radiotherapy and this treatment is reserved for metastatic disease, recurrence of the primary tumor, or incompletely excised lesions.¹⁰ Chemotherapy may have a role in the treatment of distant metastatic disease but is not used as monotherapy.

Patient Outcome

This patient was diagnosed as having an intraorbital hemangiopericytoma. She underwent uncomplicated cerebral angiography with glue embolization followed by orbitotomy and

excision of the mass. The pathologic margins were negative, and the pseudocapsule remained intact. Evaluation by the hematology-oncology and radiation oncology services was pursued and no adjuvant chemotherapy or radiotherapy was offered. Computed tomography of the orbits 1 month later revealed no evidence of residual tumor or recurrence.

Acknowledgments

Funding/Support: This work was supported by an unrestricted grant from Research to Prevent Blindness and core grant EY01792 from the National Eye Institute. Dr Aakalu was supported by a research grant from Midwest Eye Banks, a grant-in-aid from Fight for Sight, and grant K08 EY024339 from the National Eye Institute.

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WHAT WOULD YOU DO NEXT?

- A.** Fine-needle aspiration biopsy
- B.** Fluorescein angiography
- C.** Embolization followed by orbitotomy and biopsy
- D.** Observation

Figure.

Computed tomography and conventional angiography of the orbital mass. A, Computed tomography of the orbits demonstrates a homogeneously enhancing mass within the intraconal space (blue arrowhead). Smooth bony remodeling was present without erosive changes. B, Cerebral angiography demonstrated a branch of the left ophthalmic artery supplying the orbital mass (black arrowhead).