

Follicular lymphoma isolated to the superior oblique muscle

A 76-year-old female presented with bilateral peripheral visual field constriction. She had a past medical history of left common iliac vein stenting secondary to compression from the iliac artery, left lower extremity deep vein thrombosis requiring angioplasty, bilateral lower extremity lipodermatosclerosis, hyperlipidemia, vitamin D deficiency, diverticulitis, and generalized anxiety disorder. Her regular medications included apixaban, clopidogrel, venlafaxine, and montelukast. She denied smoking or alcohol use.

She presented as a telemedicine patient during the COVID-19 pandemic with a 1-year history of progressive, painless bilateral peripheral vision loss. After initial development of bilateral peripheral vision loss, she underwent bilateral cataract extraction with intraocular lens placement and subsequent yttrium aluminum garnet (YAG) laser capsulotomy of the right eye with progressive vision loss thereafter. Composite findings from external and subsequent examinations revealed that her visual acuity was 20/40 OD and 20/30 OS. Slit-lamp biomicroscopy and intraocular pressures were normal. Hertel exophthalmometry revealed proptosis—18 mm OD and 15 mm OS. Extraocular movements were normal. Her confrontation visual fields were equally narrowed at 1 m and 2 m testing (“tunnel vision”), and her saccades were accurate outside of a 5 degree island bilaterally. Fundus examination showed a cup-to-disk ratio of 0.3 OU with mild optic disc pallor and peripapillary atrophy bilaterally. Global optical coherence tomography showed retinal nerve fibre layer thickness of 81 μm OD and

85 μm OS. Automated perimetry (Humphrey visual field 24-2) was constricted to a 5 degree central island bilaterally with a mean deviation of -28.09 dB OD and -28.49 dB OS.

Magnetic resonance imaging of the brain and orbits showed asymmetric enlargement of the right superior oblique muscle, enhancement in the adjacent intraconal and extraconal fat, and enhancement of the floor of the adjacent anterior cranial fossa (Fig. 1). Right orbital biopsy and right extraocular muscle biopsy were performed via endoscopic orbitotomy and ethmoidectomy. The biopsy showed an infiltrate of predominantly small lymphocytes found to be positive for CD20 with coexpression of CD10 and BCL2 and moderate intermixed CD3 T cells. Immunohistochemistry was negative for CD5, cyclin D1, Epstein–Barr virus by in situ hybridization, c-MYC, MUM1, and kappa and lambda light chains. These findings were consistent with low-grade follicular lymphoma. Further malignancy staging revealed a negative bone marrow biopsy, and positron-emission tomography revealed fluorodeoxyglucose avidity in the right superior oblique muscle and a right inguinal lymph node (this node was negative for lymphoma on biopsy). Paraneoplastic antibodies were negative.

The patient received low-dose radiation therapy (400 cGy, 2 fractions) to the right orbit. Repeat magnetic resonance imaging after 3 months showed resolution of the orbital mass. Positron-emission tomography scan showed no avidity in the right orbit or inguinal lymph node, consistent with clinical remission. At her subsequent neuro-ophthalmic review, the patient remained clinically stable with stable constricted Humphrey visual fields that were most

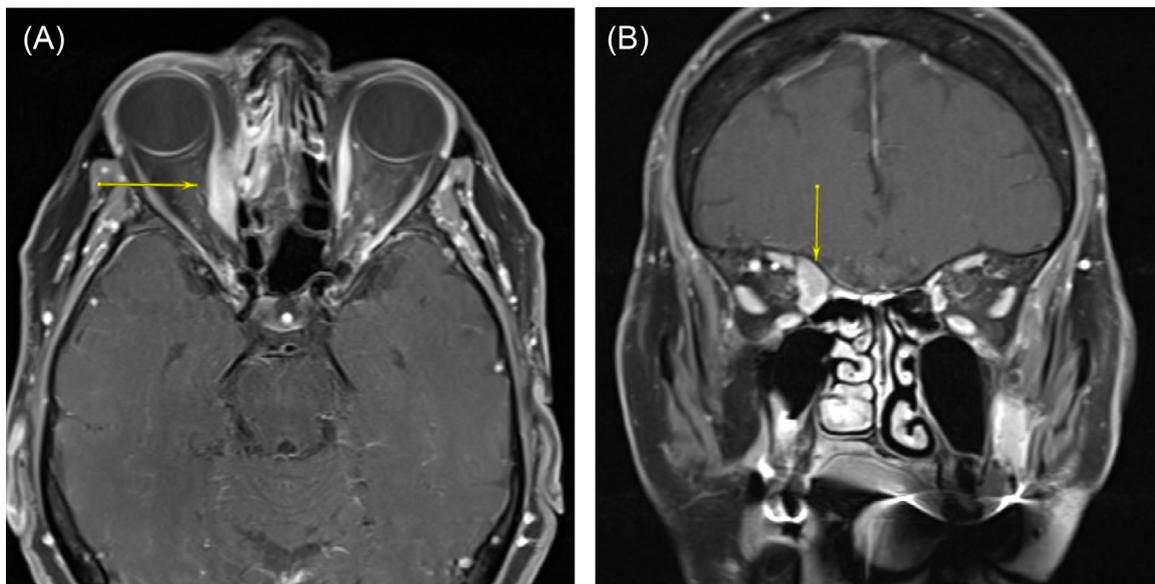


Fig. 1 – Magnetic resonance imaging of the brain and orbits. T1 fat-saturated post-gadolinium contrast sequences showing asymmetric enlargement of the right superior oblique muscle with enhancement of soft tissue in the adjacent intraconal and extraconal fat in (A) axial sequence and (B) coronal sequence.

consistent with a functional vision disorder and stable mild global optic atrophy with an optical coherence tomography retinal nerve fibre layer thickness of 79 μm OD and 80 μm OS.

Orbital lymphoma is a rare site of lymphoma, accounting for 1% of non-Hodgkin lymphoma cases.¹ Extraocular muscle orbital lymphoma accounts for <2% of orbital lymphomas.² Involvement of the superior oblique muscle is even rarer. Eade et al.³ described 25 patients with lymphoma of the extraocular muscles and found that lymphoma was most common in the levator palpebrae superioris, superior rectus, and inferior rectus. Only 4 patients (16%) had superior oblique muscle involvement.³ Watkins et al.⁴ described 57 patients with lymphoma of the extraocular muscles in which the tumour occurred in the rectus muscle (73%), the oblique muscles (17%), and the levator muscle (11%).

Lymphoma of the extraocular muscles is most commonly treated with radiotherapy or chemotherapy. In their series, Eade et al.³ noted that 24 of the 25 patients had a complete response to initial treatment, with 1 case of relapse. Watkins et al.⁴ showed that patients had good outcomes following treatment, with overall mortality of 15.8%. In summary, clinicians should be aware that while rare, isolated superior oblique muscle enlargement can be indicative of lymphoma.

Meher A. Saleem,* Subahari Raviskanthan,[†] Anirudh Mukhopadhyay,[‡] Amina I. Malik,[§] Andrew G. Lee^{†,||,¶,*,††,‡‡}

*Georgetown University School of Medicine, Washington, DC; [†]Blanton Eye Institute, Houston Methodist Hospital, Houston, Tex.; [‡]Baylor College of Medicine, Houston, Tex.; [§]Houston

Methodist Hospital, Houston, Tex.; ^{||}Weill Cornell Medicine, New York, NY; [¶]University of Texas Medical Branch, Galveston, Tex.; ^{**}University of Texas MD Anderson Cancer Center, Houston, Tex.; ^{††}Texas A&M College of Medicine, Bryan, Tex.; ^{‡‡}University of Iowa Hospitals and Clinics, Iowa City, Iowa.

Originally received May. 10, 2022. Accepted Jun. 8, 2022.

Correspondence to Andrew G. Lee, MD; aglee@houstonmethodist.org.

References

1. Ahmed S, Shahid RK, Sison CP, Fuchs A, Mehrotra B. Orbital lymphomas: a clinicopathologic study of a rare disease. *Am J Med Sci* 2006;331:79–83.
2. Sullivan TJ, Whitehead K, Williamson R, et al. Lymphoproliferative disease of the ocular adnexa: a clinical and pathologic study with statistical analysis of 69 patients. *Ophthalmic Plast Reconstr Surg* 2005;21:177–88.
3. Eade EL, Hardy TG, McKelvie PA, McNab AA. Clinicopathological features and outcomes in lymphoma of extraocular muscles. *Ophthalmic Plast Reconstr Surg* 2019;35:615–8.
4. Watkins LM, Carter KD, Nerad JA. Ocular adnexal lymphoma of the extraocular muscles: case series from the University of Iowa and review of the literature. *Ophthalmic Plast Reconstr Surg* 2011;27:471–6.

Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this correspondence.