



A Diagnostic Dilemma: Optic Nerve Sheath Meningioma Mimicking Perineuritis

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Introduction

Optic nerve sheath meningiomas (ONSM) are typically found in the subarachnoid or intradural space of the intraorbital optic nerve, and present clinically with decreased visual acuity, afferent pupillary defect, optic disc edema, and visual field loss, with no pain^{1,2}. They have a characteristic tram-track enhancement on MRI and are treated in non-diabetic patients with fractionated radiation therapy³. Optic perineuritis shares many clinical and radiological features as ONSM but is treated with high dose systemic corticosteroids instead.

Case Presentation

- 73-year-old female with no past ocular history presenting with six months progressive visual changes in left eye, no inciting events
- Transient episodes with positional changes of “snowy” area in superonasal quadrant of OS, increasing in frequency over time
- Exam significant for normal VA, choroidal folds with staining of vessel walls, mild RAPD OS, enlarged blind spot, blurred and elevated optic disc with diffuse edema
- CT and MRI showed thickening and tram-track enhancement of distal left optic nerve sheath; referred to oculoplastics and neuro-ophtho
- differential diagnosis of optic perineuritis vs ONSM
- Biopsy of optic nerve sheath was performed
- started on fractionated radiation therapy and prednisone course
- Five months after completing fractionated radiation therapy, patient had stable optic neuropathy with resolution of disc edema OS

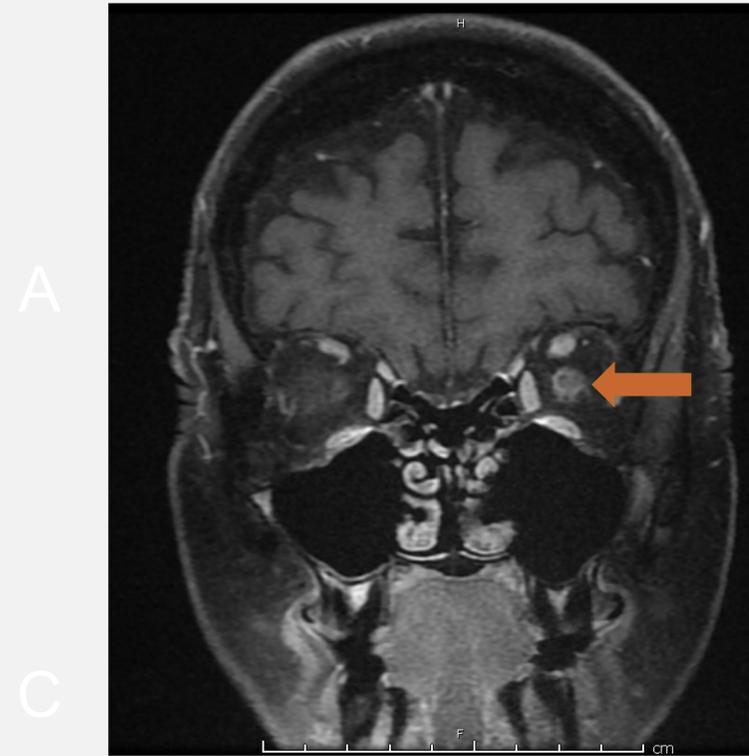


Figure 1. Postcontrast T1 coronal fat-suppressed MRI shows enhancement and thickening of the left optic nerve sheath

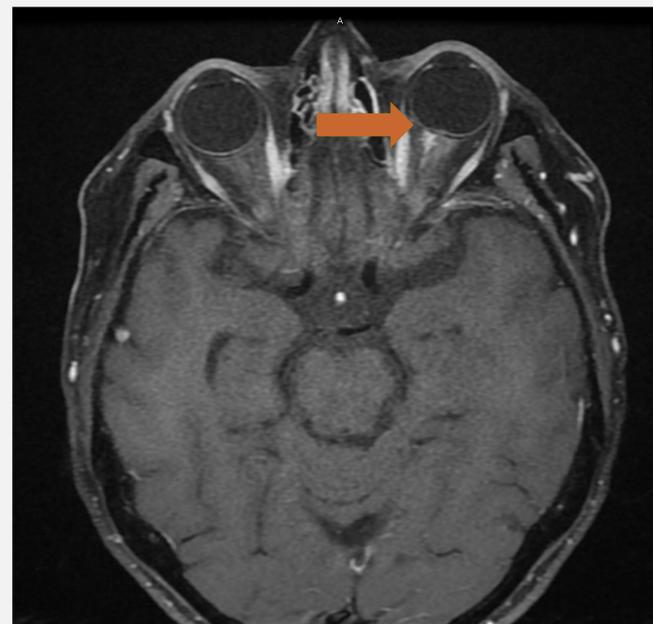


Figure 2. Postcontrast T1 axial fat-suppressed MRI demonstrates enhancement of the proximal left optic nerve sheath

Discussion

Optic perineuritis, either primary or secondary to inflammatory or neoplastic conditions, can mimic ONSM making the diagnosis difficult. Imaging with classic “tram-track” signs are nonspecific, and there are no distinguishing clinical findings.

In our case, enhancement around the optic nerve happened distally, closer to the orbit, versus a more likely location of ONSM in the optic canal. This finding made perineuritis a more likely diagnosis. A biopsy was tried first, revealing the correct diagnosis early and allowing appropriate treatment to be instituted.

To differentiate the two conditions, the typical course of treatment would have been a trial of steroids, followed by nerve biopsy if no improvement. Medical management with an initial empiric course of corticosteroids could avoid a surgical biopsy and the risks of the procedure. However, the correct dose and length of steroid therapy has not been established. Furthermore, an empiric course of corticosteroids may decrease the diagnostic yield of a subsequent biopsy, such as in cases of infiltration by lymphoma.

Conclusion

Distinguishing ONSM and perineuritis based on clinical presentation and imaging alone may be challenging. Although an empiric course of prednisone may help distinguish the two, this case illustrates how a biopsy early on may expedite reaching the correct diagnosis and treatment.

References

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3. Turbin, Roger E., et al. "A long-term visual outcome comparison in patients with optic nerve sheath meningioma managed with observation, surgery, radiotherapy, or surgery and radiotherapy." *Ophthalmology* 109.5 (2002): 890-899.