

Case Report

An unusual radiological presentation of optic nerve sheath meningioma



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Abstract

Our report describes an unusual radiological presentation of optic nerve sheath meningioma. The classic radiological appearance of optic nerve thickening with enhancement and calcification within the tumor was not seen; instead, an elongating gadolinium enhancing band-like area adjacent to the superomedial aspect of the left optic nerve sheath was identified. The diagnosis was confirmed on histopathology. Our report adds to the spectrum of presentations of this relatively common clinical entity.

Keywords: Optic nerve sheath meningioma, Radiology, MRI, Histopathology

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Introduction

Optic nerve sheath meningiomas (ONSM) account for 2% of all orbital tumors.¹ They are the second most common optic nerve tumor after gliomas and represent 1–2% of all meningiomas.^{2,3} Without treatment, continued growth of ONSM results in progressive loss of visual acuity, color blindness and eventually complete loss of vision.¹ No tumor-related deaths have been reported.³ Radiological findings are usually of an enlarged optic canal, optic nerve thickening and enhancement and calcification within the tumor.² Definitive treatment of ONSM is controversial. Fractionated radiotherapy is often first choice in an attempt to control the ONSM growth rate and to preserve vision if the diagnosis is not in question.³ Surgery remains best-treatment in cases with blindness, severe proptosis and in patients with intracranial extension.³

Case report

A 57-year-old female doctor presented with a 1-year history of blurred vision and visual field changes in her left eye. She had a history of a pituitary adenoma 30 years previously, treated by open craniotomy and post-operative radiotherapy (4200 rad over 20 treatments). She was otherwise well.

Examination revealed visual acuity of 20/17 on the right and 20/120 on the left. There was left relative afferent pupillary defect and left sided brightness desaturation. No proptosis was noted and the patient had a full range of ocular movements. Anterior segment examination was unremarkable, while fundal examination showed disk pallor of the left optic nerve head without disk swelling. There was progression of inferior visual field loss (Humphrey Field Analyzer; Zeiss, Germany) between January 2010 and February 2011 as shown in Fig. 1.

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cavernous fistula or aneurysms. The differential diagnosis was an ONSM, a diffuse glioma of the optic nerve, or less likely, an atypical recurrence of pituitary adenoma.

With her previous history of pituitary adenoma with radiation treatment, progressing symptoms and the uncertainty of the diagnosis, the patient expressed a strong preference for surgical exploration and removal of the lesion with division of the optic nerve rather than further radiation therapy. She was aware that surgical exploration would leave her with no light perception vision post-operatively.

A stereotactically guided resection of tumor was performed through an orbitozygomatic craniotomy. The left frontal lobe was gently retracted to expose the superior orbit. The roof of the orbit was decompressed to 6 mm anterior to the optic canal. After opening the periorbita and exposing the muscle cone, the supraorbital nerve was mobilized laterally. The optic nerve was localized behind the globe and the optic nerve was full inside the dura. A band-like structure was not identified; instead, a macroscopically normal-looking nerve was found surrounded by soft red/brown material. Resection of the optic nerve over a 16 mm distance was performed and sent for histopathology.

Histopathology (Fig. 3) showed fibrous thickening of the dura and arachnoid around the nerve with large collections of meningiothelial cells between the two layers with occasional psammoma bodies within the cellular collections. The meningiothelial cells show immunoperoxidase staining for progesterone receptors with a negligible proliferation rate with virtually no staining for Ki67. Taken altogether, this was consistent with an ONSM. No invasion of the optic nerve was noted. No subdural involvement of the meningioma was seen at either end of the nerve segment. Thus far, the patient has been followed up over a 12 month period and is recovering well.

Discussion

Our case demonstrated unusual radiological features of a histologically confirmed ONSM. The largest case series on record of ONSM describes five radiological patterns commonly encountered.² The commonest pattern identified was tubular (62% of cases), with subgroups of diffuse (45%), apical expansion (15%) and anterior expansion (3%), which had minimal widening of the optic nerve and more frequent

evidence of calcification.² The second group was globular tumors (23%) where growth was outside the dural sheath causing significant proptosis. 11% displayed a fusiform pattern and 4% had a focal enlargement of the optic nerve.² The radiological appearance of the ONSM in our case did not conform to any of the known patterns; rather, it appeared as a distinct band-like structure that displaced the optic nerve.

Previous ionizing radiation is a known cause of ONSM.^{4,5} Such secondary meningiomas tend to present at a younger age, present with multiple lesions commonly arising from the sphenoid ridge, tuberculum sellae or olfactory groove and demonstrate a higher recurrence rate.^{4,6,7} Histopathology also suggests that they tend to be more aggressive, with a higher tendency toward malignancy in patients who had high dose radiation therapy.^{4,8} This knowledge played a role in the decision made by our well-informed patient.

Canalicular ONSM can often pose a diagnostic challenge for clinicians as very small lesions can compress the optic nerve and cause significant and progressive visual symptoms because the bony canal walls limit expansion. Reports have shown diagnostic delays of greater than 1 year due to this variability in radiological appearance.⁴ It is strongly recommended that all cases of progressive visual loss be investigated by high-spatial-resolution MR of the orbit before and after contrast enhancement in combination with fat suppression.^{4,5}

Recent literature reviews support the safety of fractionated radiotherapy in the treatment of ONSM.^{2,3}

Patient consent

The patient has consented to the submission of the case report for submission to the journal.

Conflict of interests and source of funding

The authors declared that there is no conflict of interest or funding source for any authors.

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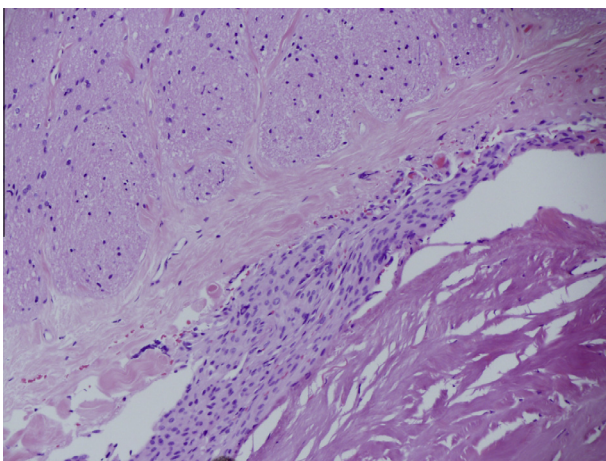


Figure 3. Histological appearance of the optic nerve sheath meningioma. The meningiothelial cells do not invade the pial/arachnoid tissue. (Hematoxylin and eosin stain.)