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Afterimage

Waiting to deliver a final diagnosis

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ABSTRACT

We report a patient with left eye positional transient visual obscurations, diplopia, a left afferent pupillary defect, and optic disk edema—all occurring during pregnancy. Non-contrast magnetic resonance imaging revealed an orbital mass encasing the optic nerve and extending through the superior orbital fissure into the cavernous sinus. An magnetic resonance imaging with gadolinium after parturition and strongly positive somatostatin receptor scintigraphy suggested the diagnosis of meningioma. Biopsy confirmed the diagnosis, and she was treated with CyberKnife stereotactic radiosurgery. In cases of atypical presentation, somatostatin receptor scintigraphy can help distinguish optic nerve sheath meningioma from alternative orbital masses.

1. Case presentation

A 33-year-old woman in week 26 of an uncomplicated pregnancy presented with diplopia in up-gaze and intermittent, 30-second episodes of painless "graying out" of vision in the left eye exacerbated by position changes. Acuity was 20/20 OD and 20/25 OS with intact color vision in both eyes and a left relative afferent pupillary defect. There was a moderate elevation deficit of the left eye. The remainder of the cranial nerve examination and anterior slit lamp examination was normal. External examination showed 4 mm of relative left proptosis. Dilated fundus examination was normal in the right eye and showed severe optic disk edema in the left eye. Humphrey Vision Analyzer 24-2 visual fields were full in the right eye and showed an inferior altitudinal defect in the left eye.

Magnetic resonance imaging (MRI) brain without gadolinium revealed a 20 × 17 × 14 mm intraconal left orbital mass that was T1- and T2-isointense, encased the optic nerve, and extended through the superior orbital fissure into the left cavernous sinus (Fig. 1). Despite a trial of oral prednisone 60 mg daily, her left eye's visual field worsened. The differential diagnosis at this point included steroid-refractory inflammation, infiltrative malignancy including lymphoma, and optic nerve sheath meningioma. The patient and care team opted for repeat imaging following Caesarean section at 37-week gestation. Postpartum MRI brain and orbits with gadolinium revealed no change in size of the homogeneously enhancing mass (Fig. 2). Orbital CT failed to reveal bone destruction, bone remodeling, or calcification.

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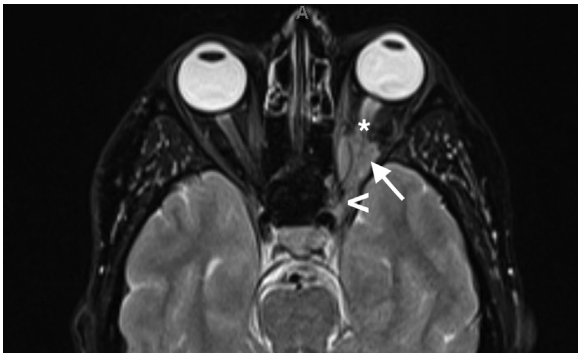

Fig. 1 – Axial T2 fat-saturated magnetic resonance imaging (MRI) without gadolinium revealed a left intraconal mass (*arrow*) encasing the optic nerve (*) and extending through the supraorbital fissure (*arrow head*) toward the left cavernous sinus.

Somatostatin receptor scintigraphy revealed focal radiotracer uptake of the mass indicating somatostatin receptor positivity and strongly suggesting the diagnosis of meningioma (Fig. 3). It is unusual for optic nerve sheath meningioma to track through the superior orbital fissure into the cavernous sinus, thus biopsy was recommended. Navigation-guided endoscopic sinus approach to the orbital apex allowed an incisional biopsy of the gelatinous mass lateral to the medial rectus muscle (Fig. 4).

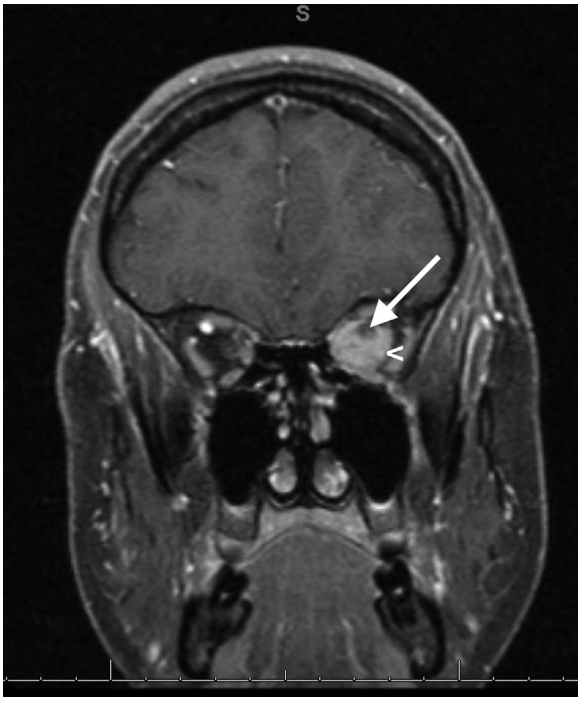

Fig. 2 – Coronal T1 fat-saturated postcontrast magnetic resonance imaging (MRI) through the midorbits demonstrating homogeneous enhancement of the mass (*arrow head*) encasing the left optic nerve (*arrow*).

Histopathology showed nests of bland meningothelial cells infiltrating skeletal muscle indicative of a WHO grade-1 meningioma (Fig. 5A). These cells were positive on immunohistochemical staining for progesterone receptors (Fig. 5B) and negative for estrogen receptors.

The mass was treated with CyberKnife stereotactic radiosurgery, administering 2500 cGy over 5 equal fractions. Four months after treatment, her visual acuity improved to 20/20, the visual field defect improved significantly, and her motility remained stable.

2. Discussion

Optic nerve sheath meningiomas (ONSMs) constitute 1–2% of all meningiomas, yet are the second most common intrinsic optic nerve neoplasm.[6] Like all meningiomas, ONSMs have a female predominance (F:M = 3:2).[6] Most ONSMs present among the middle-aged, although they may rarely present in childhood (4–7%).[1,6,19]

ONSM may be classified as primary, arising from the intraorbital or intracanalicular optic nerve, or secondary, arising from an intracranial location with extension to the optic nerve sheath.[17] Among ONSMs exhibiting both an intracranial and intraorbital segment such as our case, it may be challenging to discern the tumor's origin. Fortunately, this distinction of primary versus secondary ONSM carries little clinical significance in regards to management.

CT of the orbits can aid the diagnosis by showing thickening of the optic nerve/sheath complex, calcifications within the tumor, and hyperostotic bone remodeling. Intravenous contrast may help distinguish a hyperdense sheath and hypodense optic nerve termed the "tram-track sign." Readily available higher resolution MRI has greatly facilitated early detection of ONSMs. The "tram-track sign" is accentuated on T1-weighted postgadolinium sequences with avid enhancement of the thickened optic nerve sheath as compared to the optic nerve. The presence of tumor extension within the cavernous sinus, sellar region, and/or adjacent skull base as in our case is unusual in ONSM, but has been reported among 4/30 patients in 1 large series.[3] The extent of intracranial involvement is best assessed with MRI. In ONSMs with intracranial extension, the presence of an enhancing, thickened dural tail strongly indicates the diagnosis of meningioma with a specificity of 94% in 1 study.[18]

ONSM is now often diagnosed earlier than in the pre-MRI era.[13] In unusual cases such as ours, intracranial tumor and rapid symptom progression may lead to diagnostic challenges. Rapidly growing intracranial meningiomas indicate a more aggressive tumor grade, seen in 1.5–30% of tumors (WHO grade II or III) and are more common in men.[5] Among ONSMs specifically, an aggressive growth pattern is more common in children.[7] Rapid vision loss or acute realization of chronic vision loss in atypical ONSM presentations may lead to an initial misdiagnosis of optic neuritis.[2,21]

Sex-hormone receptor expression is common in intracranial meningiomas. In a study of 510 meningiomas, progesterone receptor, estrogen receptor, and androgen receptor positivity occurred in 88%, 40%, and 39% of tumors, respectively.[11] Clinically, meningiomas may exhibit