Optic nerve sheath meningioma- A diagnostic dilemma

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Patient: 41 years, female

Clinical History:

A 41-year-old lady presented with visual symptoms in her left eye associated with afferent papillary defect, visual field defects and disc swelling. Initial imaging and lumbar puncture suggested optic neuritis. However, progressive worsening of symptoms despite disease modifying treatment for Multiple sclerosis led to further investigations.

Imaging Findings:

A 41-year-old female healthy school teacher presented to the Eye clinic with a two month history of diplopia and flashing lights in her left eye. Visual acuity in her left eye was 6/9 and examination of her left eye revealed afferent papillary defect, constricted visual field defect with inferior and superior arcuate loss and swollen optic disc.

Initial investigations including CT scan, MRI and lumbar puncture were reported as normal. She was referred to a neurologist for her persistent symptoms. Multiple white matter lesions on repeat MRI scan and raised CSF IgG on lumbar puncture performed five months later suggested optic neuritis as the cause of her visual symptoms.

Few months later vision in her left eye deteriorated and she also developed some symptoms of paraesthesia. She was started on disease modifying treatment. In spite of treatment her vision progressively deteriorated over time and swelling of her optic disc remained unexplained.

Two years later she was referred back to ophthalmology for a second opinion and found to have new signs of complex ophthalmoplegia and vertical diplopia on lateral gaze in the left eye along with persistent disc swelling. B scan ultrasound confirmed the presence of disc drusen. She was referred to a neuro-ophtalmologist and repeat CT scan showed left sided proptosis and thickening of anterior portion of left optic nerve/sheath complex consistent with optic nerve meningioma. This was confirmed by subsequent MRI scan.

Discussion:

Optic nerve sheath meningiomas are uncommon benign tumours which account for approximately one third of primary optic nerve tumours and 5% to 10% of orbital tumours [1]. The diagnosis of optic nerve sheath meningioma requires a high index of suspicion and close interaction with the radiologist to ensure small tumours are not missed. The presence of additional comorbidity may confuse the clinical presentation.

The common presenting features include ipsilateral visual loss, afferent pupillary defect, visual field defect and optic disc swelling. The patients may also present with additional features of colour vision disturbances, proptosis, eye motility disturbances and lower eyelid oedema [2]. Ophthalmoscopic examination may reveal optic nerve head swelling, macular oedema, nerve pallor or choroidal folds. [1] Occasionally spontaneous improvement in visual function has also been observed [5].

The diagnosis of optic nerve sheath meningiomas is usually presumptive and based on the appropriate clinical picture supported by appropriate neuroimaging, however, accurate diagnosis depends on the clinical course and occasionally biopsy [4]. In our case although the patient presented with the common presenting features of optic nerve sheath meningiomas, certain factors like young age, female sex, symptoms of paraesthesia and normal
imaging scans on initial presentation suggested diagnosis of optic neuritis. Multiple white matter lesions consistent with demyelination on repeat MRI scan and raised CSF IgG further supported the diagnosis of Multiple sclerosis. The common CT scan findings in optic nerve sheath meningiomas are; a well defined tubular thickening of the optic nerve more commonly on the orbital apex, tram-track-sign or linear calcification of the optic nerve. The diagnosis can easily be overlooked on routine neuroimaging if the size of tumour is extremely small [3]. The other reason the diagnosis was missed was the MRI scan performed was a routine scan and not the MRI scan with high field strength T1-weighted of the brain and orbit, with fat suppression and gadolinium contrast which currently remain the procedure of choice for diagnosis of optic nerve sheath meningiomas [1]. The patient was started on disease modifying treatment for Multiple sclerosis but there was no improvement in symptoms. The persistent swelling in optic disc and deterioration in visual symptoms despite treatment lead to further investigations. Repeat CT scan and MRI done four years later revealed left sided proptosis and thickening of anterior portion of left optic nerve/sheath complex consistent with optic nerve sheath meningioma. The symptoms of paraesthesia and raised CSF IgG suggested that patient had multiple sclerosis as a co-morbidity which made diagnosis of optic nerve sheath meningioma more complex. Our case illustrates that ONSM can be a diagnostic dilemma in the presence of a co-morbidity. A high index of suspicion along with positive interaction with the radiologists can be instrumental in quick diagnosis.

**Differential Diagnosis List:** Optic nerve sheath meningioma

**Final Diagnosis:** Optic nerve sheath meningioma

**References:**


**Figure 1**

**a**

*Description:* CT scan showing proptosis and thickening of left optic nerve/sheath complex.

**b**

*Description:* Thickening and enhancement of left optic nerve sheath.
Description: MRI showing area of enhancement in left optic nerve Origin: