CASE REPORT

A Rare Case of Optic Nerve Sheath Meningioma

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Abstract

Optic nerve sheath meningiomas (ONSMs) are benign neoplasms originating from the arachnoid cap cells located in the dural sheath surrounding the orbital or intracanalicular portion of the optic nerve. Multiple meningiomas have most commonly been found to be associated with neurofibromatosis type 2 (NF2). Although optic nerve sheath meningiomas are benign and slow growing, they tend to cause severe vision loss at later stages. Patients with meningiomas at one site should be screened for meningiomas at other locations and also for the presence of neurocutaneous stigmata.

Keywords: Meningiomas, Neurofibromatosis, Optic nerve sheath.

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Introduction

Optic nerve sheath meningiomas are rare benign neoplasms originating from the arachnoid cap cells located in the dural sheath surrounding the orbital or intracanalicular portion of the optic nerve.[1] Meningiomas are also the second most common intradural spinal tumors, accounting for about 25% of all adult intraspinal tumors. Multiple meningiomas have most commonly been found to be associated with neurofibromatosis type 2 (NF2), caused by NF2 gene mutations on chromosome 22q12.[2] More than half of NF2 patients are prone to develop at least one intracranial meningioma in their lifetime.[3] Here we report a rare presentation of a middle-aged female found to have optic nerve sheath meningioma associated with a spinal meningioma and neurocutaneous stigmata.

Case Vignette

We report this case of a 40-year-old lady who presented to us with two month's history of gradually progressive spastic paraparesis. She had a tight band-like sensation over the upper abdomen, poorly localized low backache, flexor spasms and numbness in both her lower limbs, and urinary disturbance in the form of detrusor sphincter dyssynergia. She also gave a history of painful protrusion of her right eye associated with gradually progressive vision loss and also hearing a disturbance in the right ear for the past 15 years. Their family history was unremarkable.

On examination her higher mental status was normal. General examination showed multiple hyperpigmented macules and patches of size ranging from 3 to 15 mm with smooth borders present over her back and abdominal region. Ophthalmological examination showed right eye exophthalmos (Fig. 1) with periorbital edema and congestion. Visual acuity in the left eye was 20/40. The right eye showed absent light perception with optic atrophy on fundus examination. Extraocular movements were restricted in all the directions in the right eye. Motor examination showed symmetrical spastic paraparesis with motor strength of muscle power assessment (MRC) Grade 1/5 in both lower limbs with

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Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images

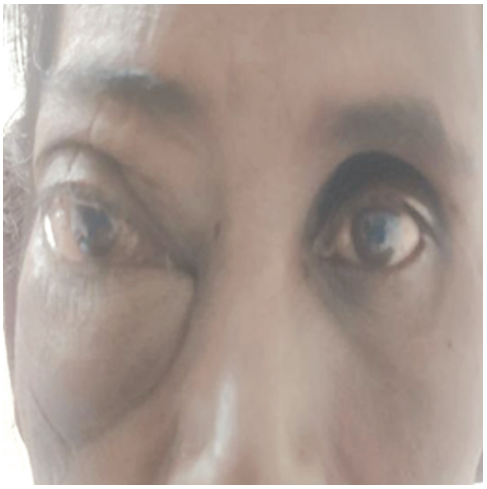


Fig. 1: Patient image showing right eye exophthalmos with periorbital edema and congestion

exaggerated deep tendon reflexes and extensor plantar response bilaterally. Sensory examination revealed reduced perception for all modalities below the D4 level.

On evaluation, all her routine blood investigations came out to be normal. Audiometry revealed a moderate conductive hearing loss in the right ear. Computed tomography imaging of the brain showed intraocular space occupying a lesion in the right eye isodense to brain parenchyma without hemorrhage or calcification (Fig. 2). Magnetic Resonance Imaging (MRI) of the spinal cord showed intradural broad-based T1 Isointense, T2 Isointense, uniformly enhancing space-occupying lesion in the spinal canal from C7 to T1 levels, not extending to spinal foramen suggesting a spinal meningioma (Figs 3 and 4). Similar T1 and T2 Isointense, uniformly enhancing space-occupying lesion also was seen in the right intraorbital region (Figs 5 and 6). Myelogram performed on the patient showed undisrupted cerebrospinal fluid (CSF) flow (Fig. 7). Histopathological examination of the resected spinal cord lesion confirmed it to be a meningioma. Post resection surgery patient power improved to MRC Grade 4-/5 in both lower limbs.

Discussion

Optic nerve sheath meningiomas (ONSMs) are rare accounting for only 2% of all orbital tumors and 1% of all meningiomas.[4] They usually are slow growing, painless, and benign but can cause severe visual loss owing to their critical location, directly affecting the anterior visual pathway. Patients with ONSMs tend to present earlier to the hospital due to vision disturbances compared to intracranial meningiomas. Our patient had long-standing vision disturbance, which was neglected and she presented to the hospital only after she developed paraparesis due to spinal meningioma. Due to this, her vision could not be salvaged with ONSM tumor resection. Patients with ONSM can present with any of these components of the classic triad (if not all three) vision disturbance, optic atrophy, and optociliary shunt vessels. As the tumor expands in size patient can

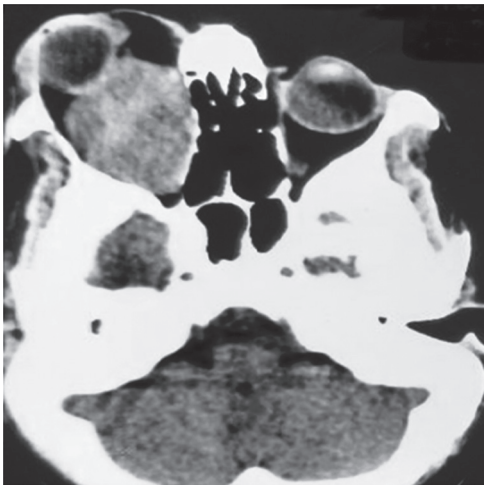


Fig. 2: Computed tomography imaging of the brain showing intraocular space occupying lesion in the right eye isodense to brain parenchyma without hemorrhage or calcification

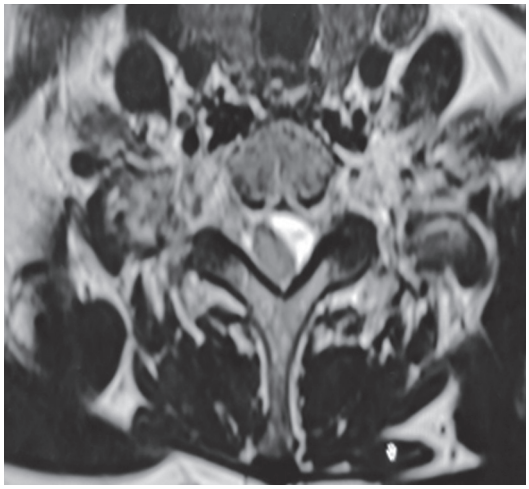
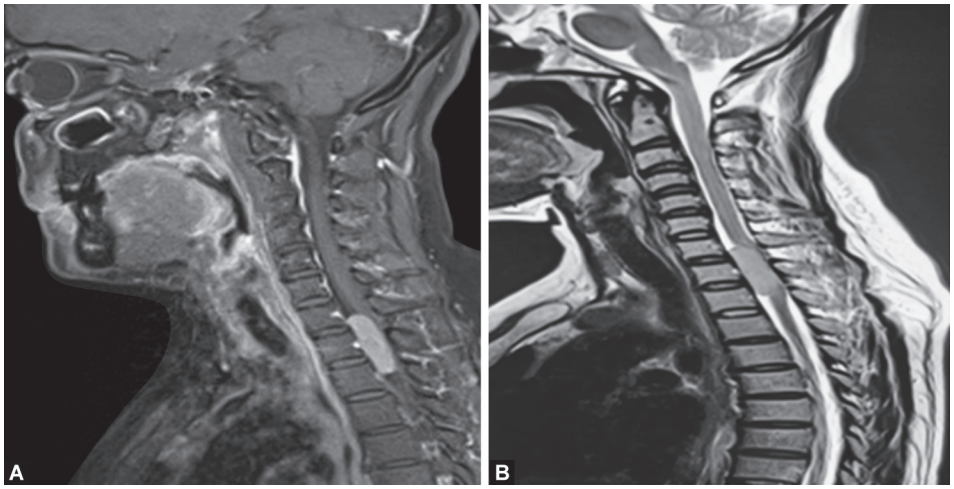
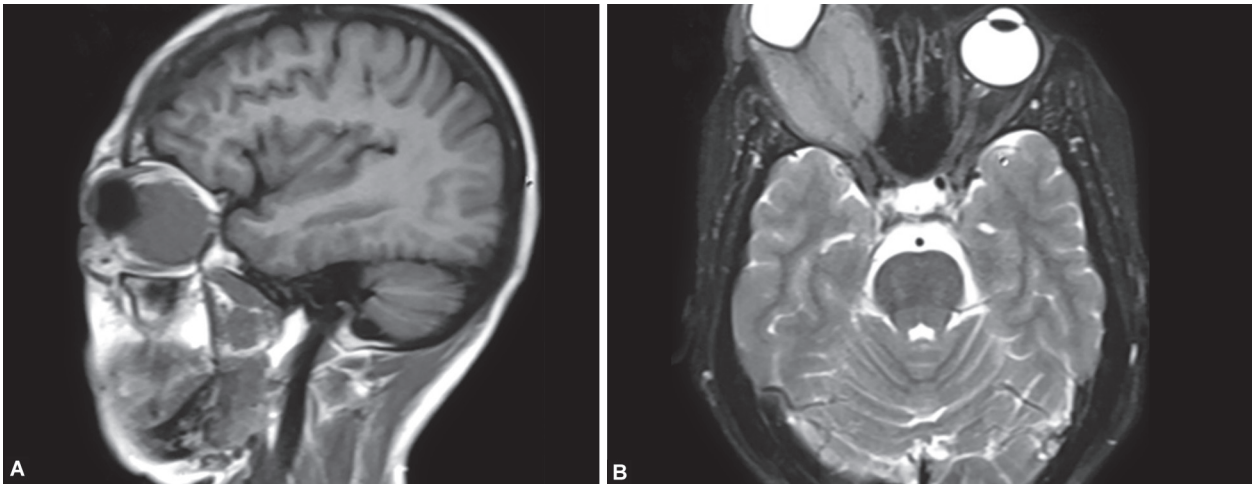


Fig. 3: MRI T1 and T2 weighted images show an isointense intraocular space-occupying lesion in the right eye



Figs 4A and B: Axial postcontrast T1 weighted image showing enhancement of intraocular space-occupying lesion with tram track sign appearance (linear non-enhancing optic nerve with surrounding enhancement)



Figs 5A and B: Axial T1 postcontrast MRI spine image showing an intradural broad-based uniformly enhancing lesion encroaching and compressing spinal cord from the left side

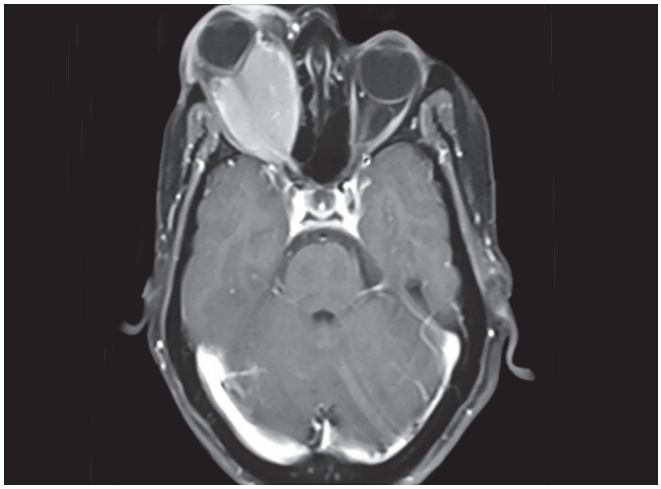
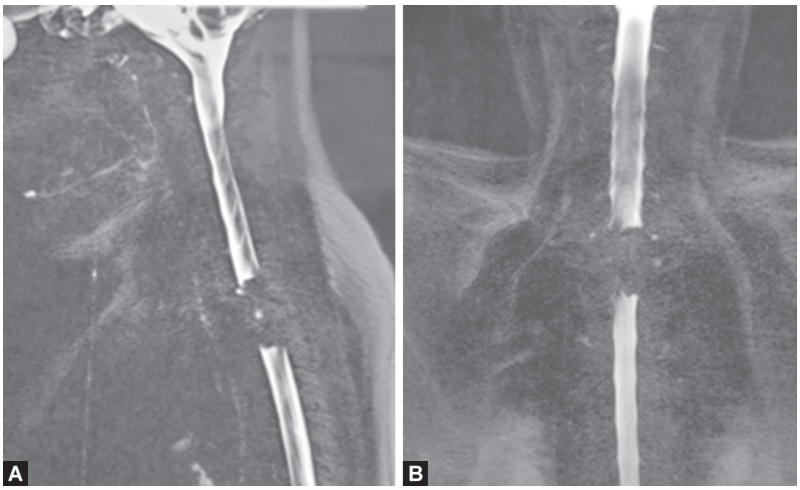


Fig. 6: Sagittal T1 postcontrast and T2 weighted images showing intradural uniformly enhancing space-occupying lesion from C7 to T1

also develop proptosis and restricted extraocular movements, both of which were present in our patient. The majority (95%) of ONSMs are unilateral. Rarely they may be bilateral or may be associated with meningioma at other locations, which is particularly seen with NF2 mutation.[5] In addition to ONSM and spinal meningioma, our patient also had multiple hyperpigmented macules and patches of varying sizes over her back and abdominal region suggestive of cafés au lait spots. Although café au lait spots are known to be classically seen in NF1, very rarely patients with NF2 also can have a few café au lait spots.[6] Due to the presence of meningiomas at two different locations and also the above-mentioned neurocutaneous stigmata, the patient was screened for the presence of other tumors, especially vestibular schwannoma. Despite her audiometry showing a hearing loss in her right ear, imaging was negative for schwannoma. Genetic testing for NF2 mutation could not be done in our case due to financial constraints. A final possible diagnosis of NF2 was made and the patient was asked to be on regular follow-up screening for neoplasms.



Figs 7A and B: Myelogram showing the corresponding space-occupying lesion spanning the entire spinal canal with normal CSF flow

Conclusion

Although ONSM are benign and slow growing, they tend to cause severe vision loss at later stages. Observation is an acceptable strategy when a visual function is intact, but with worsening acuity surgery may be considered. Magnetic Resonance Imaging is the gold standard for the diagnosis of ONSM. Patients presenting with multiple meningiomas at different locations should be thoroughly examined for the presence of neurocutaneous markers and also meticulously screened for vestibular schwannoma even if they are otherwise asymptomatic.

References

1. Dutton JJ. Optic nerve sheath meningiomas. Surv Ophthalmol 1992; 37(3):167–183. DOI: 10.1016/0039-6257(92)90135-g.
2. Kerr K, Qualmann K, Esquenazi Y, et al. Familial syndromes involving meningiomas provide mechanistic insight into sporadic disease. Neurosurgery 2018;83(6):1107–1118. DOI: 10.1093/neuros/nyy121.
3. Smith MJ. Germline and Somatic Mutations in Meningiomas. Cancer Genet 2015;208(4):107–114. DOI: 10.1016/j.cancergen.2015.02.003.
4. Parker RT, Ovens CA, Fraser CL, et al. Optic nerve sheath meningiomas: prevalence, impact, and management strategies. Eye Brain 2018; 10:85–99. DOI: 10.2147/EB.S144345.
5. Bosch MM, Wichmann WW, Boltshauser E, et al. Optic nerve sheath meningiomas in patients with neurofibromatosis type 2. Arch Ophthalmol 2006;124(3):379–385. DOI: 10.1001/archopht.124.3.379.
6. Kresak JL, Walsh M. Neurofibromatosis: A Review of NF1, NF2, and Schwannomatosis. J Pediatr Genet 2016;5(2):98–104. DOI: 10.1055/s-0036-1579766.