A rare case of bilateral optic nerve sheath meningioma

Somen Misra, Neeta Misra, Pratik Gogri, Rajen Mehta

A 60-year-old female presented with gradual, painless, progressive diminution of vision, and progressive proptosis of left eye since 7 years. Ophthalmological examination revealed mild proptosis and total optic atrophy in the left eye. Magnetic resonance imaging (MRI) and computed tomography (CT) brain with orbit showed bilateral optic nerve sheath meningioma (ONSM) involving the intracranial, intracanalicular, intraorbital part of the optic nerve extending up to optic chiasma and left cavernous sinus.

Key words: Cavernous sinus involvement, chasmal involvement, optic atrophy, optic nerve sheath meningioma

Optic nerve sheath meningioma (ONSM) is a benign neoplasm of meningothelial cells of the arachnoid tissue. ONSMs account for less than 1% of all meningiomas and 5-10% of all orbital tumors.[1,2] They most commonly present in patients between 30 and 50 years of age. There is a female dominance in the ratio of 3:1.[3] These lesions can arise primarily from the optic nerve sheath or may involve the optic nerve secondarily after arising from the cavernous sinus, falciform ligament, sphenoid wing, pituitary fossa, planum sphenoidale, frontoparietal area, or the olfactory groove.

Bilateral cases of ONSMs are rare and represent just 5% of all reported cases.[4-6] We present a case of bilateral secondary ONSM involving both the optic nerves, optic chiasma, and the left cavernous sinus. Besides being bilateral, this case was unusual since it showed different growth patterns on the two sides: Tubular around the right optic nerve and fusiform around left. This is a rare presentation of ONSM.

Case Report

A 60-year-old female presented with gradual, progressive, painless diminution of vision and mild protrusion of the left eye since the last 7 years. On clinical examination, her visual acuity was 20/30 in the right eye and no perception of light in the left eye. Left eye was deviated inwards [Fig. 1] with restriction of movement in all directions of gaze except adduction. Movements of right eye were free and full in all directions of gaze. There was 4 mm proptosis of the left eyeball [Fig. 1]. The left pupil was mid-dilated and fixed. Fundus showed total optic atrophy [Fig. 2]. In the right eye, anterior segment and fundus examination were within normal limits. Systemic examination also was within normal limits with no evidence suggestive of neurofibromatosis type 2.

Post-contrast T1-weighted axial magnetic resonance imaging (MRI) with fat saturation showed a well-defined mass encasing the orbital portion of the left optic nerve showing bulbous enlargement at the apex with distal tubular enlargement [Fig. 3]. Also, there was intracranial extension through the left optic canal into the suprasellar cistern involving the optic chiasma and the left cavernous sinus. There was a similar mass surrounding the intraorbital, intracanalicular, and intracranial part of the right optic nerve in a tubular fashion extending up to the optic chiasma. Both the masses showed intense heterogeneous enhancement with central relatively lesser enhancement of the optic nerve, producing the "tram-track sign". Non-contrast computed tomography (CT) brain and orbit showed areas of calcification around both optic nerves and optic chiasma [Fig. 3].

In our patient, surgical intervention did not appear to be a wise choice because of the extensive nature of involvement. Since total optic atrophy had already developed in the left eye, preservation of vision of the right eye was of prime importance. The patient was hence advised fractionated radiation therapy, that too preferably stereotactic or conformal type as these are better than the conventional form. The patient was lost to follow-up after referral for radiation therapy.

Discussion

ONSMs are rare tumors of anterior visual pathway.[3] Primary ONSMs arise from meningoepithelial cap cells of the arachnoid villi and can develop at any location along the entire course of the optic nerve sheath.[3,7] Secondary ONSMs may arise from tissues outside the orbit, namely the cavernous sinus, falciform ligament, clinoid processes, sphenoid wing, pituitary fossa, planum sphenoidale, tuberculum sellae, frontotemporal dura, and/or olfactory groove; and secondarily grow into the optic nerve sheath.

Dutton reported the mean age at presentation of ONSM patients to be 40.8 years (42.5 years in women and 36.1 years in men; range 3–80 years), with 61% of the patient population being female.[3] ONSM is usually unilateral. Only 5% of ONSMs present bilaterally, and 65% of these bilateral lesions are intracanalicular.[3,4] Approximately 50% of the patients who present with bilateral ONSMs also have tumors along the planum sphenoidale in continuity with these lesions, a finding that raises questions about the true origin of bilateral ONSMs. Bilateral and multifocal presentations of ONSMs are most commonly found in patients with neurofibromatosis type 2.[3,7,8]

As ONSMs progress, it is thought that they compromise optic nerve function mainly by mass effect on the pial vasculature

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/0301-4738.136238

Department of Ophthalmology, Rural Medical College of Pravara Institute of Medical Sciences, Loni, Maharashtra, India

Correspondence to: Dr. Somen Misra, Department of Ophthalmology, Rural Medical College, Loni-413 736, Taluka Rahata, District Ahmednagar, Maharashtra, India. E-mail: drsomenmisra@hotmail.com

Manuscript received: 26.03.13; Revision accepted: 16.07.13

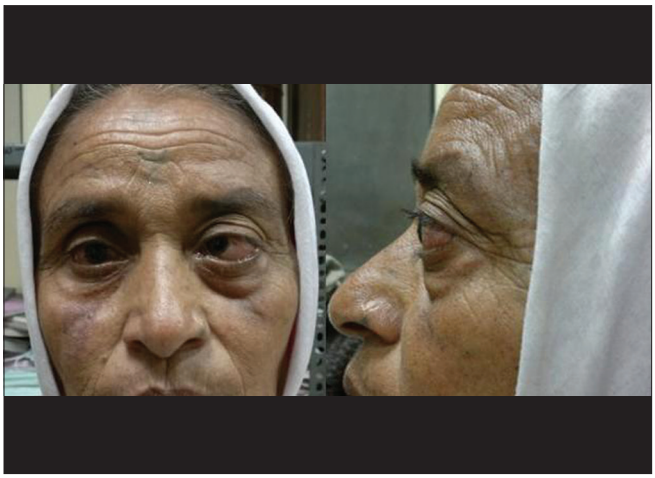

Figure 1: Left eye esotropia and proptosis of left eye

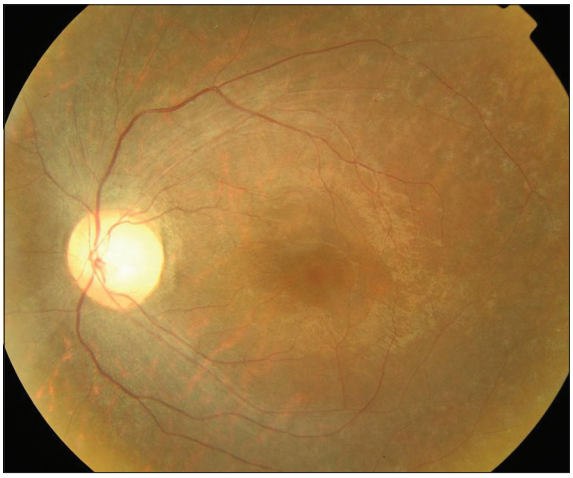

Figure 2: Total optic atrophy in left eye

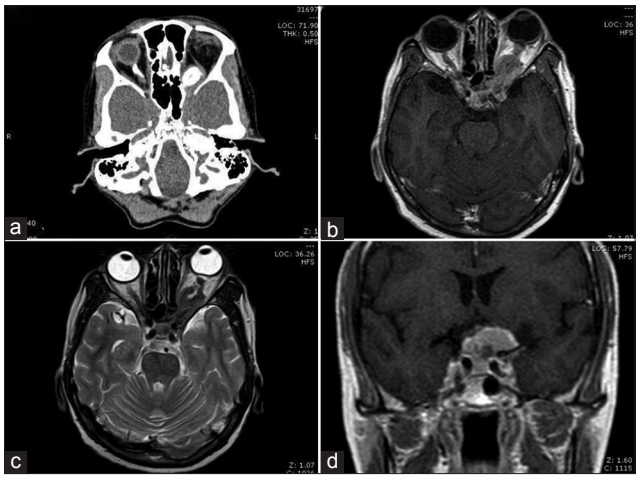

Figure 3: (a) Computed tomography brain and orbit showing calcification around both the optic nerves. (b-c) Magnetic resonance imaging brain with orbit showing optic nerve sheath meningioma having intraorbital, intracanalicular, and intracranial involvement on both sides. (d) MRI brain with orbit post-contrast showing suprasellar cistern and left cavernous sinus involvement

which induces ischemic changes as well as interferes with axonal transport in the nerve.[3,7] Patient may present with classic triad of visual loss, optic atrophy, and the presence of opticociliary shunt vessels on the disc. However, this is present in only a minority of patients.[7,8] Fundus examination almost always demonstrates a pathological appearance of the optic disc, which may consist of disc edema suggesting some manifestation of a compressive optic neuropathy or frank optic atrophy.

Once the diagnosis of an ONSM is suspected, the diagnosis can usually be established using MRI or high-resolution CT scan.[3] The characteristic features of ONSMs include the presence of calcification surrounding the nerve along with different radiographic growth patterns: Tubular, globular, fusiform, and focal.

Definitive treatment of ONSMs is challenging because of the lesions' intimate circumferential relationship with the optic nerve and its vascular supply. Surgical excision has almost always resulted in blindness in the affected eye. This could be due to excision of the tumor along with the affected optic nerve intraoperatively or damage to the pial vasculature.[3,7,8] Conservative management is indicated if there is no significant progressive visual dysfunction, or intracranial extension of the tumor. Recently, it has been highlighted by Miller that surgery to remove an ONSM is rare if ever indicated and radiation is the optimum therapy.[9] Also Turbin *et al*., from his comprehensive study of comparison of various treatment options for ONSM concluded that fractionated radiation beam therapy was preferable over surgery or conservative approach of management especially when preservation of vision was of prime importance as is the case in our patient.[10]

The above reported case of secondary ONSM involving the left cavernous sinus, optic chiasma and intracranial, intracanalicular as well as intraorbital portions of both the optic nerves is rare because it is bilateral and showed different growth patterns on the two sides, being tubular on the right and fusiform on the left side. We recommend considering ONSM as one of the differential diagnosis in a case of very slowly progressive unilateral optic neuropathy especially when presenting along with proptosis.

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Cite this article as: Misra S, Misra N, Gogri P, Mehta R. A rare case of bilateral optic nerve sheath meningioma. Indian J Ophthalmol 2014;62:728-30.

Source of Support: Nil. **Conflict of Interest:** None declared.