Optociliary Shunt Vessel: An Ancillary Ophthalmic Sign Harbingering Optic Nerve Sheath Meningioma

A 61-year-old male presented with gradual loss of vision in his right eye for one year at neuro ophthalmology clinic. On clinical examination, his visual acuity was perception of light and intraocular pressure was 12 mm Hg in right eye. Also, there was relative afferent pupillary defect in the right eye. Fundus examination showed peripapillary atrophy, temporal pallor in optic disc with dilated and tortuous optociliary shunt vessels in right eye. Presence of optociliary shunt, visual loss, relative afferent pupillary defect and optic atrophy opt us for further evaluation by visual field and magnetic resonance imaging. Visual field showed unilateral blindness suggesting lesion of optic nerve. Magnetic resonance imaging brain revealed optic nerve sheath meningioma.

Key words: Meningioma, Optociliary shunt, Relative afferent pupillary defect

Optociliary shunt are the collateral retino-choroidal vessels straddling the optic disc border. They carry venous blood from the retinal circulation to the choroidal circulation. These are seen in central retinal vein occlusion (CRVO), optic disc drusen, optic nerve sheath meningioma, sphenoid-orbital meningioma, optic nerve glioma, meningocele of the optic nerve, chronic papilloedema, high myopia, chronic glaucoma and congenital anomaly. The disturbance in the blood draining function of the central retinal vein due to any disease process is supposed to be major pathology for the development of optociliary shunt. This can be intraocular as well as retrobulbar. Optic nerve sheath meningioma causes the compression on optic nerve along with central retinal vein leading to formation of optociliary shunt. It presents typically with a triad of vision loss, optic...
nerve pallor, and optociliary shunt vessels\cite{4,5}. Optic disc photograph and Magnetic Resonance Imaging (MRI) helps the clinician to reach a diagnosis\cite{5}. Timely referral by an ophthalmologist and coordination with a neurosurgeon is vital in the management of this disease. Literature on this very important but rare sign from Nepal could not be found. Therefore, this paper would be knowledgeable to the ophthalmic and neuroscience worker.

Case Report

A 61-year-old male farmer presented with gradual loss of vision in his right eye for a year at neuro ophthalmology clinic of Lumbini Eye Institute. He did not experience headache, nausea, vomiting, floaters or flashes of light during this period. He did not have any history of systemic illness, surgery and trauma. On clinical examination, his visual acuity was perception of light (PL) and 6/6 in right and left eye respectively. The intraocular pressure (IOP) was 12 mm of Hg in both eyes. There was relative afferent pupillary defect (RAPD) in the right eye.

Slit-lamp bio microscopy did not reveal abnormality. Fundus examination showed peripapillary atrophy, temporal pallor in the optic disc with dilated and tortuous optociliary shunt vessels in right eye whereas left eye was within normal limits (Figure 1). Extraocular movements were full without ptosis. Corneal sensation was intact.

Visual field test revealed right sided unilateral blindness suggesting lesion of optic nerve. MRI brain revealed optic nerve sheath meningioma arising from the meninges at sphenoid wing compressing optic nerve in tram track fashion with calcification (Figure 2). The patient was referred to the neurosurgeon for further management.

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Figure 1: Fundus photo showing temporal optic disc pallor and optociliary shunt vessel in right eye and a normal left eye.

Figure 2: MRI brain showing a meningioma at sphenoid wing compressing intracranial optic nerve.
Discussion

Visual loss, optic atrophy and optociliary shunt vessels are the classical triad of Optic nerve sheath meningioma (ONSM) although, this triad presenting in its entirety is rare. According to Wright, presenting acuity of “no perception of light” (NLP) is seen in 24% of patients. Disc swelling, optic atrophy and optociliary shunt are the optic disc abnormality seen in ONSMs.

Optociliary shunt vessels are seen in only 30% of ONSMs. They are dilated normal anastomoses between the retinal venous system at the optic disc and the choroidal venous circulation. It is thought that they develop due to compression of the central retinal vein by tumor as it passes through the optic nerve. Shrinkage of optociliary shunt vessels has been observed after surgical removal and radiotherapy treatment supporting this thought.

Optociliary shunt vessels are generally missed by ophthalmologists as they resemble collaterals and neovascularization at disc (NVD). They can be differentiated by their larger caliber, tortuosity and lack of bleeding from them. MRI is the standard investigation in these cases to detect the cause. Thus detection of optociliary shunt helps in early diagnosis of meningioma.

This case highlights that optociliary shunt vessel is an ancillary ophthalmic sign harbingering optic nerve sheath meningioma. Though there are many causes of optociliary shunt, optic nerve sheath meningioma should be kept in mind in setting of visual loss and optic atrophy. Observation, radiotherapy and surgery are the modality of treatment for these patients, thus needing care from a neurosurgeon, whereas an ophthalmologist has role in early detection of it and in monitoring of treatment outcome.

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