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Primary Optic Nerve Sheath Meningioma with Choroidal Neovascular Membrane-A Rare Association

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Abstract *- **Primary optic nerve sheath meningiomas (ONSMs) are rare tumors arising from meningoendothelial cells of anterior visual pathway and constitute approximately 2% of all orbital tumors and 1–2% of all meningiomas. A 25 year old male presented with painless diminution of vision and slowly progressive outward protrusion of right eye since 12 months. His visual acuity was 20/120 in right eye and 20/20 in left eye. Reaction to light was sluggish in right eye and relative afferent pupillary defect was present. Proptosis was 25 mm in right eye and 18 mm in left eye. On automated perimetry, right eye showed enlargement of blind spot while left eye was normal. On fundus examination, right eye showed marked optic nerve head swelling, venous dilatation, tortousity along with extensive superficial and deep haemorrhages and a hypopigmented lesion superiorly around half disc diameter from optic disc. Fundus flouroscein angiography showed disc hyperflourescence, hypoflourescence due to haemorrhages and hypoflourescence in a lesion superior to optic disc with late leakage, suggestive of scarred CNVM. Contrast enhanced CT scan orbit showed tram track appearance of right optic nerve suggestive of right primary optic nerve sheath meningioma. Systemic examination revealed no other evidence of neurofibromatosis and there was no family history of hearing loss or central nervous system tumours. Thyroid hormones were within normal limits. Definitive treatment of primary ONSMs is challenging. Surgical excision has almost always result in blindness in affected eye. This could be due to excision of tumor along with affected optic nerve intraoperatively or damage to pial vasculature. Conservative management is indicated if there is no significant progressive visual dysfunction or intracranial extension of tumor.***

Keywords *- Primary Optic Nerve Sheath Meningioma, choroidal neovascular membrane, proptosis, hyperfluouroscence.*

Introduction

Primary optic nerve sheath meningioma (ONSM) is a benign neoplasm of meningothelial cells of arachnoid tissue. Primary 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 predominance in ratio 3:1.[3] It may cause gradual painless visual loss and is typically present for 1-5 years before clinical presentation.[4] Slowly progressive monocular visual loss, optic atrophy and presence of optociliary collateral vessels known as Hoyt Spencer triad are classic sign of primary ONSM.[5] The most common visual field defect is peripheral constriction although other field defects such as blind spot enlargement, altitudinal field defects and central scotomas have been described.[6] In patients with primary ONSM,optic disc abnormalities are nearly always visible at time of presentation[7] (98%). Chronic disc swelling occurs when tumor surrounds or compresses intraorbital part of optic nerve.[8] Optic atrophy may be subtle and is a late finding as patient's optic disc swelling resolves and optociliary collateral vessels may appear on disc surface.

Subretinal choroidal Neovascular Membrane formation is an important feature of older adults as a part of wet age related macular degeneration. Other conditions that can predispose to CNVM include ocular toxocariasis, optic disc drusen, traumatic choroidal rupture, radiation retinopathy, degenerative myopia, histoplasmosis, angioid streaks and optic nerve pit.[9] It is very rare with optic nerve sheath meningioma. The exact pathogenic association of disc swelling with subretinal neovascularisation is still unclear. Morse et al[10] suggested two plausible factors for its

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occurrence in papilloedema, the first being an anatomical dehiscence due to physical deformation of peripapillary tissue creating a pathway for ingrowth of new vessels, and the second, hypoxia caused by axonal swelling leading to impaired vascular perfusion of the tissues and hence neovascularisation

Radiological findings of primary optic nerve sheath meningioma are usually an enlarged optic canal, optic nerve thickening and enhancement and calcification within the tumor.[11] Definitive treatment of primary ONSM is controversial. Fractionated radiotherapy is often the first choice in an attempt to control the ONSM growth rate and to preserve vision. Surgery remains the best treatment in cases with blindness, severe proptosis and in patients with intracranial extension.[12]

Case Report

A 25-year-old male presented with gradual progressive, painless diminution of vision and outward protrusion of right eye since past 12 months. On ocular examination, his visual acuity was 20/120 in right eye and 20/20 in left eye. Reaction to light was sluggish in right eye while reaction to light was brisk in left eye. Relative afferent pupillary defect was present in right eye. Extraocular muscles movements were restricted in all directions of gaze in right eye while were full range in all directions in left eye. The degree of axial proptosis for right eye was 25 mm which was significantly different from value of 18 mm in left eye. The proptosis was not reducible and there was no bruit over it.On automated perimetry, right eye showed enlargement of blind spot (Figure 1) while left eye was within normal limits.

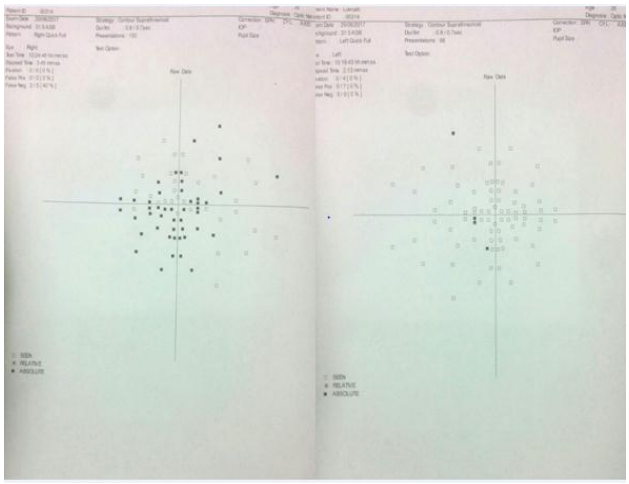

FIGURE-1

On fundus examination, right eye showed marked optic nerve head swelling, venous dilatation and tortousity along with extensive superficial and deep haemorrhages and a hypopigmented lesion half disc diameter superior to optic disc (Figure 2).

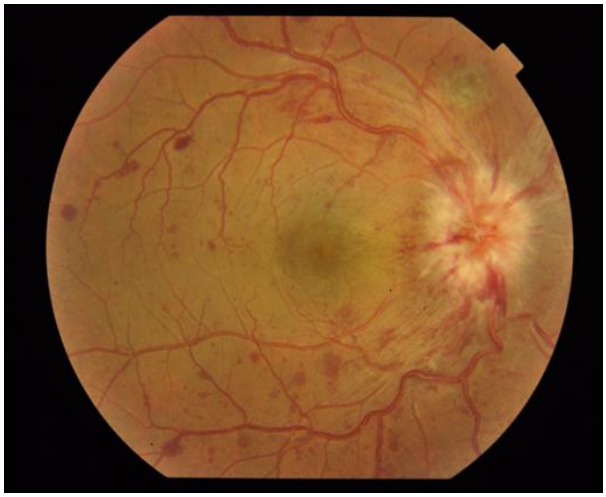

FIGURE-2

Fundus flouroscein angiography showed disc hyperflourescence,hypoflourescence due to haemorrhages and hypoflourescence in a lesion superior to optic disc with late leakage, suggestive of scarred CNVM(Figure 3)

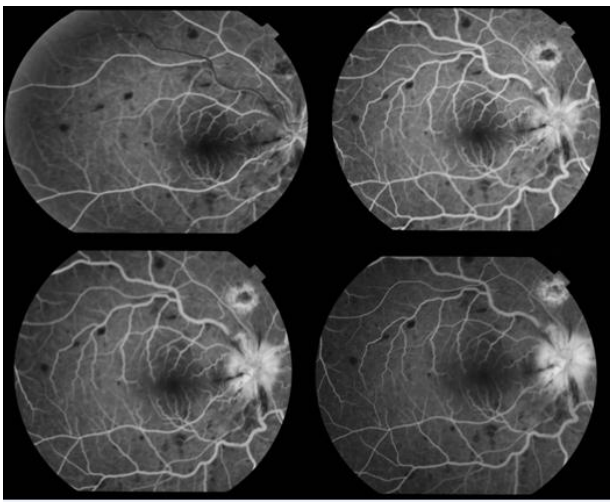

FIGURE-3

Contrast enhanced CT scan orbit showed moderate enlargement and peripheral enhancement of right optic nerve giving rise to tram track appearance suggestive of right primary optic nerve sheath meningioma (Figure 4)

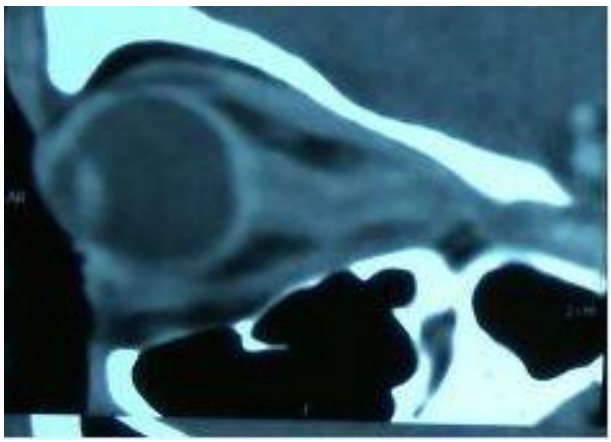

FIGURE-4

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CT Scan Brain was within normal limits. Systemic examination revealed no other evidence of neurofibromatosis, and there was no family history of hearing loss or central nervous system (CNS) tumours. Thyroid hormones were within normal limits.

Discussion

Primary ONSM are rare benign tumors of optic nerve. The tumors grow from cells that surround the optic nerve and as tumor grows, it compresses the optic nerve. This causes loss of vision in affected eye. Rarely, it may affect both eyes at same time. It is typically a slow growing tumor and has never been reported to cause death. However, there is concern that the tumor can grow into the brain and cause other types of neurological damage. In some patients, the tumor grows so slowly that the treatment is not necessary.[13] 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] Primary ONSM is usually unilateral. Only 5% of ONSMs present bilaterally and 65% of these bilateral lesions are intracanalicular. Approximately 50% of the patients who present with bilateral ONSMs also have tumors along planum sphenoidale in continuity with these lesions. As ONSMs progress, it is thought that they compromise optic nerve function mainly by mass effect on the pial vasculature which induces ischemic changes as well as interferes with axonal transport in the nerve.[14] Patient may present with classic triad of visual loss, optic atrophy and presence of opticociliary shunt vessels on the disc. 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. Definitive treatment of primary ONSMs is challenging because these 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.[15] Conservative management is indicated if there is no significant progressive visual dysfunction or intracranial extension of 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.[16] 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. Despite the extensive literature on the subject, there is still considerable controversy regarding the natural history of these tumors and their appropriate management. Loss of vision and progressive proptosis are the two most common symptoms, present in the vast majority of patients. Whether proptosis or visual dysfunction is the initial complaint probably depends on the site of the original tumor. Children with primary optic nerve sheath meningiomas generally present a greater optic canal, sphenoid wing and/or intracranial involvement on presentation as well as occasionally bilateral optic nerve involvement, recurrence after treatment and even intraocular extension.[17]

While we cannot eliminate the possibility that the association of CNV with meningiomas and CNV in our patient is merely by chance, several observations suggest otherwise. In our patient, the fundus of the fellow eye was free of drusen, let alone more substantive evidence of macular degeneration. Patient does not had a disorder known to predispose to CNV. Schatz et al[18] published the histopathological findings in a patient with a primary optic nerve sheath meningioma in which there was a CNV. However, their patient had chronic disc oedema and venous collaterals and had antecedent age related macular degeneration in both eyes. Shields et al[19] described an instance of CNV in a child with an optic nerve glioma. That patient's disc was also oedematous. How might a meningioma cause an ipsilateral CNV? The pathophysiological mechanism by which these two conditions occur together is unclear. It is possible that the tumour tissue could have invaded the eye. CNV has been associated with other tumours involving the choroid.[20] Ocular invasion was not evident on ultrasound or MRI scans but absence of proof is not proof of absence. In the case of Schatz et al there were small foci of the meningioma in the peripapillary sclera and retrolaminar optic nerve, which were not seen before enucleation. Dutton reviewed meningiomas involving the optic nerve and primary optic nerve sheath meningiomas. CNV was not mentioned as a presenting sign. None the less, he calculated 3.7% of 477 reported cases described intraocular invasion by meningiomas. Other, authors have reported histopathological cases of meningioma invading the optic nerve and disc. We believe that the association of CNV with ipsilateral meningiomas in our patients was not one of chance. The presumed mechanism is invasion of the globe by the tumour sufficient to cause the CNV but below threshold for detection by MRI, ultrasound, or ophthalmoscopy.

Conclusions

Based on observations of this patient ,it can be believed that a young patient with unilateral CNVM, should be evaluated for optic disc swelling, optic disc atrophy or proptosis.If any of these conditions is present,then orbital imaging studies should be considered to exclude the possibility of an intrinsic neoplasm of an optic nerve.

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References

[1] Beccari S, Cima L, Posenato I, Sala F, Ghimenton C, Brunelli M, Eccher A. Pediatric optic nerve sheath meningioma. J Neuroophthalmol 2014; 34 (3):315-316.
[2] Zhou HF, Wei SH. New view in the disgnosis and therapy of optic nerve sheath meningiomas Guoji Yanke Zazhi(Int Eye Sci)2008;8(7):1451-1453.
[3] Litre CF, Colin P, Rousseaux P, Civit T. Optic nerve sheath meningiomas. Neurochirurgie 2010; 56(2-3):132-136.
[4] W. B. Wilson, "Meningiomas of the anterior visual system," Survey of Ophthalmology, vol. 26, no. 3, pp. 109–127, 1981
[5] N. R. Miller, "Primary tumours of the optic nerve and its sheath," Eye, vol. 18, no. 11, pp. 1026–1037, 2004
[6] J. W. Kim, J. F. Rizzo, and S. Lessell, "Controversies in the management of optic nerve sheath meningiomas," International Ophthalmology Clinics, vol. 45, no. 4, pp. 15–23, 2005
[7] C. S. Eddleman and J. K. Liu, "Optic nerve sheath meningioma: current diagnosis and treatment," Neurosurgical Focus, vol. 23, article E4, 2007
[8] J. W. Kim, J. F. Rizzo, and S. Lessell, "Controversies in the management of optic nerve sheathmeningiomas," International Ophthalmology Clinics, vol. 45, no. 4, pp. 15–23, 2005
[9] Goshorn EB, Hoover DL, Eller AW, Friberg TR, Jarrett WH, 2nd, Sorr EM. Subretinal neovascularization in children and adolescents. J Pediatr Ophthalmol Strabismus. 1995;32:178–182
[10] Morse PH, Leveille AS, Antel JP, Burch JV. Bilateral juxtapapillary subretinal neovascularisation associated with pseudotumor cerebri. Am J Ophthalmol 1981;91:312-7
[11] Saeed P, Rootman J, Nugent R. Optic nerve sheath meningiomas. Ophthalmology 2003;110:12
[12] Pacelli R, Cella L, Conson M, Tranfa F, Strianese D, Liuzzi R, et al. Fractionated stereotactic radiation therapy for orbital optic never sheath meningioma – a single institution experience and a short review of the literature. J Radiat Res 2011;52:6
[13] Turbin RE, Pokorny K. Diagnosis and treatment of orbital optic nerve sheath meningioma. Cancer Control 2004;11:334-41
[14] Khan SN, Sepahdari AR. Orbital masses: CT and MRI of common vascular lesions, benign tumors, and malignancies. Saudi J Ophthalmol 2012;26(4):373-383
[15] Misra S, Misra N, Gogri P, Mehta R. A rare case of bilateral optic nerve sheath meningioma.Indian J Ophthalmol 2014;62(6):728-730
[16] Kothari NA, Kulkarni KM, Lam BL. Untreated bilateral optic nerve sheath meningiomas observed for 27 years. J Neuroophthalmol 2013;33(1): 45-47
[17] Amoli FA, Mehrabani PM, Tari AS: Aggressive orbital optic nerve meningioma with benign microscopic features: a case report. Orbit 2007;26:271-274
[18] Schatz H, Green WR, Talamo JH, et al. Clinicopathologic correlation of retinal to choroidal venous collaterals of the optic nerve head. Ophthalmology 1991; 98:1287-93.
[19] Shields JA, Shields CL, De Potter P, et al. Choroidal neovascular membrane as a feature of optic nerve glioma. Retina 1997; 17:349-50.
[20] Green WR, Wilson DJ. Choroidal neovascularization. Ophthalmology 1986;93:1169-76

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