Case Studies

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OPTIC PATHWAY GLIOMA: A CHALLENGE FOR CLINICIAN

Shachi Jain Taran[1], Lubna Khan[2*] and Ravindra Kumar[3]
[1]Department of Pediatrics; [2]Department of Ophthalmology; [3]Central Research Laboratory,
Sri Aurobindo Medical College and PG Institute, Indore, Madhya Pradesh, India

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Abstract: Optic pathway gliomas are rare neuro-ophthalmic cause of vision loss. About half of optic gliomas occur as a component of neurofibromatosis type 1. Here we are reporting an unusual case of optic pathway glioma without neurofibromatosis (NF).

Keywords: Optic pathway gliomas (OPG), Retrobulbar, Canalicular and Prechiasmal Segments

INTRODUCTION

Optic pathway gliomas (OPG) are rare astrocytic neoplasms arising from the optic pathway that occur most commonly in the pediatric population [1, 2]. OPGs were first described in 1833 by a surgeon in Edinburgh who published a case report of a 13-year-old patient with significant proptosis. With the advent of modern technologies, OPG have been described to comprise 0.6 to 5.1% of all intracranial tumors, 1.7 to 7% of all gliomas, up to 3.5% of all orbital tumors and 66% of all primary optic nerve tumors [2]. The majority of patients are diagnosed below the age of 10 years and the most common histology in pre-pubertal children is pilocytic astrocytoma [3]. OPGs also have a known association with neurofibromatosis type 1[4].

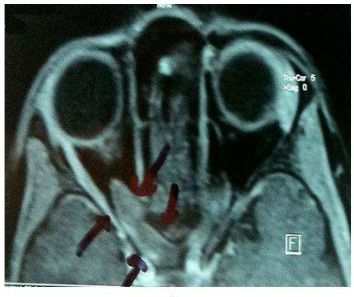
Case:

A five year old girl presented to us with complaints of squint right eye since age of two years. Parents noticed diminution of vision in the same eye progressing since last three months. There was no history of trauma, pain, redness of eye and any other systemic illness. On detail history she was full term vaginally delivered, second birth order with uneventful perinatal period. Milestones achieved at appropriate age and normal school performance at present. On examination there was one hyper pigmented macule at right hip 1.5x2 cm size, no freckling, no spine or leg deformity, no bumps and no other neurocutaneous marker. There was no history suggestive of hearing impairment. Other two siblings were normal. Family history was not suggestive of neurofibromatosis.

On ocular examination there was mild proptosis in right eye. The eyes were orthophoric in primary position for distance and near, but at intermediate distance there was alternating 15 degree exotropia. Anterior segment was within normal limits but right eye had an ill sustained pupillary reaction. Swinging flash light test revealed relative afferent pupillary defect in right eye. Ocular movements were full in all directions bilaterally.

On dilatation and retinoscopy an astigmatic error was detected bilaterally, more in right eye than in the left. Fundus examination showed temporal pallor bilaterally, oculus dexter was more affected than oculus sinister.

MRI revealed fusiform enlargement of right optic nerve involving posterior retrobulbar, canalicular and prechiasmal segment [figure 1A-D]. Intraorbital part of optic nerve just behind the globe appeared to be normal and was isointense to normal brain grey matter. No significant enhancement was seen. This fusiform segment measured two centimeter in length and eight millimeter in width. There was widening of optic canal on ipsilateral side with no evidence of hyperostosis or reactive bone changes. Rest of intraconal fat and content appeared normal. Left optic nerve, left orbital content and optic chiasm seemed to be normal. No intrinsic brain parenchyma abnormality was seen [figure 1 E&F]. Findings were suggestive of right optic nerve glioma.



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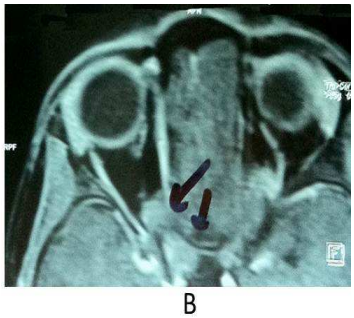

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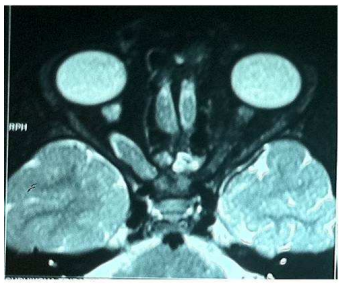
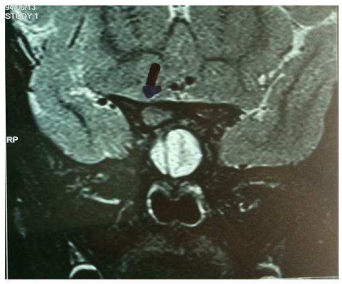
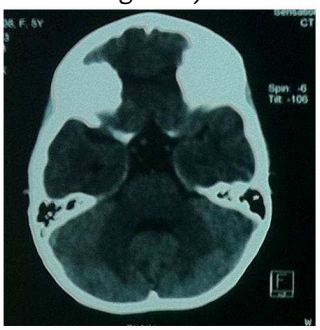
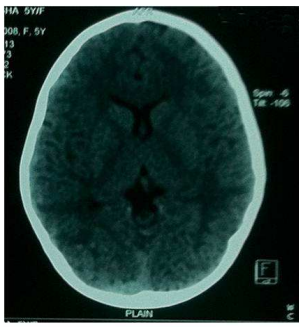
***Corresponding Author:**
Dr. Lubna Khan,
Associate Professor,
Department of Ophthalmology,
Sri Aurobindo Medical College and PG Institute,
Indore, Madhya Pradesh, India.

C D

Figure.1 A-D: showing fusiform swelling of right optic nerve (retrobulbar, canalicular and prechiasmal segment)

E F

Figure.1E&F: Showing no intrinsic Brain Parenchymal Lesion

It was considered appropriate to offer surgical management which was advised to the parents. However we could not procure consent from them, possibly since conservative management had already been advised elsewhere.

DISCUSSION

Optic pathway glioma is a very rare childhood neoplasm with peak age incidence between 2 to 6 years; 75% of all patients are less than 10 years old [5]. Its prevalence in the general population has been estimated to be about 1in 100000 [6].

These may be divided into three clinicopathologic entities: first anterior lesions- confined to the optic disc and nerve (25%); second chiasm lesions- may or may not involve the optic nerves (20-40%); third posterior lesions- involving the hypothalamus with possible extension to the optic tracts (33-60%) [7]. The most common histotype is low grade astrocytoma which in 10-70% of patients is associated with neurofibromatosis. WHO has classified them predominantly as grade 1 pilocytic astrocytoma.

Kornreich *et al.,* 2001 [8] studied several morphologic features that distinguish NF-OPG from non–NF OPG. They showed in the patients with NF, the most common site of involvement was the orbital nerve and the tumor was smaller than in the non-NF patients, the original shape of the optic pathway was preserved, and cystic components were uncommon. In the non-NF group, the chiasm and hypothalamus were the most common sites of involvement, the tumor was mass like and cystic components were frequently seen, as was extension beyond the optic pathways. The prognosis is also significantly different in these two groups-half the NF patients remained stable compared with only 5% of the non-NF patients.

Though our case belonged to non NF OPG but had MRI features and clinical course similar to that of neurofibromatosis group. So this case emphasizes that there may be certain patients who lie in grey zone. Possibly there is a third subset which has features common to both groups.

The clinical course and natural history of optic gliomas are highly variable, making treatment paradigm very difficult. Management includes observation with serial monitoring using MRI for asymptomatic slowly growing tumor. Chemotherapy is usually the first line treatment for progressive tumor in young children and radiotherapy for progressive disease in children older than 5-7 years. Surgery is reserved for large tumor causing mass effect or hydrocephalous and tumor confined to the orbit or unilateral optic nerve [7].

There are many researches ongoing for the diagnosis and management of these tumors like diffusion tensor tractography for diagnosis and surveillance, correlation of optical coherence tomography parameter with clinical and radiological progression, role of diffusion weighted MRI in predicting outcome, PLAN scoring system of anatomical classification, MRI internal segmentation, role of conformational radiotherapy, gamma knife surgery, medically induced hypothyroidism and carboplatin in the management. Though optic pathway gliomas are benign tumors, still a challenge for clinicians as no consensus guideline for management has been yet made.

REFERENCES

1. Liu GT, Katowitz JA, Rorke-Adams LB, Fisher MJ. Optic pathway gliomas: neoplasms, not hamartomas. JAMA Ophthalmol. 2013, 131, 646-50.

2. Mishra MV, Andrews DW, Glass J, Evans JJ, Dicker AP, Shen X, Lawrence YR. Characterization and outcomes of optic nerve gliomas: a population-based analysis. J Neurooncol. 2012, 107, 591-7.

3. Bilgiç S, Erbengi A, Tinaztepe B, Onol B. Optic glioma of childhood: clinical, histopathological, and histochemical observations. Br J Ophthalmol. 1989, 73, 832-7.

4. Sylvester CL, Drohan LA, Sergott RC. Optic-nerve gliomas, chiasmal gliomas and neurofibromatosis type 1. Curr Opin Ophthalmol 2006, 17, 7–11.

5. Dahlia F, Elisabetta P, Barbara T, Gianluca I, Ebtesam H, Sara T, Grazia T and Riccardo S. Radiation Therapy in Optic Gliomas: A Case Report and a Review of Literature. J Nucl Med Radiat Ther 2012, 3, 4.

6. Singhal S, Birch JM, Kerr B, Lashford L, Evans DG. Neurofibromatosis type 1 and sporadic optic gliomas. Arch Dis Child 2002, 87, 65-70.

7. Binning MJ, Liu JK, Kestle JR, Brockmeyer DL, Walker M. Optic pathway gliomas: a review. Neurosurg Focus. 2007, 23, E2.

8. Kornreich L, Blaser S, Schwarz M, Shuper A, Vishne TH, Cohen IJ, Faingold R, Michovitz S, Koplewitz B, Horev G. Optic pathway glioma: correlation of imaging findings with the presence of neurofibromatosis. AJNR Am J Neuroradiol. 2001, 22, 1963-9.

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