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Case Report
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OPTIC NERVE GLIOMA PRESENTING AS NON-AXIAL PROPTOSIS AND OPTOCILIARY SHUNT

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ABSTRACT

Optic nerve gliaoma is a slow growing tumor,which typically affects children. Nonaxial proptosis and optociliary shunt usually rare in such case. Here we present a case of optic nerve glioma in a 14 year old girl who presented with loss of vision and nonaxial proptosis and optociliary shunt on fundus examination in left eye in M.G.M.Medical College, Ophthalmology Department, Indore, India.

KEYWORDS: Glioma, Optic Nerve, Proptosis, Optociliary shunt, RAPD, EOM.

INTRODUCTION

Optic nerve glioma is a tumor of childhood [1] and presentation in adulthood may suggest malignant glioma. [2] Diagnosis is usually made before 5 years of age, but occasionally the presentation is in early adulthood[3]. Optic nerve glioma accounts for 0.6% to 1.2% of all intracranial tumors. [4] The incidence is 1 in 100,000 patients, with 90% presenting within the first two decades and 70% in the first decade. [5] It represents about 17% of all the orbital tumors encountered in childhood. [6]

About half of the optic nerve gliomas are confined to the orbit, whereas half demonstrate intracranial extension. [7] Intracranial extension may be suspected clinically on the basis of precocious puberty, somnolence, or diabetes insipidus. Any part of the optic pathway may be involved in a glioma, and prognosis depends in part upon the extent and location of the tumor. Patients with gliomas that both infiltrate and compress the optic nerve can eventually develop optociliary shunt vessels–congenital venous channels that enlarge in the setting of chronic compression of the optic nerve and that shunt blood from the retinal to the choroidal circulation. In general, the more anterior the lesion, the better the prognosis.

CASE REPORT

A 14 year old girl presented in the Ophthalmology Department in M.G.M. Medical college & MYH Hospital Indore, India with progressive, painless loss of vision in left eye, slowly progressive bulging of left eye (Fig.1) since 3 year ,which is associated with intermediate redness and watering. There was no associated history of trauma, pain, discharge, headache, vomiting, rhinorrhea, nasal obstruction.

There was no association of increase in the swelling on coughing or bending. There is no significant past and family and personnel history. Clinically she is well oriented to time ,person and place. On ophthalmic evaluation of left eye vision PL+ve, PR inaccurate in all 4 quadrants**,** nonaxial proptosis (on exophthalmometry 23mm axial and 7mm laterally displacement), 15 degree exotrophia, ocular movement restricted nasally and pupil shows RAPD. Right eye best corrected vision is 6/9, rest within normal limit. On fundus examination left eye shows inferiorly blurring and pale di sc and optocilialy shunt (Fig.2), right eye is normal.

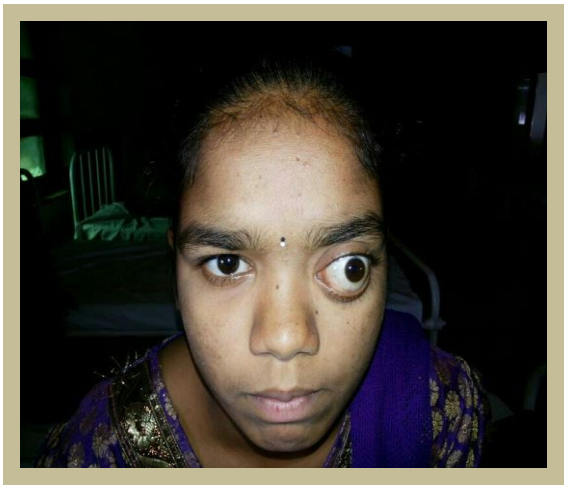

Fig. 1

Fig. 2

INVESTIGATION

1.Routine blood and urine investigations were in within normal limit.

2.MRI Brain and Orbit shows a well defined homogenously solid soft tissue mass lesion is seen in the intracranial retrobulbar space of left eye (Fig.3). Isointense on T1 (Fig.4), iso to hyper on T2 image (Fig.5). It has homogenous contrast enhancement (Fig.6). Lesion extending from globe to optic canal & normal optic nerve was not visualizes separately. Globe displaced anteriorly and Iaterally and EOM displaced peripherally.

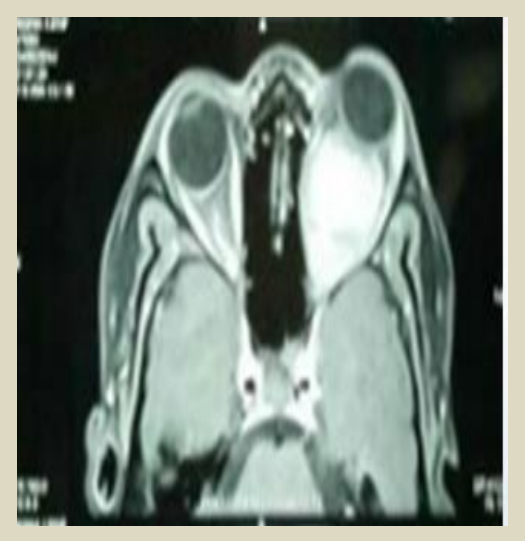

Fig. 3

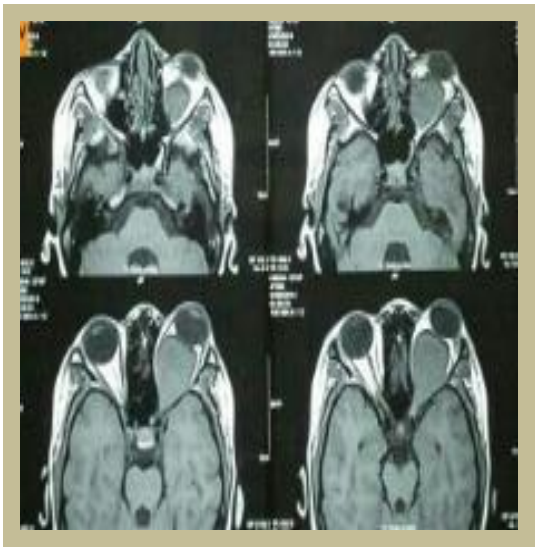

Fig.4

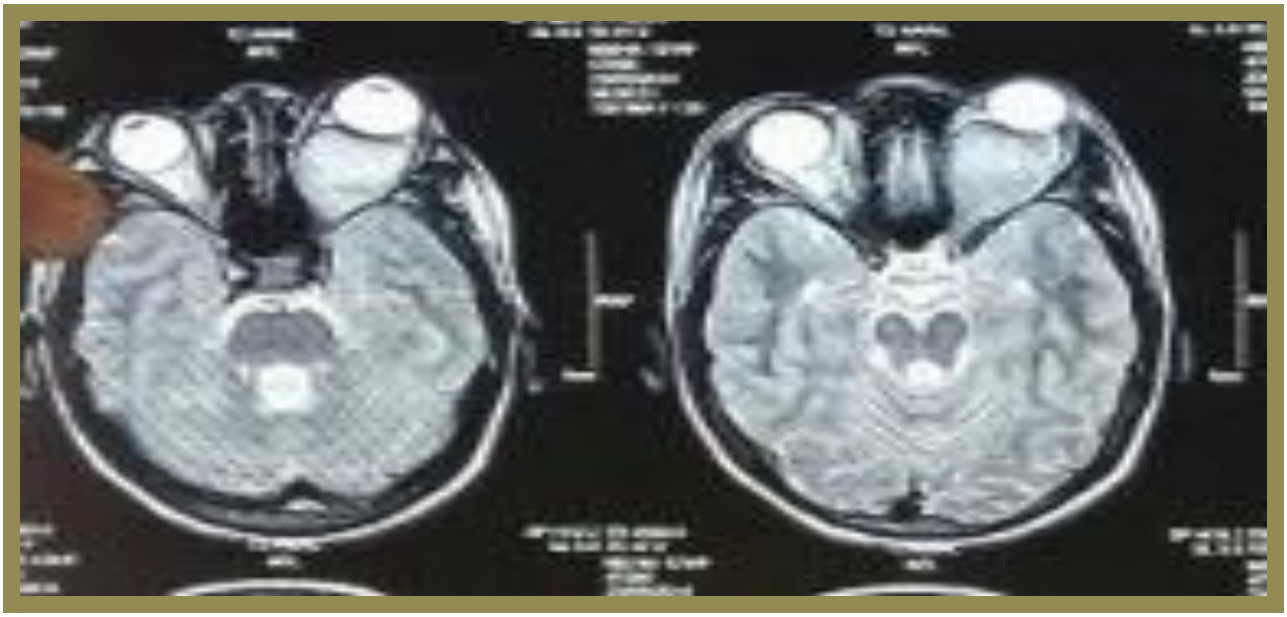

Fig. 5

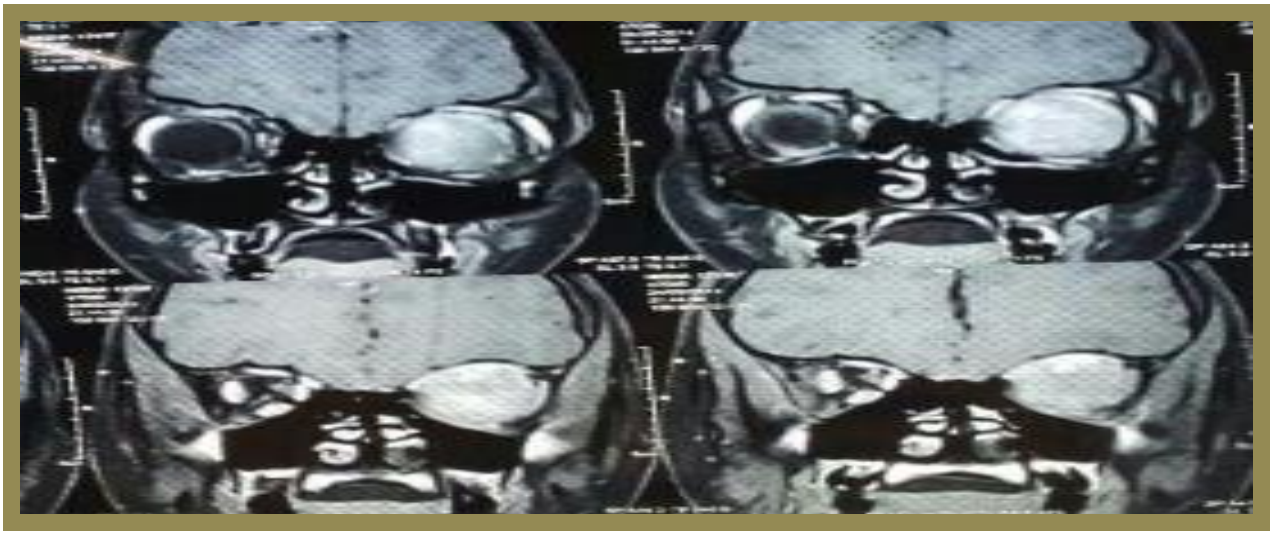

Fig. 6

TREATMENT AND FOLLOW UP

Patient was referred to neurosurgeon and surgery was done under general anesthesia with informed consent from parents. The procedure occurred without complications and the globe was sent for histological examination. Histological examination revealed juvenile pilocytic astrocytoma, characterized by cells with prominent eosinophilic processes called Rosenthal's fibres (Fig.7). There was no post –operative complication.

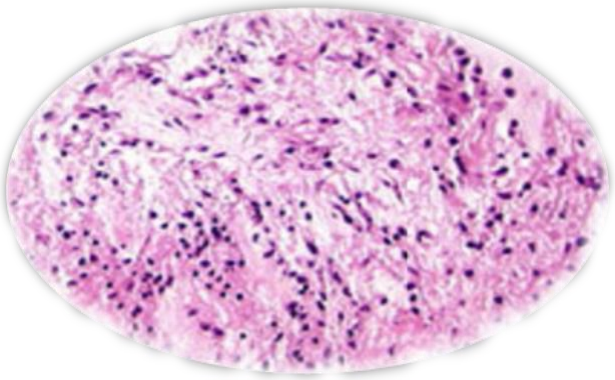

Fig 7

DISCUSSION

Optic nerve glioma is a slow-growing, pilocytic astrocytoma, which typically affects children. It is ectodermal in origin and is derived from astrocytes and oligodendroglial cells of the optic nerve. An ipsilateral afferent pupillary defect is present, visual acuity is deficient, and there is achromatopsia. Central scotomas are common, but temporal or bitemporal field loss may occur if the prechiasmal or chiasmal portion of the intracranial optic nerve is involved.[8] Optic nerve gliomas involving the orbit produce proptosis, ophthalmoplegia and painless progressive visual loss. [9] The optic nerve head may be

edematous, infiltrated with tumor, or atrophic. Rarely, optociliary venous collaterals may be present.

Neovascular glaucoma secondary to optic nerve glioma has also been reported. [10] Optic nerve glioma present as either a solitary manifestation or a component of neurofibromatosis. It commonly occurs in neurofibromatosis type 1(NF-1) and belongs to the diagnostic criteria of NF-1. [11] Histologically, optic nerve glioma is generally characterized as a juvenile pilocytic astrocytoma, characterized by cells with prominent eosinophilic processes called Rosenthal's fibres. The astrocytic nature of the tumor can be confirmed using immunohistochemical techniques with antibodies against glial fibrillary acidic protein. [4] Rarely, there is exophytic extenion of the tumor outside the optic nerve sheath. The over all histologic appearance is benign with paucity of cellular atypia, mitosis, or tumor necrosis.

Diagnosis of optic nerve glioma is usually based on orbital imaging. CT scan and MRI scan are important to establish the tumor extension, to plan treatment, and to allow radiologic and clinical follow-up. The treatment of optic nerve glioma is controversial[12]. Observation is generally recommended if there is no clinical or radiographic evidence of progression of an optic nerve glioma. [13] Surgical resection is successful in tumors confined to the optic nerve, with no useful vision or progression. The preferred technique is transcranial superior orbitotomy with preservation of the globe. [14]

Radiation therapy is generally reserved for patients over 5 years of age with progressive radiographic features or worsening clinical signs and symptoms[9].Chemotherapy is emerging as a possibly safer alternative to radiation therapy particularly in younger children. [10]

The prognosis of optic nerve glioma is quite variable, most (80%) have stable vision after an initial period of visual loss. The 10 year overall survival rate is between 85% to 100% in various series and spontaneous regression may occur. [1,16]

REFERENCES

1. Liu GT, Lessell S. Spontaneous visual improvement in chiasmal gliomas. Am J Ophthalmol, 1992; 114:193-201.
2. Brodovsky S, ten Hove MW, Pinkerton RM et al. An enhancing optic nerve lesion: malignant glioma of childhood. Can J Ophthalmol, 1997; 32:409-413.

3. Chutorian AM, Schwartz JF, Evans RA, et al: Optic gliomas in children. Neurology, 1964; 14: 83.
4. Janss AJ, Grundy R, Caan A et al. Optic pathway and hypothalamic/chiasmatic gliomas in children younger than age 5 years with 6 years follow-up. Cancer, 1995; 75:1051-1059.
5. Dutton JJ. Gliomas of the anterior visual pathway. Surv Ophthalmol, 1994; 38:427-452.
6. Porterfield JF. Orbital tumors in children: A case report on 214 cases. Int Ophthalmol Clin, 1962; 2:319
7. Alvord EC Jr, Lofton S: Gliomas of the optic nerve or chiasm: Outcome by patients' age, tumor site, and treatment. J Neurosurg, 1988; 68:85.
8. Trobe JD, Glaser JS: Quantitative perimetry incompressive optic neuropathy and optic neuritis.Arch Ophthalmol 1978; 96:1210.
9. Lee AG, Dutton J. A practice pathway for the management of gliomas of the anterior visual pathway: an update and an evidence-based approach. Neuro-Ophthalmology 1999; 22:139-155.
10. Hovland KR, Ellis PP: Hemorrhagic glaucoma with optic nerve glioma. Arch Ophthalmol 1966; 75:806
11. Dutton J. Optic nerve glioma. In: Dutton JJ, ed. Diagnostic Atlas of Orbital Diseases. Philadelphia: WB Saunders, 2000; 120-121.
12. Friedman JM, Birch P. An association between optic nerve glioma and other tumors of the central nervous system in neurofibromatosis type I. Neuropediatrics, 1997; 28: 131-132.
13. Imes RK, Hoyt WF. Childhood chiasmal gliomas: update on the fate of patients in the 1969 San Francisco study. Br J Ophthalmol, 1986; 70:179-182.
14. Housepian EM: Surgical treatment of unilateral optic nerve gliomas. J Neurosurg, 1969; 31: 604.
15. Fuss M, Hug EB, Schaefer RA et al. Proton radiation therapy (PRT) for pediatric optic pathway gliomas: comparison with 3D planned conventional photons and standard photon technique. International J Radiat Oncol Biol Phys, 1999; 45:1117-1126.
16. Parsa CF, Hoyt CS, Lesser RL et al. Spontaneous regression of optic gliomas. 13 cases documented by serial neuroimaging. Arch Ophthalmol, 2001;119: 516-529