Case Report

Optic Nerve Glioma – A Rare Case Report

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Abstract

An optic nerve glioma is a slow growing tumour which typically affects children. Malignant gliomas are rare and almost always occur in adult males with a very poor prognosis. It is a brain tumour that arises in or around the optic nerve, which connects the eye to the brain. We describe a case of a 5 year old boy with painless progressive enlargement of the left eye since the past 1 year. He also complained of a loss of vision in the left eye. The father had a history of Neurofibromatosis type 1. The child was clinically evaluated and required investigations were carried out. This was consistent with the diagnosis of optic nerve glioma. Early diagnosis is the key factor and an integrated approach should involve neurosurgeons, plastic surgeons, ophthalmologists and radiologists.

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Introduction

Optic nerve glioma (also known as optic pathway glioma) is the most common primary neoplasm of the optic nerve. Along with reducing visual acuity in the affected eye, the tumor sometimes produces additional symptoms as it grows. A low-grade form of this neoplasm, benign optic glioma, occurs most often in pediatric patients. Another form, aggressive glioma, is most common in adults; it is frequently fatal, even with treatment.[1,2,3]

Many children with optic nerve glioma are also known to have neurofibromatosis type 1 (NF-1) or, in some cases, hybrid phakomatosis.[4]

Case Report

We describe a case of a 5 year old boy with painless progressive enlargement of the left eye since the past 1 year. He also complained of a loss of vision in the left eye. The father had a history of Neurofibromatosis type 1. Presenting visual acuity was absence of light perception in the left eye and the proptosis was measured of 25mm which was in the downward and outward direction. A relative afferent pupillary defect was noticed in the left eye and extraocular movements were restricted superiorly and medially. Funduscopy of the left eye revealed a pale disc, blurring of the disc margins and presence of optociliary shunt vessels. CT scan of the left eye showed a fusiform optic nerve swelling. MRI showed a fairly large fusiform swelling in left optic nerve, pressure effect on ocular bulb with proptosis, no extension of the lesion in optic canal.

The patient was posted for lateral orbitotomy. Biopsy was taken from the same. It revealed low grade spindle shaped pilocytic (hair like) astrocytes & glial filaments, with the presence of numerous Rosenthal's fibers.

Discussion

Optic nerve glioma is a rare but important cause of proptosis in paediatric patients. Early diagnosis is mandatory in order to ensure a favourable prognosis.

Optic nerve glioma usually presents with slowly progressive visual loss, followed later by proptosis. There may be an acute loss of vision due to hemorrhage into the tumour. Proptosis is usually non- axial with temporal or inferior dystopia. The optic nerve head, initially swollen, subsequently becomes atrophic. Symptoms of an optic nerve glioma are caused by the tumor pressing against the nerves. Common symptoms of this kind of tumor include:

- nausea and vomiting
- balance problems
- vision disturbances
- headaches

Other symptoms can include:

- involuntary eye movements
- memory impairment
- daytime sleepiness
- loss of appetite
- growth delays

Hormone problems may also appear because the tumors can occur near the base of the brain where hormones are controlled.[7]

When the diagnosis is in question, the presence of an intraconal mass can often be detected through CT scanning. MRI, however, is the preferred method for definitive evaluation of optic nerve glioma. Both the intraorbital lesion and its intracranial extent can be effectively characterized through MRI. When evaluating the orbit, gadolinium-enhanced T1-weighted images with fat saturation can define the extent of aggressive glioma. Intracranially, MRI allows better evaluation of the optic nerve, chiasm, tracts, geniculate body, and optic radiations than does CT.[5]

Although MRI may reveal even subtle lesions of the optic nerve, CT scanning can detect a subtle erosion or expansion of the optic canal. In addition, fine calcification, which may help to identify a lesion as a meningioma rather than a glioma, is visualized best through CT scanning.

In children, unenhanced CT scans typically reveal a marked, diffuse enlargement of the optic nerve, with characteristic kinking or bending. The enlargement may be tubular, fusiform, or excrescent.[6] Areas of lucency may result from mucinous or cystic changes. Approximately 50% of the lesions demonstrate enhancement; this characteristic is

Figure 1: *The image of the boy with proptosis of the left eye*

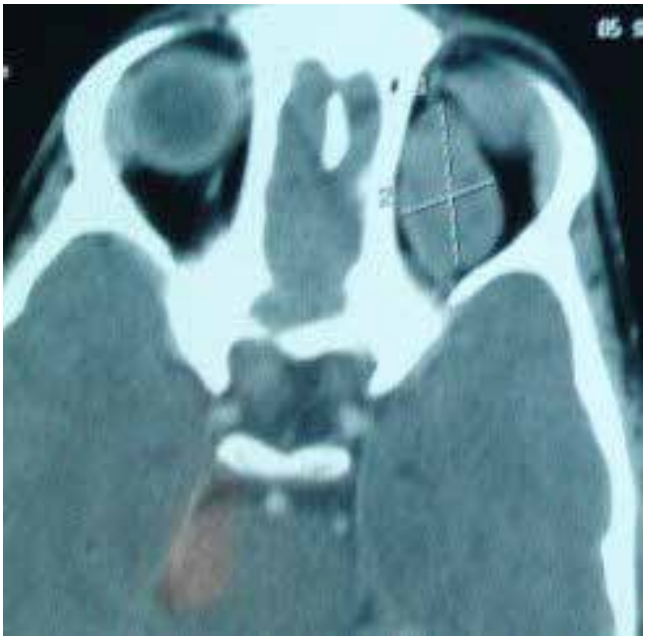

Figure 2: *CT Scan showing fusiform swelling of the optic nerve*

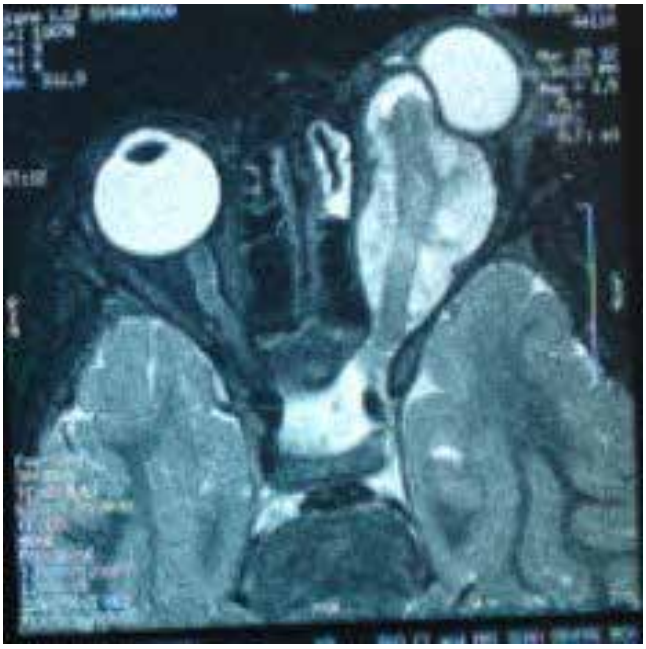

Figure 3: *MRI scan showing fairly large fusiform swelling in left optic nerve, pressure effect on ocular bulb, with proptosis, no extension of the lesion in optic canal*

more common with intracranial (especially retrochiasmatic) extension. Calcifications are rare.

The diagnosis may be made with a high degree of confidence when the lesion involves the optic chiasm and retrochiasmatic optic pathway. When confined exclusively to the orbit, the lesion may mimic optic neuritis or optic nerve meningioma; in this setting and in most instances, MRI is a better diagnostic tool than is CT scanning. CT scanning is the better modality for identifying uncommon meningioma with flecks of calcification, which are rare in optic nerve glioma.

On T1-weighted images, optic nerve gliomas are usually isointense to the cortex and hypointense to white matter. Invariably, the lesions are hypointense to orbital fat.

On T2-weighted images, lesions demonstrate a mixed appearance that is isointense to hyperintense relative to white matter and the cortex. Following contrast administration, intense enhancement is common.

Treatment options, depend on clinical context, as well as the location of a tumor at presentation. If it is isolated to one optic nerve and does not extend to the chiasm, then resection is curative (albeit with the loss of vision in that eye). If a tumor extends to the chiasm or more posteriorly, then curative resection is not possible, with resection reserved for treatment of mass effects (proptosis, intracranial mass effect).[7]

Treatment options include observation,surgical resection, high dose radiotherapy and chemotherapy.

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