

Optic nerve glioma presenting as non-axial proptosis

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Abstract

Optic nerve glioma is a relatively rare tumor of childhood. It usually present with axial proptosis. Non-axial proptosis is rare in such cases. We present a case of optic nerve glioma in a five year old child who presented with non-axial proptosis of right eye.

Keywords: Glioma, Optic Nerve, Proptosis

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] There is no gender predilection.^[4] Optic nerve glioma accounts for 0.6% to 1.2% of all intracranial tumors.^[5] The incidence is 1 in 100,000 patients, with 90% presenting within the first two decades and 70% in the first decade.^[4] 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. One or both optic nerves are involved in 24%, the optic disc in 1.6% and the optic chiasma or tract in 75%.^[6] In general, the more anterior the lesion, the better the prognosis.

Case Report

A five years old male patient presented with history of right eye squinting intermittently since two months and

forward displacement of right eye since one month, There were pigmented skin lesions present since birth. On ophthalmic evaluation, the best corrected vision in the right eye was finger counting at one meter with accurate projection of rays, and a vision of the left eye as 6/6. There was non-axial proptosis with exotropia in the right eye [Figure 1].



Figure 1: Showing exotropia in right eye

Corneal reflex test showed exodeviation of 15 degrees. The head posture was normal and the extraocular movements were restricted in all gazes. Direct ophthalmoscopy revealed disc edema. The skin showed multiple caf'e au lait spots larger than 5 mm in diameter [Figure 2]. The left eye was within normal limits. Systemic examination was within normal limits.



Figure 2: Showing caf'e au lait spots

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CT scan of the orbit revealed, 3.60x1.48 cm, heterogenous, hyperdense, mildly enhancing, fusiform, soft tissue lesion involving right optic nerve in retro-bulbar intraconal region on medial side in right orbit, without calcific focus, suggesting optic nerve glioma involving right optic nerve [Figure 3].



Figure 3: CT Scan : Showing fusiform, soft tissue density mass lesion involving right optic nerve in retro-bulbar intraconal region on medial side in right orbit

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. Except when optic nerve gliomas are discovered incidentally and vision is normal, the eye findings are typical of chronic optic nerve dysfunction. 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] NF-1 is a rare condition characterized by hamartomas of neural crest origin. It has an autosomal dominant inheritance although half the cases are new mutations. The incidence

of optic nerve glioma in children with NF-1 is as high as 15% to 20%, with symptomatic visual loss in approximately 20% of affected patients.^[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 extension of the tumor outside the optic nerve sheath. The overall 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. On CT scan, the tumor presents as an enlarged, fusiform, non-calcified optic nerve mass with frequent kinking and cystic areas.^[12] MRI scan is superior to CT scan for demonstrating intracranial extension.

Differential diagnosis of optic nerve glioma should include idiopathic optic neuritis, sarcoidosis, demyelinating disease and optic nerve sheath meningioma.^[13]

The treatment of optic nerve glioma is controversial.^[14] Observation is generally recommended if there is no clinical or radiographic evidence of progression of an optic nerve glioma.^[15] 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.^[16] 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.^[9]

The prognosis of optic nerve glioma is quite variable and is in part based upon location.^[17] 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, 18]

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