Case report of optic nerve glioma in a case of neurofibromatosis type-1.

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Abstract:

A 12 year old female patient presented with diminution of vision from childhood and more so since 1 week following fever, chills and vomiting. The patient was uncooperative for visual assessment. On slit lamp examination, anterior segments of both eyes were normal. Indirect ophthalmoscopy showed disc pallor in both eyes. The contrast enhanced computed tomography showed homogenously enhanced thickening of optic nerve predominantly in intracranial part also involving optic chiasma and associated tortuous intraorbital part of optic nerve. On magnetic resonance imaging both optic nerves, optic chiasma, and both optic tracts appeared enlarged, with widening of optic canals on both sides and subarachnoid spaces along both optic nerves appeared dilated. The patient was referred to neurosurgery department where intranasal trans-sphenoidal endoscopic biopsy from left optic nerve was taken. Histopathology examination was suggestive of low grade glioma. Neurofibromatosis type-1 is a rare condition with bilateral optic nerve glioma being a rare condition.

Keywords: Chemotherapy, Glioma, Neurofibromatosis, Orbital tumour.

Introduction:

Optic nerve glioma is a pilocytic astrocytoma and occurs primarily in the younger age groups (median age 6.5 years).[1]It is a slowly growing, low-grade neoplasm of the visual pathway that represents 2-5% of childhood brain tumors.[2] Children with neurofibromatosis type 1 (NF-1) are prone to the development of low-grade gliomas and experience significant visual loss. Majority of NF-1 associated gliomas arise in the optic nerves, chiasma, tracts, and radiations and affected children present with reduced visual acuity. The brain magnetic resonance imaging (MRI) is helpful for diagnosis of optic nerve glioma and to delineate the tumor extent.[3] Biopsy of optic nerve gliomain those of atypical locations carries a high risk of vision loss during the procedure. The management of optic nerve glioma is challenging and chemotherapy remains the mainstay of treatment which helps in the preservation of vision in the majority of children.[4, 5]Wepresent a case of a child presenting with NF-1 with bilateral optic

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nerve glioma which is a rare condition.

Case report:

A 12 year old female patient residing at Mehsana came with complaint of diminution of vision in both eyes since childhood which gradually increased following high-grade fever with chills and rigor associated with copious vomiting from last 1 week.

The patient was the youngest child of two children and had a history of her mother having Neurofibromatosis type-1.

There was no history of trauma, glasses, foreign body, previous eye disease or previous eye surgery.

On examination, the patient was uncooperative for visual assessment. Torch-light and slit lamp examination showed normal anterior segments in both the eyes. Eye movements were full and the intraocular pressures were normal. On dilated indirect fundoscopic examination, both eyes showed optic disc pallor.

Image 1: Contrast enhanced computed tomography of brain with orbit showing homogenously enhanced thickening of optic nerve on either side predominantly in intracranial part.

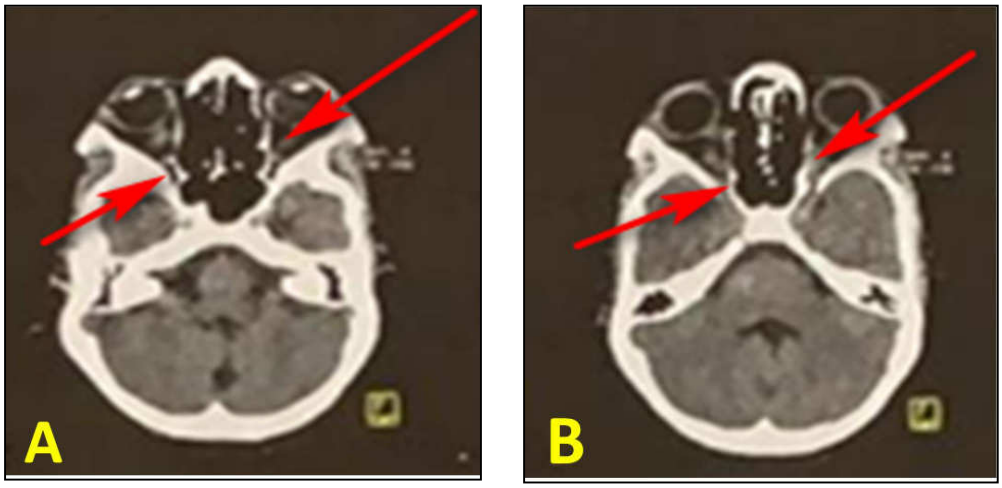
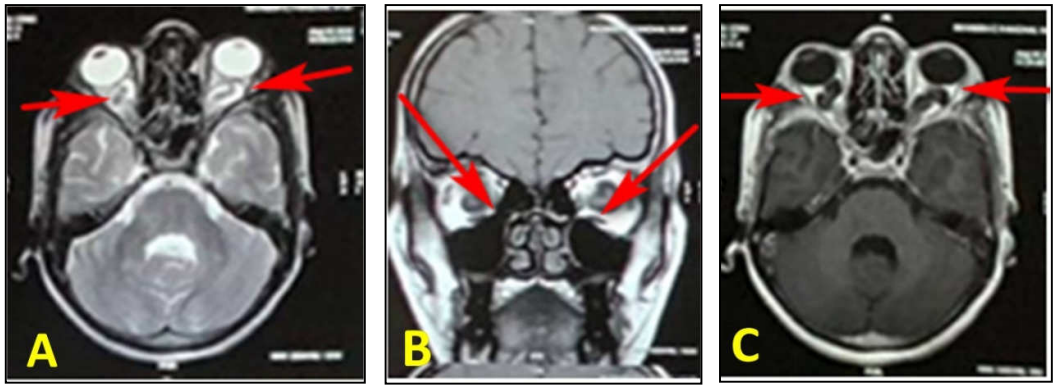


Image 2a,2b,2c: Magnetic resonance imaging(MRI) of brain with orbit showing enlarged optic nerves, optic chiasma and optic tracts on both side and widening of optic canals.



Contrast enhanced computed tomography (CECT) scan of brain with orbit showed homogenously enhanced thickening of optic nerve on either side predominantly in

intracranial part, involving optic chiasma which was suspicious of optic nerve glioma(?NF-1). Associated tortuous intraorbital parts of optic nerves were also seen (Image 1)

Magnetic resonance imaging (MRI) of brain with orbits with contrast showed both optic nerves, optic chiasma and both optic tracts to be enlarged. Lesion was noted extending into both thalamo-capsular regions (right more than left) with perilesional vasogenic oedema surrounding both thalamo-capsular and medial temporal regions; widening of optic canals was noted on both sides; subarachnoid spaces along both optic nerves appeared dilated; with the findings suggestive of optic pathway glioma (Image 2).

Then patient was referred to neurosurgery department where a transnasal transsphenoidal endoscopic biopsy was taken whose histopathological examination showed low grade glioma and patient was advised chemotherapy.

Discussion:

Carlos E. Prada, Robert B. Hufnagel, et al. noted that chiasmatic and postchiasmatic optic pathway glioma in children with NF-1 had the highest risk of progression and vision loss. Early identification of optic pathway glioma by MRI screening prior to the development of vision loss may lead to improved visual outcomes.[6] But as our patient presented late in the second decade of life, she had a poor visual outcome.

De Blank PMK, et al. noted that the indications for chemotherapy were based on a comprehensive, quantitative assessment of vision and the goal was to preserve vision in NF-1 associated optic pathway glioma, but vision loss in such patients can be unpredictable and the ability to identify and predict impending vision loss could improve management decisions and visual outcomes.[7]Hence as our patient presented with established visual deficit, the visual outcome was unpredictable even after the biopsy and chemotherapy.

Marta Campagna, Enrico Opocher, Elisabetta Viscardi, et al noted that children with optic pathway glioma had unsatisfactory visual prognosis and younger children treated with radiotherapy had poor visual outcome than older children and the indication for poor prognosis was the severe optic pallor at diagnosis or during follow-up.[8] Our patient also had severe optic pallor at diagnosis and an unsatisfactory visual prognosis..

Michael J Wan noticed that approximately 50% of optic pathway gliomas arose in NF-1 patients and there was a significant risk of visual impairment in these patients but the overall survival rate was high for optic pathway gliomas. The definite indication for urgent treatment was the combination of functional deterioration and tumor enlargement and the first-line treatment for children with optic pathway glioma was the chemotherapy. The effective treatment to control newly diagnosed, progressive low-grade gliomas in children was weekly carboplatin and vincristine and had improved visual outcomes in optic pathway glioma patients associated with neurofibromatosis type-1.[9, 10]Our patient with NF-1 also had been advised chemotherapy as a first-line treatment for her optic pathway gliomas.

Cynthia J. Campen and David H. Gutmann had considered chemotherapy as a first-line treatment of NF-1 optic nerve glioma, especially when clinical progression occurred. Due to the unusual locations of these tumors, diagnostic biopsy was done and complete surgical resection was impossible. Due to the risk of secondary tumors (glioma and MPNSTs)

in children with NF-1, radiation was not recommended.[11]Our patient was also advised chemotherapy after confirmation of diagnosis on biopsy.

Conclusion:

NF-1 is a rare disease further in which bilateral optic nerve glioma is rarely seen. Visual prognosis is poor in such patients and chemotherapy should be considered as a first-line therapy.

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