Challenges in Diagnosis and Management of Optic Nerve Glioma: A Case Report

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Introduction: Optic nerve glioma is a slowly growing, pilocytic astrocytoma that typically affects children (median age 6.5 years)[1]. About 30% of patients have associated Neurofibromatosis type 1 and those have better prognosis.[2]

Case Report: A 13yr old boy presented with a gradual, painless visual loss in the left eye with associated bulging of the eyeball which started about 7 years before presentation. Surgery was done at a peripheral hospital (nature of surgery unknown) 1 year after onset of symptoms, however tumor regrowth over the past 6 years was noticed. Examination revealed a large ulcerated left orbital mass measuring 8x7x5cm, firm, not tender, the mass was not attached to the orbital rim or skin, no dilated vessels, no bruit, and no eyeball seen. Right Eye had a visual acuity of 6/24 (unaided) and 6/18 with pinhole, normal anterior segment but pale disc, CDR=0.3, flat retina. Magnetic resonance imaging (MRI) done 3 months before presentation showed a well circumscribed lesion of mixed consistency at suprasella region compressing the left ventricle with extension into left orbit. A diagnosis of optic pathway (optic nerve & optic chiasma) glioma was made. Orbital and intracranial tumour excision was done in conjunction with neurosurgeons. Histology revealed a pilocytic astrocytoma.

Fig. 1: Optic nerve glioma (a) Left Orbital tumour protrusion (b) MRI showing a large tumour of mixed consistency compressing left ventricle and extending into left orbit

Discussion: Optic pathway gliomas represent approximately 3-5% of childhood intracranial tumors[3], the chiasm is involved in roughly half of cases of optic nerve glioma. Intracranial involvement may be associated with intracranial hypertension. The diagnosis is made by clinical findings with associated typical radiological findings especially in those with neurofibromatosis type 1[4,5] but in some cases where MRI findings are not conclusive, histological diagnosis is essential[6,7]. The available treatment modalities for optic pathway gliomas include radiotherapy, chemotherapy and surgery, however there is significant controversy regarding the optimal management and outcome of these patients[8] The prognosis is worse for those with intracranial extensions and malignant gliomas[9] Challenges in management of the patient include delay in presentation, uncertainty about the nature and extent of the first surgery which was further worsened by lack of histological examination after that surgery and lack of a more recent MRI to ascertain the extent of intracranial extension and damage to adjacent structures as of the time of the second surgery due to financial constraint.

Conclusion: Tertiary centres may be faced with multiple challenges in the management of patients with optic pathway gliomas. It is often necessary to involve multidisciplinary specialists in the management of such cases.

References

1. Adel Alsuhaibani MD, A. H. A. N. Optic Nerve Glioma - EyeWiki. (2016). Available at: http://eyewiki.aao.org/Optic_Nerve_Glioma. (Accessed: 1st October 2017)
2. Jack, Kanski Brad, B. Clinical Opthalmology: A systemic approach. (Elsevier Saunders;).
3. Binning, M. J., Liu, J. K., Kestle, J. R. W., Brockmeyer, D. L. & Walker, M. L. Optic pathway gliomas: a review. Neurosurg. Focus 2007; 23, E2.
4. Grill, J. et al. When do children with optic pathway tumours need treatment? An oncological perspective in 106 patients treated in a single centre. Eur. J. Pediatr. 2000; 159, 692–696.
5. Leonard, J. R. *et al.* The role of surgical biopsy in the diagnosis of glioma in

individuals with neurofibromatosis-1. Neurology 2006; 67, 1509–1512.
6. Sutton, L. N. *et al.* Long-term outcome of hypothalamic/chiasmatic astrocytomas in children treated with conservative surgery. J. Neurosurg. 1995; 83, 583–589.
7. Fouladi, M. et al. Survival and functional outcome of children with hypothalamic/ chiasmatic tumors. Cancer 2003; 97, 1084–1092.
8. El Beltagy, M. A. *et al.* Treatment and Outcome in 65 Children with Optic Pathway Gliomas. World Neurosurg. 2016; 89, 525–534.
9. Kaiser, P. K. Prospective evaluation of visual acuity assessment: a comparison of Snellen versus ETDRS charts in clinical practice (An AOS Thesis). Trans. Am. Ophthalmol. Soc. 2009; 107, 311–24.