ORIGINAL RESEARCH PAPER

Radio-Diagnosis

A RARE CASE OF OPTIC PATHWAY GLIOMA PRESENTING WITH LIMB WEAKNESS

KEY WORDS: Optic pathway glioma, limb weakness, low grade astrocytoma with piloid differentiation – WHO grade II

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ABSTRA

Optic pathway gliomas(OPG) are low-grade neoplasms arising from the pre-cortical optic pathways. OPG can involve the optic nerve, optic chiasm, optic tracts, optic radiations, or the hypothalamus. Most commonly affecting children, they present usually with headache and vision related complaints . In this, we describe the a rare case of optic glioma presenting with limb weakness and role of MRI in it's diagnosis

INTRODUCTION:

Optic pathway gliomas are low-grade neoplasms arising from the pre-cortical optic pathways involving the optic nerve , optic chiasm, optic tracts, optic radiations, or the hypothalamus.These neoplasms may arise sporadically or in association with neurofibromatosis type 1 (NF1).[1] When arising sporadically, the most common genetic alteration identified is a BRAF-KIAA1549 fusion.[2]

Optic pathway gliomas most commonly affect children under ten years old and account for 3 to 5% of childhood central nervous system tumors.[3] However, gliomas have been found in patients ranging from birth to 79 years of age.

Histopathology, Gliomas arise from astrocytes of the optic nerve and visual pathway-usually WHO grade I pilocytic astrocytoma with immature astrocytes.

The location of the tumor determines the presenting symptoms and signs. The most common presenting symptom of OPG is headache . Overall, 85% of patients with glioma will lose some vision, and over time , approximately 25 % will retain vision between 20 /20 and 20 / 40. Patients may have an afferent pupillary defect and visual field defects. Optic nerve atrophy and optic d isc edema may be seen.

Most common presenting sign is Proptosis seen in 95% of patients.
Other symptoms and signs include limitation of ocular motility, nystagmus, spasmus nutans, convulsions, nausea, dizziness, strabismus, developmental regression, and growth retardation . Hydrocephalus may develop when t he t umor spreads from the chiasm. Hypothalamic-pituitary dysfunction with precocious puberty , growth hormone deficiency, and deficiency of gonadotropin, TSH , and ACTH may also be seen.

Initial Work-up includes magnetic resonance imaging (MRI) of the brain and orbit which is almost diagnostic because of pathognomonic features.

On MRI, gliomas will demonstrate slightly prolonged T1 relaxation times , and the image of the tumor will be isointense or slightly hypointense compared to the normal o ptic nerve . T2 - weighted images will show a hyperintense image with prolonged T2 relaxation time . Enlargement of the optic canal will be seen in 80% of patients where the optic nerve is involved . In 25% of patients with chiasmal glioma, sella turcica enlargement and J-shaped excavation may be found [4]

CT also shows enlargement of the optic nerve and/or chiasm. Mild enhancement may be seen on contrast study. The optic nerve will show a fusiform swelling, but rounded and exophytic changes may also be seen. Calcification in optic pathway gliomas is uncommon but may be seen. [4]

A biopsy is typically unnecessary because the diagnosis can be made based on imaging and clinical examination.

Close differential to be considered is Optic Nerve Meningiomas . Both optic nerve gliomas and meningiomas can show a diffuse , globular or fusiform enlargement of the optic nerve. However, the thickened and denser optic nerve sheath in meningiomas will result in tram track appearance with a central lucency , which is the residual optic nerve . Calcification is seen in 20 to 50% of meningiomas but very rarely in gliomas . Optic nerve gliomas will show an isointense or slightly hypointense signal on T1 but hyperintense on T2 sequences whereas meningiomas will appear hyperintense compared to normal nerve on both T1- and T2 -weighted sequences.

Another differentials of sellar and suprasellar mass includes pituitary macroadenoma , craniopharyngioma which has typical feature of calcification and a prominently cystic component or signal intensity of mixed cystic and solid components in most patients. [5]
Rathke's cleft cyst, originating from pars intermedia , typically shows midline anterior infundibular displacement. [5]

The management of gliomas depends on the extent of involvement and clinical presentation- ranging from simply observing clinically and radiologically to chemotherapy, radiotherapy or combination of both to surgery. Surgery should be limited to obtaining a biopsy for diagnostic purposes , or to resect tumors when they cause excessive proptosis and pain or when they extend posteriorly, threatening involvement of the optic chiasm.

CASE STUDY:

A 3 year old male patient presented with complaint of headache since 6 months and left sided weakness since 2 months.

MRI showed lesion in suprasellar and right parasellar mass lesion involving hypothalamic region and extending along right optic tract.

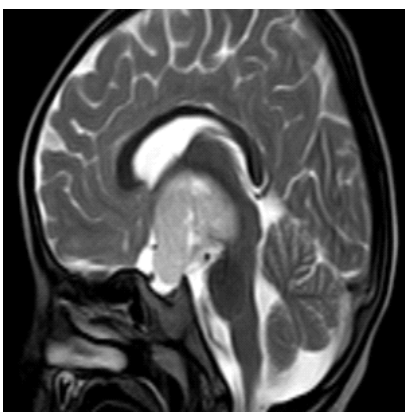
The lesion appears hypointense on T1WI and hyperintense on T2WI with heterogenous post contrast enhancement.

The lesion causes mass effect in the form of compression over medial temporal lobe and compression of temporal horn of right lateral ventricle and third ventricle and midline shift of approximately 3 mm towards left side.

The lesion displaces right midbrain region inferomedially and posteriorly and superior displacement of right thalamus.

Pituitary gland is seen separately from the lesion.
Optic chiasma could not be seen separately from the lesion - possibility of hypothalamic – optochiasmatic glioma with extension along right optic nerve.

Figure 1: Sagittal and coronal T2WI shows hyperintense lesion in suprasellar and right parasellar region



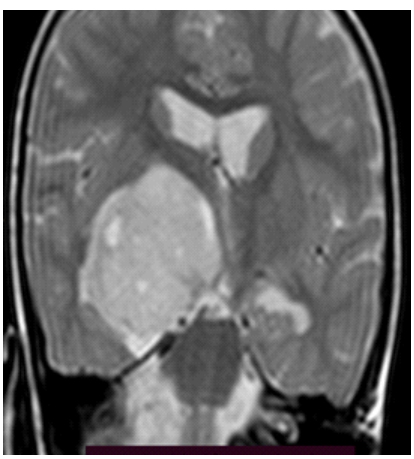
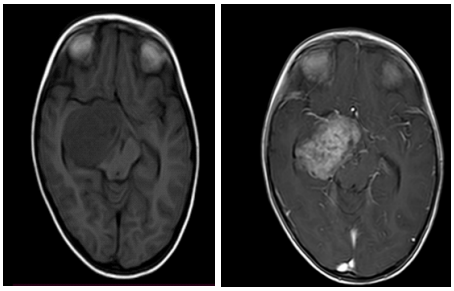


Figure 2: Axial T1WI (left) show hypointense lesion and postcontast image (right) shows heterogenous postcontrast enhancement.

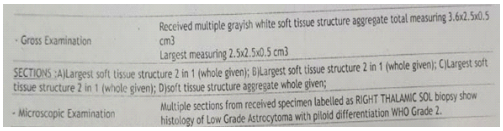
Lesion shows mass effect in the form of compression over medial temporal lobe and right midbrain region.



Due to complain of limb weakness and location of tumor as described on MRI , patient underwent surgical resection of the tumor.

Figure 3: The histological examination shows a glial neoplasm composed of a monomorphic population of cells with oval nuclei and elongated cytoplasmic processes in a myxoid background. No mitotic figures or atypical cells were found . There was no evidence of Rosenthal fibers or eosinophilic granular bodies —

Above findings suggested low grade astrocytoma with piloid differentiation –WHO grade II



CONCLUSION:

MRI has a very crucial role in diagnosis of optic pathway glioma based on the specific appearance of the lesion and also in deciding the further management based on the extent of involvement.

REFERENCES

1. https://pubmed.ncbi.nlm.nih.gov/28847625/
2. https://pubmed.ncbi.nlm.nih.gov/23624918/
3. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6169473/
4. https://www.ncbi.nlm.nih.gov/books/NBK557878/
5. https://www.ajronline.org/doi/pdfplus/10.2214/ajr.167.3.8751659?src=recsys