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Multicentric Malignant Optic Nerve Glioma-A Rare and Challenging Diagnosis

Case Report

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Introduction

Optic nerve gliomas are predominantly benign tumors, commonly arising in children and young adults. Malignant transformation is a rare occurrence in optic nerve gliomas, and multicentric involvement is even rarer. Herein, we present a case of multicentric malignant optic nerve glioma to increase awareness of this uncommon variant and contribute to the existing literature on optic nerve gliomas.

Case Presentation

A 44-year-old male presented with a 3-month history of progressive bilateral visual loss. The patient complained of reduced visual acuity, visual field defects in both eyes and color vision impairment. On examination, afferent pupillary defects were noted in both eyes. Visual field analyser revealed bitemporal hemianopia. Fundus examination showed no evidence of disc edema. Visual acquity at the time of presentation was 6/6. MRI revealed thickening of the optic chiasma with altered T2/FLAIR hyperintense signal and early post contrast enhancement (Figure 1).

However, contour of optic chiasma was preserved. Pituitary stalk and tuber cinerium were also thickened showing altered signal intensity with intense post contrast enhancement. The lesions demonstrated infiltrative growth patterns, resulting in fusiform thickening of the intracranial parts of B/L optic nerves with mild post contrast enhancement. Altered signal intensity was also seen involving post chiasmatic left optic tract in the region of left temporal lobe, left lateral geniculate body in the posterior part of thalamus with involvement left hypothalamus, left hippocampus and left insular cortex. No evidence of any hemorrhage/calcification/necrosis was seen within it (Figure 2 and Figure 3).

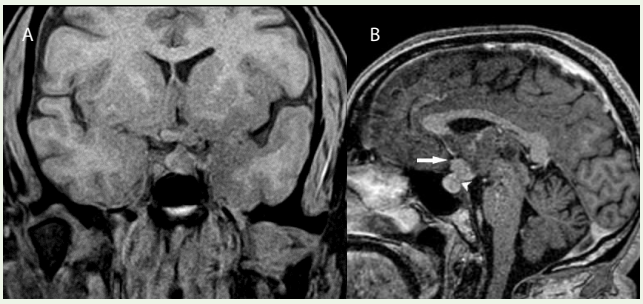

Figure 1: Coronal(A) and sagittal (B) T1W image shows thickened optic chiasma (arrow) with thickened pituitary stalk (arrowhead).

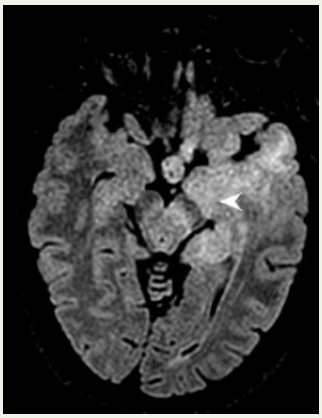

Figure 2: Axial FLAIR image shows altered signal intensity in the region of post chiasmatic left optic tract in the region of left temporal lobe,left hypothalamus and left hippocampus.

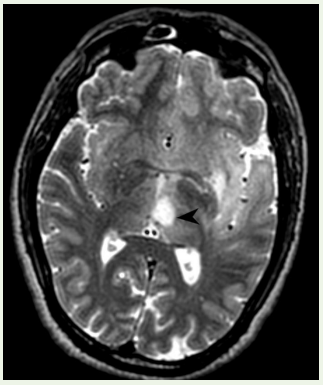

Figure 3: Axial T2W image shows a well-defined lesion in left thalamus (arrowhead). On post contrast images, no enhancement was seen. This lesion was described as a multicentric low grade glioma.

Another well-defined lesion appearing hyperintense on T2W/FLAIR images, hypointense on TlW images, measuring approximately 1.2 x 2 cm was noted in left thalamus suggesting the multicentricity of tumour. MRI with gadolinium contrast showed enhancement in optic chiasma, pituitary stalk, tuber cinerium and intracranial parts of bilateral optic nerves. Diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) maps showed restricted diffusion in the lesions, indicating high cellularity.

On the basis of MRI, differential considerations included bilateral optic neuritis or multifocal hypothalamic chiasmatic optic nerve glioma or metastatic disease. Rare possibility of lymphoma was also considered after reviewing literature which seemed unlikely due to the pattern of involvement.

The radiological findings were consistent with the diagnosis of multicentric malignant optic nerve glioma.

Surgical interventions, histopathological examination of the optic nerve lesions are confirmatory in such cases. However, both of them could not be performed due to multicentric and infiltrative nature of the lesion making it a risky option when the risk versus benefit ratio was outweighed.

Due to the multicentric nature of the tumour and involvement of both optic nerves, complete surgical resection was not feasible. Poor prognosis was explained to the patient. The patient refused to undergo radiotherapy. Regular ophthalmological follow-up was scheduled to monitor visual acuity and optic nerve function.

Discussion

Optic nerve gliomas are relatively benign when they occur in childhood. Malignant gliomas of optic nerve are more common in adulthood.The more common benign optic nerve glioma is considered a low- grade astrocytoma and is frequently associated with neurofibromatosis type [1]. Malignant optic glioma is a very rare optic pathway tumor of the adulthood [2].

Optic nerve sheath is spared in optic neuritis and optic nerve ischemia. In these conditions, there is diffuse enlargement of the optic nerve. In conditions such as meningioma and pseudotumour, the optic nerve is spared whereas the optic nerve sheath is enlarged.[3,4] Those processes that involve both the optic nerve and sheath would be favoured in the differential diagnosis of the lesion in this case.

Due to the extreme rarity and very rapid disease progression, the point of origin cannot be certainly defined.[5] Secondary invasion of the hypothalamus causes symptoms like polyuria and polydipsia. [5,6] One report with computed tomography findings described a case of malignant optic glioma of adulthood that extensively enlarged the optic nerve but not the optic chiasm [7].

A syndrome of Optic nerve glioma in adults has been described by Hoyt et al which is very similar to the case presented by us here [6].

In such cases, clinical examination does not favour optic nerve or intraorbital mass, rather due to optic nerve involvement, the diagnosis of optic neuritis is made and intravenous steroids are initiated. And if there is clinical improvement after initiation of steroids, the diagnosis is further stressed. But due to the rapid progression of the mass, there is rapid deteroiration in visual acuity which leads to unilateral blindness in rapid succession. Chiasmal involvement occurs in rapid progression suggested by bilateral reduced visual acuity and bilateral optic disk edema. The mass further progresses and invades the hypothalamus, basal ganglia, and internal capsule. Hypothalamic symptoms usually occur late in the clinical course. [5] There can also be lepto-meningeal and subpial spread of malignant optic glioma to the medial temporal lobes and brain stem.[7]The current case demonstrates spread of malignant optic glioma of adulthood to intracranial bilateral optic nerves, optic chiasma, pituitary stalk and post chiasmatic left optic tract.The multicentricity of the tumour was diagnosed as a lesion was seen in left thalamus. No other similar case has been found in literature.

Conclusion

Multicentric malignant optic nerve glioma is an extremely rare variant of optic nerve glioma, with limited published reports. Its diagnosis is challenging due to its rarity, variable clinical presentation, and radiological features overlapping with other optic neuropathies. The role of MR imaging plays a pivotal role in the early diagnosis of this disease.

Treatment involves a multimodal approach, including surgical resection, radiation therapy, and chemotherapy, tailored to individual patients.

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