Suprasellar Ganglioglioma: Report of a Rare Case

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Gangliogliomas are rare central nervous system tumors composed of intimately admixed neuronal and glial components. Gangliogliomas are intra-axial masses located predominantly in the temporal lobe, but they can also arise from frontal, parietal and occipital region, and rarer sites include the cerebellum, brainstem and spinal cord. We report a case of a 22 years old female who presented with severe headache, bilateral blurring of vision, generalized weakness and history of convulsion. The lesion was radiologically indistinguishable from meningioma. Histologically the tumor was diagnosed as ganglioglioma by the presence of dual population of neoplastic ganglionic and glial components.


Key words: Ganglioglioma, Sellar-suprasellar tumor.

Introduction

Gangliogliomas are rare central nervous system tumors composed of intimately admixed neuronal and glial components. Their incidences range from 5-8% of all brain tumors, but they are more common in the pediatric age group.1,2 Seizures are the commonest presentation reflecting involvement of temporal lobes commonly, but they can involve any part of the neuraxis including spinal cord.2 Sellar or suprasellar gangliogliomas are very rare. We report a case of suprasellar Ganglioglioma with histopathological and MRI features. This case suggests that though Ganglioglioma is rare but can occur in this location.

Case Report

A twenty two years old female was admitted in the department of Neurosurgery of National Institute of Neurosciences and Hospital (NINS & H) with the complains of severe headache and vomiting for 25 days. She had history of loss of consciousness and convulsion and difficulty in walking associated with generalized weakness. She also complained of blurring of vision. On examination her Glasgo Comma Scale was 15/15. Higher psychic function and speech were normal. There was no sign of meningeal irritation. Bilateral blurring of vision was present.

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Her Magnetic Resonance Imaging (MRI) revealed a fairly large, lobulated extra axial lesion of (3.22 x 2.34) cm with signal intensity change, in the suprasellar and right parasellar region. It was iso to hypo-intense on T1 and mildly hyperintese on T2. Post contrast scan revealed intense enhancement of the lesion. It extends upwards through the floor of the third ventricle causing ventricular obstruction (Fig.1 and 2). Radiology suggested the lesion as a suprasellar meningioma. All the pituitary hormone levels were within the normal limit. A clinical diagnosis of craniopharyngioma was made.

The patient had a left sided V-P shunt (Fig.3) inserted two months back to relieve hydrocephalus at an outside hospital. Her previous ultrasonogram (USG) report of whole abdomen was normal. Recent USG showed a large thin walled cystic lesion in left lobe of liver suggesting CSFoma. Lower end of VP shunt tube was within the lesion. She was anaemic and had high ESR. Her serum amylase level was within normal limit and gastric biopsy revealed features of chronic gastritis.

The patient underwent right frontal craniotomy with subtotal removal of the tumor. The tumor was encapsulated, lobulated, firm, gritty and haemorrhagic. Peroperative findings suggested it to be a suprasellar meningioma. Part of the specimen was sent for routine histopathological examination.

Microscopic examination of the tumor showed collections of large ganglion cells with prominent nucleoli, abundant cytoplasm admixed with glial component. The glial component was mostly pilocytic in nature with some astrocytes in a fibrillary background. No evidence of meningioma or craniopharyngioma was seen. Finally it was diagnosed as Ganglioglioma, WHO grade I/IV.

After 18 days of surgery, the patient became quite healthy and symptom free. Her vision improved. She got discharged from hospital with almost full recovery.
Discussion

Temporal lobes are the commonest location for supratentorial gangliogliomas, but they can also arise from frontal, parietal and occipital region, and rarer sites include the cerebellum and brainstem. Sellar and suprasellar gangliogliomas are very rare and a few cases have been reported.2,3,4,5

The differential diagnoses of suprasellar lesions in children include neoplastic conditions like hypothalamic glioma, craniopharyngioma, germ cell tumors and pituitary adenomas; or non-neoplastic conditions such as granulomatous diseases and benign cyst. The most common lesions in adults are meningioma and pituitary adenomas3,4. Our case was also diagnosed radiologically as meningioma due to suprasellar location. All the pituitary hormone levels were done to exclude the possibility of pituitary adenoma. The hormone levels were within the normal limit and finally preoperative diagnosis was Craniopharyngioma, which is a common tumor in this location.

The radiographic appearance of gangliogliomas is variable, but certain characteristics prevail. It may be solid or cystic. The cystic appearance varies from a single large cyst with a mural nodule to a multicystic mass. Imaging studies reveal a well-circumscribed lesion situated in the peripheral cortex. On MRI, it is iso-to-hypointense on T1 weighted images, hyperintense on T2 weighted and FLAIR images and shows variable contrast enhancement either a nodular rim or a solid pattern. Calcification is common. The cyst margins can enhance, mimicking the ring enhancement of malignant glioma.3,4,6 The present case has similar radiological findings, but calcification and cyst formation are absent. The pathologic criteria of ganglioglioma includes irregular groups of large, dysplastic, multipolar neurons admixed with glial...
component surrounded by a reticulin network. The glial component is generally pilocytic or fibrillary, but ependymal and even oligodendroglial components have been described. Eosinophilic granular bodies, hyaline bodies, microcystic changes, calcification, desmoplasia and perivascular lymphocyte infiltration may be present variably. 2, 8

The case under discussion presented with a suprasellar lesion having complaints of headache, bilateral blurring of vision and history of convulsion. 15-25% patient of ganglioglioma undergoing surgery usually presents with a history of seizure. 7 Histopathology of our case showed combination of both neuronal and glial cell elements. The ganglion cells were large having vesicular nuclei, prominent nucleoli and abundant cytoplasm arranged in sheets and groups. The glial elements showed mostly pilocytes, scattered astrocytes and occasional foci of reticulin formation in a fibrillary background. Immunohistochemistry for CD34 antigen expressed in neuronal cells in 70-80% of gangliogliomas. 7 But as this case showed very typical and prominent ganglion cell component in a large area of the tumor, immunohistochemical analysis was not necessary. Jalali R et al., Siddique K et al., Shuangshoti S et al. also found ganglioglioma in sellar-suprasellar location which were radiologically diagnosed as other common entities of this site. 3, 4, 5

Conclusion
Gangliogliomas are mostly benign tumors with good prognosis. A complete surgical resection is necessary for recurrence free survival of the patient. But in most of the cases the sellar gangliogliomas are misdiagnosed both clinically and radiologically. A well-demarcated lesion with signal intensity changes in the sellar-suprasellar region should alert the clinician in considering gangglioma as a possible differential diagnosis, which will help in proper surgical management of the patient.

References