CASE REPORT

Lateral ventricular gliosarcoma with attachment to septum pellucidum

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ABSTRACT

Gliosarcomas are lesions with both glial and sarcomatous elements and are usually seen abutting the dura. Intraventricular location is extremely rare. Such an unusual intraventricular gliosarcoma is being reported.

Key words: Gliosarcoma, intraventricular lesion, septum pellucidum

Introduction

Gliosarcomas are rare malignant lesions of the brain and resemble glioblastomas in their clinico-pathological profiles, existing treatment options and dismal prognosis. Gliosarcomas are characterized histologically by the presence of both glial and sarcomatous elements and is believed to be arising from a common precursor stem cell. These tumors are usually located peripherally, and intraventricular gliosarcomas are extremely rare condition.[1-4] Here, we present a rare case of gliosarcoma located in bilateral lateral ventricles with attachment to the septum pellucidum.

Case Report

A 28-year-old male presented with raised intracranial pressure for two months. Neurological examination of the patient revealed bilateral papilledema. However, there were no focal deficits. MR of the patient revealed T1 hypointense, T2 hyperintense intraventricular lesion filling the bilateral lateral ventricle with avid contrast uptake and central non-enhancing area [Figure 1a-e]. There was a non-enhancing area in the right frontal lobe contiguous with the intraventricular lesion as apparent on T2 and FLAIR images [Figure 1f]. Patient underwent a parasagittal craniotomy and near total excision of the tumor. The tumor was pinkish gray, very vascular and had areas of necrosis along with thrombosed vessels within. Septum pellucidum was involved and was resected partly along with the tumor. The floor of both lateral ventricles could be appreciated after resection. The right frontal lobe appeared normal intraoperatively. Histopathological evaluation of the resected specimen revealed features of a high-grade tumor comprising of biphasic population of malignant cells. Tumor cells were highly pleomorphic comprising of malignant glial cells including many bizarre tumor giant cells intermixed with bundles of malignant spindle cells. Mitosis was brisk. There was intense endothelial cell proliferation and large areas of palisaded necrosis. Tumor cells showed immunoreactivity for glial fibrillary acidic protein (GFAP) and vimentin [Figures 2a-c]. The patient had a prolonged post-op course due to venous edema which gradually resolved, and he has been subjected to radiotherapy.

Discussion

Lesions of the body of lateral ventricle are commonly attached to the septum pellucidum, and differential diagnosis varies from benign lesion like neurocytoma, subependymomas and meningiomas to more malignant high-grade gliomas. These lesions may erode the septum pellucidum to grow into both lateral ventricles or may grow into one ventricle pushing the septum to the other side,

Access this article online	
Quick Response Code:	Website: www.asianjns.org
	DOI: 10.4103/1793-5482.145090

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How to cite this article: Salunke P, Singh H, Vaiphei K. Lateral ventricular gliosarcoma with attachment to septum pellucidum. Asian J Neurosurg 2017;12:82-4.

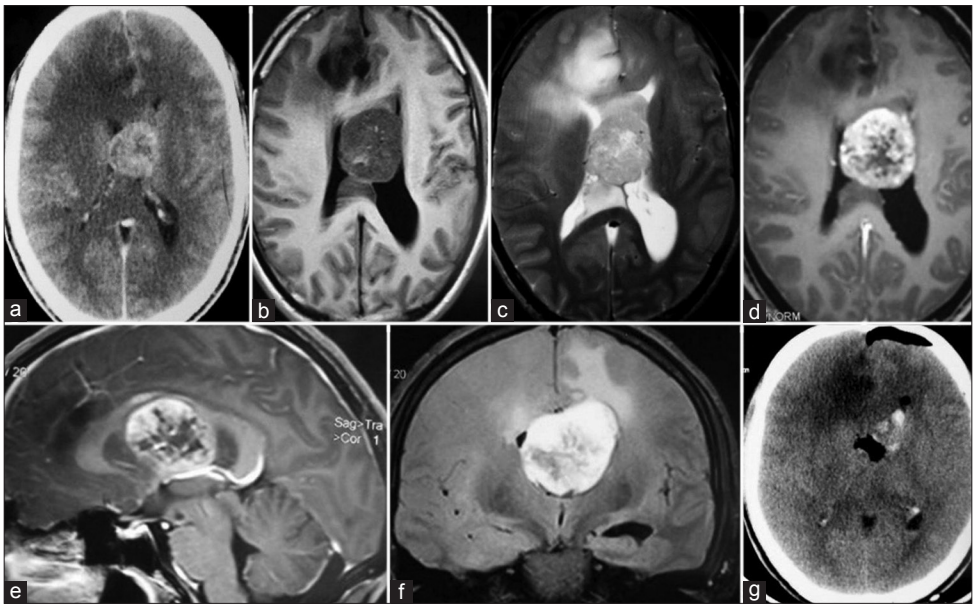

Figure 1: (a) Contrast enhanced computed tomogram head showing a well-defined hyperdense intraventriuclar lesion. (b-d) MRI axial images showing well-defined T1 hypo, T2 hyperintense lesion occupying the body of both lateral ventricles well enhancing with central hypointensity and another ill-defined lesion in the right frontal lobe. (e) Sagittal MRI showing the lesion pushing the body corpus callosum superiorly. (f) FLAIR coronal image showing hyperintense lesion. (g) Postoperative Non contrast computed tomogram showing total excision

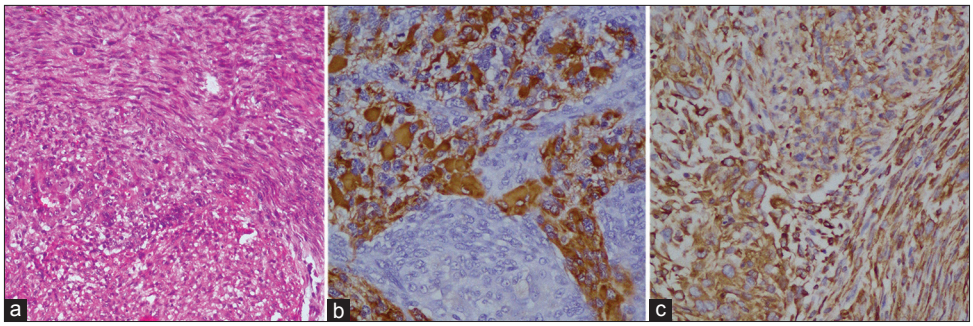

Figure 2: (a) Photomicrograph of the tumor showing distinctly two populations of cells, one of a high-grade glial cell types showing marked nuclear pleomorphism and multinucleated tumor giant cells merging with bundles of malignant spindle-shaped cells (H and E, ×250). (b) Photomicrograph showing selective cells stained positive for glial fibrillary acidic protein (PAP, ×450). (c) Photomicrograph showing vimentin positivity by both types of tumor cells i.e., spindle-shaped cells and malignant glial cells with bizarre nuclei (PAP, ×450)

masquerading as a biventricular lesion. The clinical course usually decides the approach irrespective of the pathology of the tumor. Gliosarcomas are well defined and can have features of both intra- and extra-axial lesions. These tumors are commonly seen as peripheral masses abutting the meninges with dense dural adhesion, exhibiting intense dural enhancement on contrast imaging uneven thick-walled rim or ring enhancement. There can also be an intra-tumoral strip enhancement.[2]

Intraventricular gliosarcomas are extremely rare and aggressive tumors. In spite the aggressiveness, these tumors are usually well defined that aids in complete resection. However, metastasis is more often seen with gliosarcomas then glioblastomas, possibly because of the propensity of the sarcomatous element to disseminate hematogenously. Trans ependymal spread is known giving rise to ependymal enhancement in imaging.[4] However, the present indexed patient did not have any ependymal enhancement in radiology and the post-operative craniospinal screening was negative for any tumor spread.

Origin of intraventricular gliosarcoma is still a matter of speculation. Gliosarcoma in general is presumed to have a common precursor cell, which subsequently differentiates histogenetically into glial and sarcomatous components.[4] The glial elements are high-grade lesions demonstrating numerous mitosis resembling glioblastomas. Occasionally ependymoma has been observed to undergo transformation into gliosarcoma.[3] The biopsy in our index patient did not show any feature suggestive of ependymoma. It has been speculated that the subependymal portion may

undergo a malignant transformation and protrude inside a ventricle. The tissue along the septum may be involved resembling a true intraventricular tumor. Gliosarcomas, like glioblastomas, have a very poor prognosis with an average survival of 6-14 months.[4]

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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