

Extra-axial ependymoma mimicking a meningioma

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ABSTRACT

Supratentorial ependymomas are rare and are usually intra axial in location. We report a case of extra axial supra tentorial ependymoma in a girl which mimicked a falcine meningioma. Imaging, pathological features and management are described in this case report along with review of the literature. The possible origin of these tumors is discussed.

Key words: Extra axial ependymoma, meningioma, supra tentorial

INTRODUCTION

Ependymomas represent 2-9% of all intracranial tumors. They typically present in the pediatric population.^[1] Nearly 65% of these lesions are seen in midline posterior fossa. The remaining are seen in supra tentorial location. Most of these supra tentorial ependymomas are intraventricular in location. A small minority of them is intra axial without any ventricular attachment. The origin of these lesions is controversial. Some authors have theorized that intra axial lesions develop from ventricular extensions that subsequently have cut-off.^[2] There is another opinion that these lesions arise from ependymal embryologic remnants.^[3] Primary extra axial ependymomas are rare.^[2]

CASE REPORT

The present case report is about an 18-year-old female patient who presented with clinical features of raised intracranial pressure and blurring of vision. Clinical examination revealed bilateral papilledema and left hemiparesis. Magnetic resonance imaging revealed a falx-based extra axial lesion with significant vasogenic edema. The lesion was isointense on T1 weighted imaging and hyperintense on T2 weighted imaging. It was surfacing and abutting the superior sagittal sinus. It was enhancing on contrast. There were some non-enhancing

areas inside the lesion suggestive of necrosis [Figure 1]. Patient was operated with a pre-operative diagnosis of falcine/para sagittal meningioma and the lesion was completely excised. The tumor was firm, vascular with a good arachnoid plane on the parenchymal side. The sagittal sinus was not involved. The part of the falx where the tumor was attached was also excised along with dura over the convexity of the tumor. There was no parenchymal invasion by the tumor. Patient had an uneventful post-operative stay in the hospital and was discharged on the 5th post-operative day. Her neurological deficits had improved to normalcy at the time of discharge.

The material was subjected to routine histopathology and immunohistochemistry (IHC). Routine histopathology showed tumor composed of monomorphic oval to round cells arranged in nests and around blood vessels forming rosettes. There were spindle cells in some areas and clear cells in some places. Nuclei were oval to round in appearance. Nuclear atypia and mitoses were not seen. Glial fibrillary acidic protein was strongly positive suggesting glial origin of the tumor. MIB-1 was 2-3% in areas of high proliferation. Epithelial membrane antigen was positive in many cells. Vimentin and S-100 were positive [Figure 2]. The histopathology and IHC favored a diagnosis of Ependymoma World Health Organization (WHO) grade II. Patient has been coming for regular follow-up and there has been no recurrence of symptoms. Repeat imaging was done 2 years post-operative and it did not show any tumor recurrence [Figure 3].

DISCUSSION

Ependymomas are glial neoplasms arising from the ependymal lining of the ventricular system and central

Access this article online	
Quick Response Code:	Website: www.ijns.in
	DOI: 10.4103/2277-9167.138918

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canal of the spinal cord. They are more common in the infra tentorial region. Supratentorial ependymomas are seen in the adult population. They are either in the ventricular system or in the cerebral parenchyma without any obvious association with the ventricular system.^[4]

Extra axial ependymomas are extremely rare. In our case the lesion was completely extra axial, supra tentorial and dural based. Intra-operatively no parenchymal extension was seen. In the literature, no definite cause has been postulated. One hypothesis involves the extension of subcortical sub ependymal rests extra axially with the subsequent growth of tumor.^[2] Necrosis and calcification of the originating sub ependymal rests would then follow, leaving a predominantly extra axial ependymoma. Some authors postulated that a microscopic cellular tract existed between the ventricular system and the extra axial ependymoma.^[5] Heterotopic placement of ependymal cell rests during fetal development with subsequent growth of tumor has also been discussed as a possible mechanism.^[6]

Supra tentorial ependymoma and infratentorial ependymoma are different in clinical and radiological presentation. Patients with infra tentorial ependymomas present with hydrocephalus where as those with supra tentorial ependymomas present with raised intra cranial pressure, seizures and focal deficits.^[7]

Radiologically ependymomas are iso to hypo intense on T1 weighted imaging and hyper intense on T2 weighted imaging. They are contrast-enhancing lesions. Infra tentorial lesions are solid whereas cystic components are common in supra tentorial ependymomas. Calcification, necrosis and areas of hemorrhage are common to both. Astrocytoma, primitive neuro ectodermal tumors, oligo dendroglioma are to be considered in the differential diagnosis.

Total surgical resection is the mainstay of treatment.^[7] Supra tentorial lesions are more amenable for surgery. In our case, the tumor was resected completely. No adjuvant therapy was considered since histopathology was ependymoma WHO grade II. Follow-up imaging done 2 years post-surgery did not reveal any tumor recurrence. Role of radiotherapy in WHO grade III ependymomas has been emphasized in earlier studies.^[2] Rancoroli *et al.* proposed that cortical supra tentorial low-grade ependymomas should be treated with surgery alone, if they are amenable to gross total resection and are not prone to local recurrences.^[7] The grade of the tumor and extent of surgical resection have been found to be important prognostic predictors of survival, as patients with gross total resection tend to fare better than those with subtotal resection.^[8]

Niazi *et al.*, concluded that all patients with WHO grade III ependymoma require radiotherapy, where as those

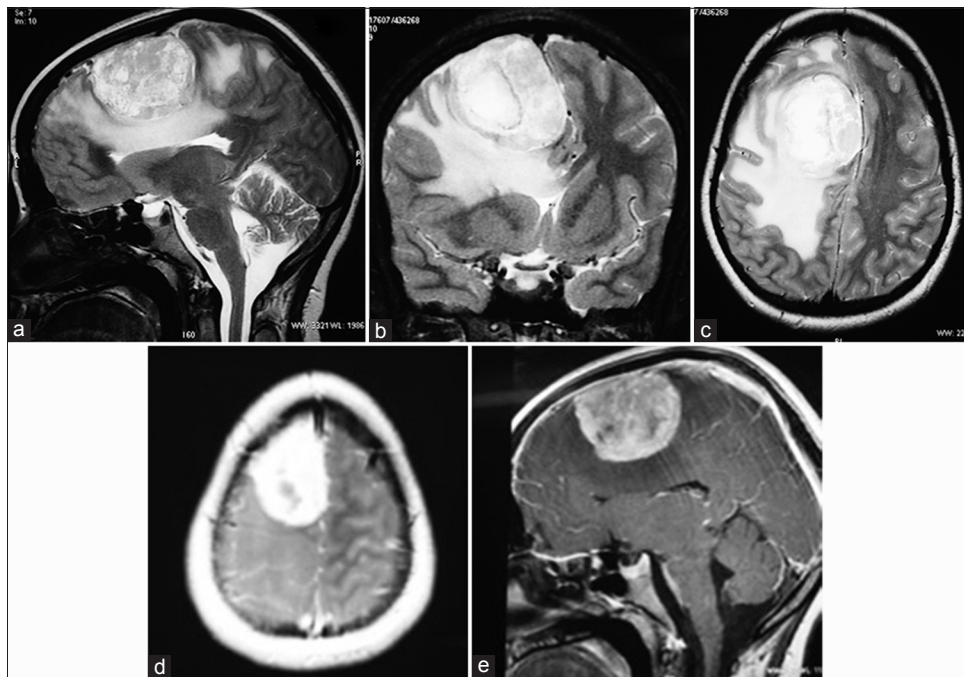


Figure 1: Magnetic resonance imaging images showing an extra axial falx based enhancing lesion (a) T2 sagittal view of the extra axial tumor (b) T2 coronal view of the extra axial tumor (c) T2 axial view of the extra axial tumor (d) pre-operative contrast enhanced axial image showing an extra axial falx based enhancing lesion (e) pre-operative contrast enhanced sagittal image showing an extra axial falx based enhancing lesion

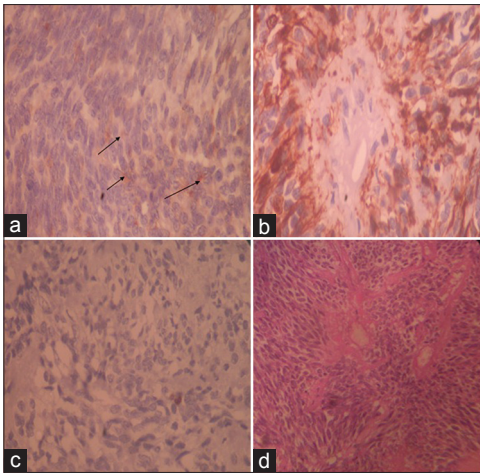


Figure 2: Histopathology and immunohistochemistry (a) dot like perivascular positivity (x40) (b) perivascular tumor cells showing positivity for glial fibrillary acidic protein (c) low MIB-1 activity (x40) (d) perivascular arrangement of tumor cells (H and E, x40)

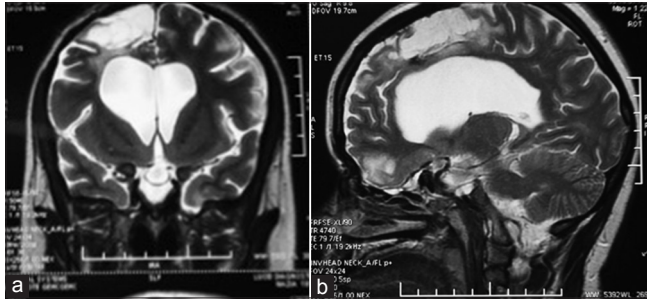


Figure 3: Post-operative imaging at 2 years follow-up showing no tumor recurrence (a) coronal image (b) sagittal image

with grade II lesions can be managed with total resection alone.^[9] Previous studies also report that total resection alone is sufficient for WHO grade II ependymoma. Adjuvant treatment is required only if subtotal resection was done. Mansur *et al.* in a series of 60 patients showed that in patients with supra tentorial tumor with subtotal resection had a worse outcome, regardless of tumor type.^[10] The role of chemotherapy in management of ependymoma is still ill-defined. There has been no survival advantage with chemotherapy.

Ependymomas can occasionally occur outside the central nervous system (CNS). They may be found in the following situations.

- As metastatic extension from a primary CNS neoplasm

- As a direct extension into the soft-tissue of the sacro coccygeal area from a primary ependymoma of the spinal cord, filum terminale or cauda equina
- As a primary tumor of the skin and subcutaneous tissue without any demonstrable connection with the spinal cord
- As a primary presacral pelvic or abdominal tumor.

The treatment of choice for these extra CNS ependymomas is total excision.

CONCLUSION

Extra axial ependymomas are very rare lesions. Total surgical resection is the treatment of choice. Long-term follow-up is required to look for recurrence. In our case, we followed the patient for 2 years with no tumor recurrence.

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How to cite this article: Puduru SS, Rajesh BJ, Shah M. Extra-axial ependymoma mimicking a meningioma. *Indian J Neurosurg* 2014;3:103-5.

Source of Support: Nil, **Conflict of Interest:** None declared.