Case Report

Extra-axial giant falcine ependymoma with ultra-rapid growth in child: Uncommon entity with literature review

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ABSTRACT

Ependymoma tends to occur commonly along either on ventricular surfaces of the brain or central canal of the spinal cord. Rarely, ependymoma can develop in the cerebral cortex without attachment to the ventricular structures. However, such occurrence in the purely extra-axial compartment in the falcine region mimicking falcine meningioma is exceedingly rare. The detailed search of primary falcine ependymoma (PEFE) in Medline and PubMed yielded only five isolated case reports. All cases occurred in patient older than 17 years of age; however, our case was a 9-year-old girl. PEFE tends to occur more commonly in male with a male to female ratio of 3:2, with the mean age of 28.8 years (range 17–43 years), lesion had intra-operatively had solid consistency in all cases. All cases were subjected to surgical resection followed by adjuvant radiotherapy; however, in addition, one case also received chemotherapy, with an average follow-up period of 9 months, however, missing in one case. The mean size of the tumor was 5.25 cm (range 1.8–7.2 cm). The authors present a unique case of PEFE in a 9-year-old girl with 8.6 cm × 6 cm × 5.4 cm-sized giant primary falcine ependymoma, managed surgically successfully. To the best of the authors’ knowledge, the current case is the first case of pediatric extra-axial falcine ependymoma occurring within the first decade of life in the western literature, showing rapid evolution over 1½ month’s period into a giant size. Brief pathogenesis, clinical feature, and management along with the pertinent literature are reviewed briefly.

Key words: Anaplastic ependymoma, extra-axial falcine ependymoma, first decade

Introduction

Ependymoma is the primary neoplasm of the central nervous system, and in the adult population, it accounts for about 5% of all intracranial gliomas.[1-3] Age is an important factor in the development as ependymomas in the supratentorial compartment tend to occur more frequently in the adult than pediatric populations.

Ependymoma arises from the lining cells, ventricular system of the brain, and central canal of the spinal cord. However, ependymoma can rarely originate without connection or continuity with the ventricular system in the supratentorial compartment typically called as cortical, ectopic, extraventricular, or lobar ependymoma.[1-4]
Management still remains debated due to the paucity of literature; further primary ependymoma located in the falx region (PEFE) is extremely rare. We report a unique parafalcine ependymoma occurring in a 9-year-old girl. She underwent successful surgery for the suspected preoperative diagnosis of falcal meningioma and tolerated surgical intervention very well.

**Case Report**

A 9-year-old girl, born to nonconsanguineous parents, reported to our neurosurgical services with the complaints of left-sided focal seizure for the last 1 year and progressive headache with visual deterioration for the last 2 months. She had sought several local treatments before being referred to our tertiary care center. There was no history of fever, evening rise of temperature, weight loss, or head trauma. She had no other significant comorbid medical illness. On admission, she was conscious, oriented with visual acuity was absent perception of light on both eyes with fundi showing bilateral secondary optic atrophy and left-sided upper motor neuron seventh cranial nerve paresis. She had left-sided spastic Grade-4 hemiparesis with brisk reflexes and Babinski positive.

Preliminary investigations including urine examination were normal.

The initial contrast-enhanced computed tomography (CT) scan of the head revealed a large heterogeneous mass, size measuring approximately 3 cm × 2.1 cm with perilesional edema in the middle third right falcal region mass with the presence of calcification [Figure 1a]. Magnetic resonance imaging (MRI) brain axial section image also revealed heterogeneous enhancing parafalcine mass of 2.2 cm × 3 cm with attachment to the falx with the presence of arachnoid cap with associated perilesional parenchymal edema [Figure 1b]. Parents were advised about the need for urgent surgical intervention to decompression and find out histological diagnosis and need of any further requirement of adjuvant therapy; however, parents were not willing for any surgical intervention including biopsy, in view of poor literacy level, they flatly refused surgical intervention.

However, over the period of about 1½ months, headache continued to worsen with an associated increase in the focal seizure frequency, and then again reported to our emergency neurosurgical services. A repeat MRI brain was carried out to plan for corridor and approaches of surgical intervention, revealed massive growth of the size of lesion [Figure 2a] compared to the previous scan; size was 8.6 cm × 6 cm × 5.4 cm [Figure 2b], heterogeneous with marked mass effect causing effacement of ipsilateral ventricle and associated subfalcine herniation and dilatation of contralateral left lateral ventricles [Figure 2c]. MR venography was suggestive of partial occlusion [Figure 3]. She was kept on intravenous steroids and cerebral decongestant. She underwent surgery under general anesthesia, a three-fourth of bicoronal scalp incision was made, and scalp flap was raised; a frontoparietal free bone flap only crossing the mid-line was raised to get access to the falx and superior sagittal sinus. After bone flap elevation, the dura was opened based on the superior sagittal sinus. A reddish, highly vascular mass with markedly dilated veins, tortuous veins, and engorged veins was present on the tumor surface. It was attached to the falx, which was gradually severed, slow devascularization from falx, and internal debulking was done. At few places, the lesion had poor dissection planes with the brain parenchyma. After securing hemostasis, at the end of surgery, the brain was lax, and further lax duraplasty was done with pericranium. Bone flap was loosely fixed, and wounds closed in layers. The patient was ventilated overnight and extubated the next morning. CT scan head showed complete excision. She was discharged on the 6th day following the surgery, the histopathological examination was suggestive of anaplastic ependymoma, and she received adjuvant therapy. MRI brain at 12-month follow-up showed no residual ependymoma [Figure 4a-c], and screening of spine did not reveal any drop or metastatic lesion. At the last follow-up at 16 months, she was doing well with no evidence of recurrence.

![Figure 1: (a) Contrast-enhanced computed tomography of the head showing heterogeneous mass lesion in middle third part of right falcal region, (b) Magnetic resonance imaging, axial section, contrast-enhanced image of brain showing heterogeneous mass lesion in middle third of right falcal region (size 3.1 cm × 2.1 cm × 2 cm)](image1.png)

![Figure 2: Magnetic resonance imaging, contrast-enhanced image of the brain (a) axial section showing heterogeneous mass lesion of size 8 cm × 6 cm × 5 cm in middle third of right falcal region, (b) coronal section, showing heterogeneous mass lesion, (c) sagittal section, image of brain showing heterogeneous mass lesion of with massive perilesional edema)](image2.png)
Discussion

Supratentorial ependymomas occur most commonly in adults; ependymoma usually originates from the ependymal linings of the ventricular system in the brain and central canal of the spinal cord. Few cases of ependymal can originate neither from ventricle nor contain continuity to the ventricular structures are also commonly called as ectopic ependymomas, however, extremely rarely can occur in the purely extra-axial compartment only attached to falx.

In the detailed PubMed literature search of extra-axial falcine ependymoma, the authors could find out only five isolated case reports, being with more common in male with a male to female ratio of 3:2; however, our case was female. The mean age of the patients was 28.8 years with the range varying from 17 to 43 years, and the youngest were 17-year-old females. The primary treatment modality was surgical resection. All cases underwent primary surgical resection followed by adjuvant radiotherapy; however, in addition, one case reported by Hanchey et al. also received additional chemotherapy as an adjuvant. The mean follow-up period was 9 months with range being 2–12 months, but not mentioned in one report [Table 1].

Still, origin of the extra-axial ependymomas is debated. According to one postulate extension of subependymal cell rests from subcortical location to the extra-axial compartment promote the development of the extra-axial located ependymoma. Further, Donich et al. hypothesized about the existence of microscopic cellular tract in between the site of occurrence of extra-axial ependymoma and the ventricular system. During fetal development, the aberrant placement of ependymal cell rests and subsequent growth of extra-axial ependymoma may be another mechanism as postulated by Youkilis et al. In case of PEFE, located in the falx suggested heterotopically placed ependymal cell rests in the falx may represt the possible site of origin. Hanchey et al. reported a case of large interhemispheric ependymoma occurred in a 29-year-old male, diagnosed on the basis of cerebral angiography and technetium 99m scintigraphy, author further added although the ependymoma was extra-axial but contained definite extension toward ventricles.

Primary extra-axial ependymomas clinically present with local pressure effects such as falcine meningioma. On MRI imaging study, meningiomas usually show homogeneous isointense signal T1- and T2-weighted images and intense enhancement on gadolinium study, however, ependymomas are heterogeneous due to the presence of calcification and multiple cyst.

Management of ependymoma is usually primary radical surgical excision. Radical excision of ependymoma reduces the chances of recurrence and increases the effect of adjuvant therapy. Adjuvant radiotherapy is usually indicated with subtotal surgical or histopathological high-grade lesion or presence of craniospinal seedlings. Postoperative adjuvant therapy delays the occurrence of recurrence and increases survival period.

Various prognostic factors of ependymoma are age, location, and degree of surgical resection, histopathology findings, adjuvant therapy, and histopathological grades. Prognosis of infratentorial ependymoma is superior to the supratentorial counterpart.

Summary

Primary extra-axial ependymoma is a rare entity; however, a possibility of occurrence of PEFE in the falcine region should be one of the differentials that should be kept in mind. Early and radical surgery improves the surgical outcome, and appropriate cases should be subjected to adjuvant therapy.
Table 1: Previously published pediatric cases

| Authors (reference number) | Year | Sex/age (year) | Location | Lesion size (cm) | Cystic/solid | Surgery-resection | Dural attachment | Histology (WHO grade) | Adjuvant therapy | Follow-up |
|----------------------------|------|----------------|----------|-----------------|-------------|------------------|------------------|-------------------|-----------------|-----------|
| Park et al. [9]            | 2010 | Female/17      | Right parafalcine falx | 7 | Solid | Gross-total | Absent | Anaplastic Grade III | Radiotherapy | Alive at 2 months |
| Current case               | 2016 | Female/9       | Right parafalcine       | 8.6 | Solid | Gross-total | Present | Anaplastic Grade III | Radiotherapy | Alive at 12 months |

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Conflicts of interest
There are no conflicts of interest.

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