

Parafalcine Ependymoma in Elderly Patient- An Unique Case with Literature Review

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Abstract

Primarily extra-axial ependymoma without any visible connection of the ventricular system is very rare. In the literature there are only five cases have been reported till date. These all five cases are in the first decade of life. We are presenting a case of extra axial ependymoma in Parafalcine location without any proximity to ventricular system in a male in his eighth decade of life.

Keywords: Ependymoma; Extra-Axial; Tumour; Ventricular System; Glial Fibrillary Acidic Protein

Introduction

Primarily extra-axial ependymoma without any visible connection of the ventricular system is very rare.

Case Report

An 80-year-old gentleman presented with insidious onset progressive spastic right hemiparesis for last one year, starting from lower limb first then involved ipsilateral upper limb later on.

On neurological examination, the patient was otherwise healthy except weakness involving right lower limb for MRC grading of 3/5 at the hip, and knee flexors, 3/5 in large groups of the upper limb. Biceps, triceps, knee and ankle reflexes were exaggerated in right side. No other cranial nerves and neurological abnormality seen.

On imaging by MRI, there was dural based mass present in left Para falcine location. The mass was isointense in T1 and T2 with inhomogeneous contrast enhancement with few voids in the central areas (Figure 1). The mass was well-circumscribed but medial to brain parenchyma causing pressure over postcentral gyrus.

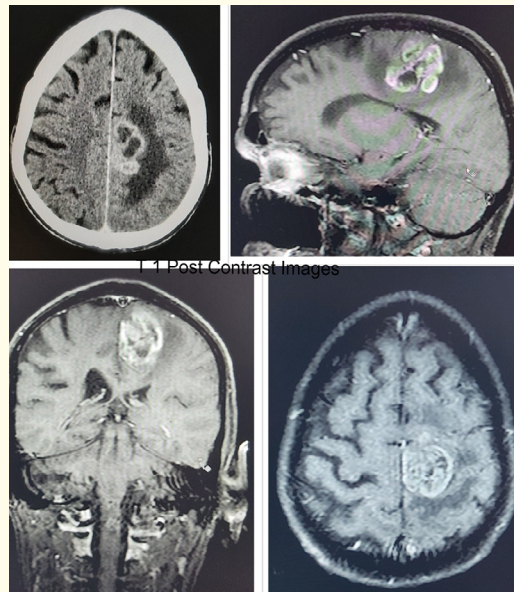


Figure 1: MRI images.

Surgery

The patient underwent surgery over Sugita Head Clamp in the supine position with neck flexed making the vertex almost parallel to the floor.

Left parietal cross midline craniotomy with stripping the superior sagittal sinus from the inner table of calvaria done via a horseshoe-shaped incision over the vertex (Figure 2). Dura was cut and reflected in curvilinear incision after taking due care for draining cortical veins. Small corticectomy (1 x 1) done anterior to paracentral gyrus and gross total resection of the tumour was done under microscopic guidance. The tumour was greyish white soft friable non adhered to adjacent brain parenchyma but loosely adhered to the falx.

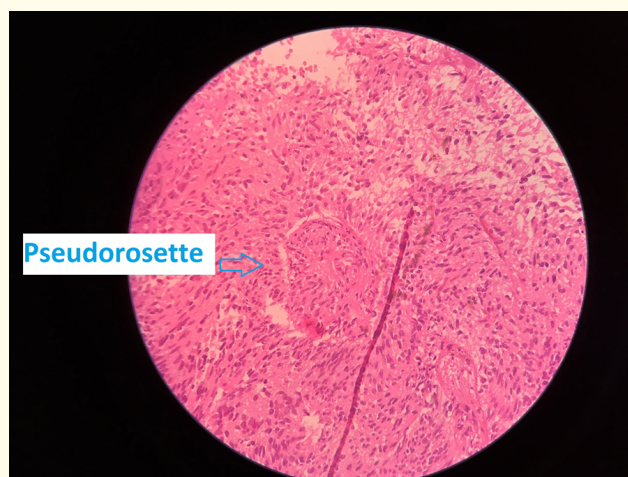


Figure 2: Microscopic view.

Postoperatively patient developed transient increment of weakness which recovered significantly over the next few days with the help of physiotherapy to the level of MRC grade 4 in both upper limb and lower limb.

Follow up scan revealed total removal of mass without any significant damage to surrounding structures.

The histopathological examination revealed Cellular neoplasm showing perivascular pseudorosette formation and Monomorphic cells with oval nuclei, salt and pepper chromatin and peripheral palisading. The GFAP stain was positive. These findings were consistent with Ependymoma (Figure 3).

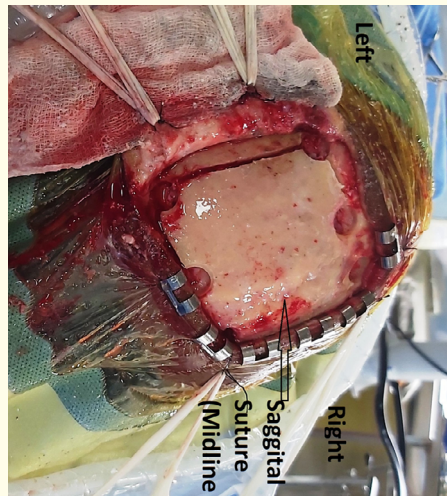


Figure 3: Intraoperative images.

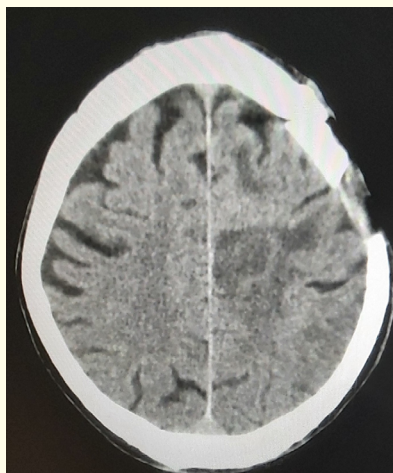


Figure 4: Postoperative scan.

Discussion

Ependymoma a rare type of brain tumour constitutes about 3 - 4% of all intracranial tumours. Majority of these arise in first three decades of life with predominance in posterior fossae location. However few shows deviation from this trend and presents in supratentorial compartment.

About half of supratentorial ependymoma presents closely in relation to ventricular system. The most amenable theory states that fetal rests of ependymal cells produce such ependymomas.

In our case the ependymomal mass was located in parafalcine region in an 80 year old gentlemen. Epidemiologically this type of pattern is not mentioned anywhere in literature. We are the first to report this.

Extra axial location of ependymoma is extremely rare. In our exhaustive search we find only five cases of extra-axial ependymoma. Two among these five were infratentorial extra axial while three were supratentorial extra axial. All of them were from upto third decade of life. Our case is the only one with supratentorial extra-axial in elderly patient of 80 years of age.

Case no.	Location	Author	Age
1	Infratentorial	Cosgrove., <i>et al.</i>	
2	Infratentorial	Donich., <i>et al.</i>	
3	Supratentorial	Hanchey., <i>et al.</i>	29 years
4	Supratentorial	Hayashi., <i>et al.</i>	13 years
5	Supratentorial	Andrew., <i>et al.</i>	20 years

Exact mechanism of development of these extra-axial tumours is not known. However, Hayashi postulates that subcortical growth of subependymal rest in the extra-axial location may be the cause of growth of these tumours. Later on calcification and necrosis takes place due to scant blood supply, making it completely extra-axial with or without parenchymal tailing. Donich., *et al.* [7] theorized that a microscopic tail exists between the tumour and ventricular lining connecting through cerebral parenchyma.

Another ill proved theory by Anderw., *et al.* is heterotopic placement of fetal ependymal cell rests during fetal development with subsequent growth of such tumours.

Radiographically extra-axial ependymomas are difficult to diagnose. The magnetic resonance picture can be very similar to meningioma as both may be isointense in t1 and T2 but in homogenous contrast enhancement is the character which raises first suspicion of ependymoma. Similar to other extra-axial tumours ependymomas are usually well circumscribed with some solid and cystic component within. Sometimes the appearance is more like gliomas but location and well defined margins are the key to differentiate.

The first line of definitive management of intracranial ependymoma is surgery with emphasis on complete resection. The complete resection nearly cures the tumour. The postoperative chemotherapy or Radiotherapy is not required in patients of complete resection they can be followed by regular follow up with clinical examination and/ or imaging if required (Palma., *et al.*). Post-operative radiation is thought to be effective in increasing the life span Subtotal resection but due to rarity of entity small sample size is the hindrance in definitive inference to conclude.

In our case the tumour was resected completely so no further adjuvant therapy was advocated. Though regular follow up is advised.

In the literature various factors were considered to determine the prognosis including age of presentation, location of tumour, histopathological grading and surgical resection. The most consistent factor among these study is extent of resection. Complete resection is associated with increased survival. Though older age of presentation is associated with prolonged survival in infratentorial ependymoma [8], it is not with supratentorial ependymomas [7].

Literature is also inconsistent with impact of location of ependymoma on survival but Schwartz, *et al.* reported decreased survival in association with third ventricular proximity and metastatic disease.

The rarity of extra-axial ependymoma is the reason for overlooking this pathological differential diagnosis. Immunohistochemical evaluation with Glial fibrillary acidic protein (GFAP) is of utmost importance in differentiating it from the closest diagnosis of meningioma. Some schwannomas may express GFAP but absence of perivascular rosette is helpful. Similarly, oligodendrogliomas can also be ruled out with absence of perivascular rosette and epithelial membrane antigen.

Conclusion

The extra-axial supratentorial ependymoma is extremely rare. They are usually present in first three decades of life but we encountered in a patient in eighth decades of life harbouring it. The origin of these tumours theorized to be from growth of subependymal cells in subcortical region reaching to extra-axial location.

Complete surgical resection is associated with longest survival with some role of radiation in subtotal resection.

Though supratentorial extra-axial ependymoma is rare they should be considered in all the dural based masses and surgical team should be prepared in advance to avoid intraoperative surprises.

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