

Paediatrics Supratentorial Extraventricular Ependymoma

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Abstract

Introduction: Majority of ependymomas are infratentorial and intraventricular. Supratentorial and purely cortical extraventricular ependymomas are extremely rare and only six cases were reported so far.

Case report: We report a 12-year-old female with right parieto occipital ependymoma. She had complete excision of the lesion.

Conclusion: Pure cortical, supratentorial extraventricular ependymoma in paediatrics age group is a rarity.

Keywords: Cortical; Ependymoma; Extraventricular; Paediatrics

Introduction

Ependymomas are usually infratentorial and a third of the ependymomas are supratentorial. Of the supratentorial ependymomas, extraventricular tumors account for 50% [1] the majority of them are in the vicinity of the ventricles and have some connection to ventricular margins. Purely extraventricular ependymomas with no connection to ventricular lining are very rare and only six cases were reported in the literature so far [1-3]. We report one pediatric case of supratentorial extraventricular ependymoma.

Case Report

A 12 year old girl presented with one episode of seizure for which she was treated and investigated in form of contrast computerized tomography. CT brain which showed a right parieto-occipital peripherally placed lesion with perilesional edema and hypodense space medially (Figure 1).

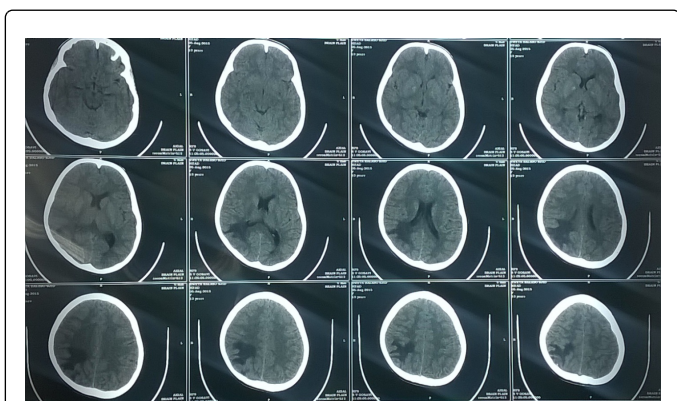


Figure 1: Right parieto occipital hypodense lesion with perilesional edema

MRI of the Brain Contrast study was done suggestive of well defined, lobulated, oval intra-axial enhancing space occupying lesion in the cortical, subcortical aspect of the right parieto occipital region. It showed solid as well as cystic component in it (Figure 2).

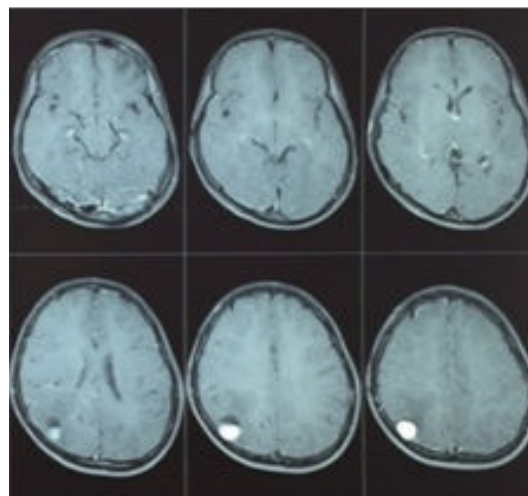


Figure 2: MRI showing right parieto occipital enhancing space occupying lesion

Right sided parieto occipital craniotomy done. Dura opened, brain bulge present. Greyish white tumour coming out. Tumour was moderately vascular and of firm to soft consistency. Complete excision of tumour done. Clear fluid gushed out after delivering the last part of tumour (Figure 3). The tumor had no connection to the ventricular ependymal lining. Margins were well defined. The postoperative period was uneventful and she recovered well. Histopathology of the excised tumor showed that the tumour was composed of round neoplastic cells arranged in rosettes, clusters and small sheets. Prominent perivascular rosettes with fibrillary structures oriented towards the blood vessels are noted. At places the tumor appears cellular and compact. The cells are round to oval having round to oval delicate vesicular nuclei with

inconspicuous nucleoli and moderate amount of eosinophilic cytoplasm (Figure 4). Some mitotic activity is noted. The neoplastic cells are separated by thin fibrovascular stroma. Immunohistochemistry of the excised tumor done for confirmation and grading. On microscopic examination EMA revealed to be strongly positive showing paranuclear dot positivity. MIB-1(Ki-67) is and 8-10%. Diagnosis of an ependymoma of World Health Organization (WHO) grade III was made.

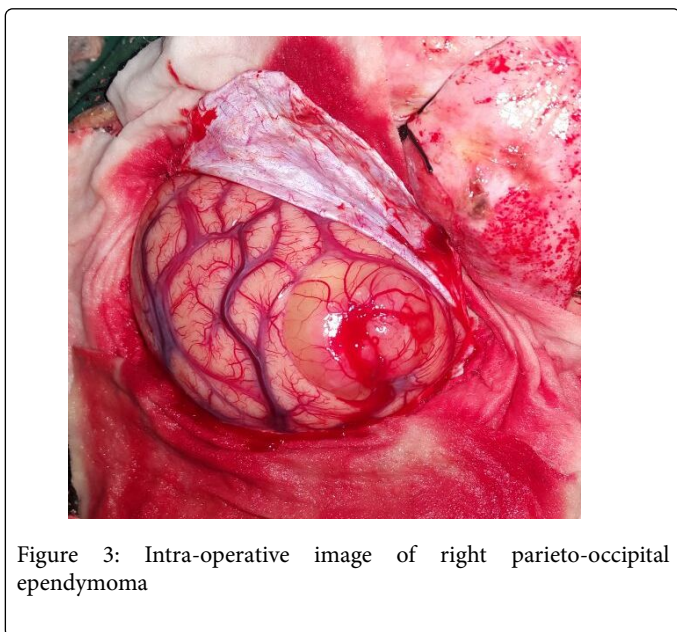


Figure 3: Intra-operative image of right parieto-occipital ependymoma

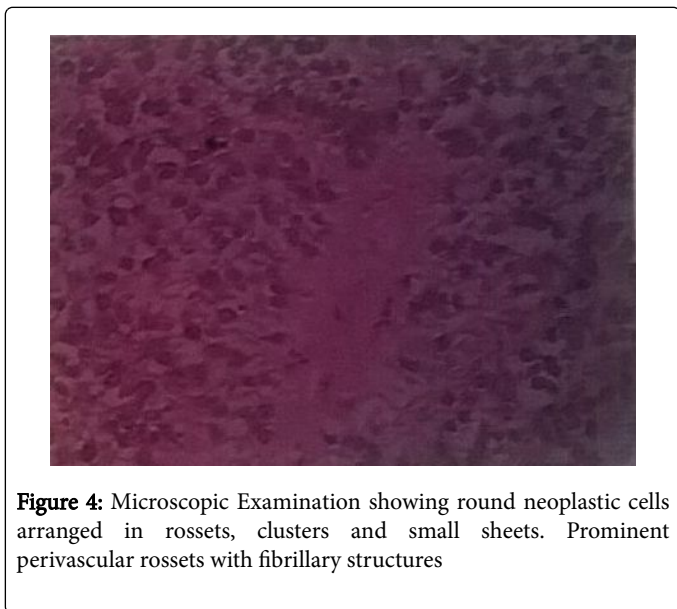


Figure 4: Microscopic Examination showing round neoplastic cells arranged in rosetts, clusters and small sheets. Prominent perivascular rosetts with fibrillary structures

Discussion

Till date, only six cases of purely cortical supratentorial extraventricular ependymomas have been reported in literature [1-3]. Supratentorial ependymomas represent a subgroup with a comparatively better prognosis when compared to infratentorial ependymomas. This may be related to the fact that total resection is more commonly achieved in supratentorial tumors. Adults have a better five-year survival than children. Children less than 2.5 years of age, have a significantly worse prognosis than older children [4]. In the young children, five-year survival is 22.40% as compared to 60.75% in older children [4]. Prognosis in children older than five years is same as that of adults [4]. Outcome can be better correlated when age at diagnosis is considered along with extent of surgical resection [4]. The need for postoperative adjuvant therapy has been controversial for supratentorial ependymomas. In general, it is considered safe to observe the patient when postoperative CT or MR shows gross total excision, particularly when the tumor is of low grade [5]. Even in the presence of recurrence some authors suggest total excision by a second operation. Thus, the option for postoperative radiation is only for high-grade tumors and tumors in locations where total excision is not possible [5].

Conclusion

Pure cortical supratentorial extraventricular ependymoma in paediatric age group is a rarity.

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