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Case Report

Supratentorial atypical teratoid/rhabdoid tumor: a case report

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ABSTRACT

Atypical teratoid rhabdoid tumor (ATRT) is a rare and highly aggressive malignant tumor of infancy and childhood with fatal outcome. Common site is kidney, but it can also occur in the liver, thymus and the CNS. The supratentorial compartment is less frequently the primary site for this neoplasm, but cases arising from the cerebral hemisphere have also been reported. Study came across a case of 18 months female child who presented with convulsions and MRI showed right frontal space occupying lesion, which after IHC confirmed as ATRT.

Keywords: ATRT, Diagnosis

INTRODUCTION

Primary CNS atypical teratoid/ rhabdoid tumor (ATRT) is a rare and highly aggressive malignant tumor comprising 2-3% with an age predilection mainly in less than 2 years. It has also been reported to occur in adults and older children.'[1]

The supratentorial compartment is less frequently the primary site for this neoplasm, but occasional cases are also arising in the pineal region'.[2-5] The histological hallmark of ATRT is the presence of rhabdoid cells.[4] Disseminated forms occur in 20 to 30% cases and generally follow a dismal course with a median time to death is first a few months after diagnosis.[5] We report an unusual case of supratentorial ATRT in 18 months female child.

CASE REPORT

An 18 months female child who presented with single episode of convulsion along with history of fever 6 days back. No history of neck stiffness, vomiting or injury and milestones were within normal limit for the age. CSF examination showed proteins - 294 mg/dl, glucose - 77 mg/dl and nucleated cells - 25 cells/microlitre. Lymphocytes - 90%, neutrophils - 10%. Gram stain and ZN stain were negative. MRI brain showed large supratentorial intra-cranial mass lesion in right frontal region causing midline shift to left side with subfalcine ancal herniation.

Lesion also showed central cavity enhancing, with thick peripheral wall, blooming and extension of cerebral haemorrhage (Figure 1). MRI findings were suggestive of PNET.

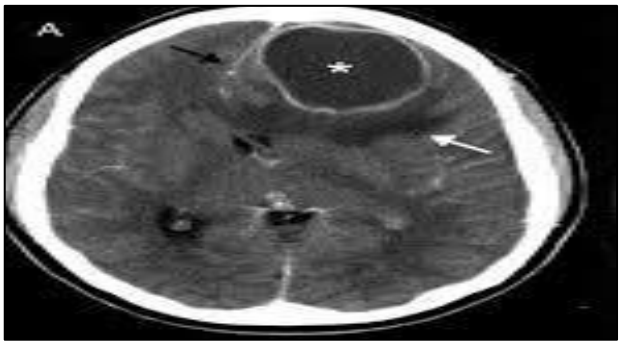

Figure 1: MRI brain of large frontal mass.

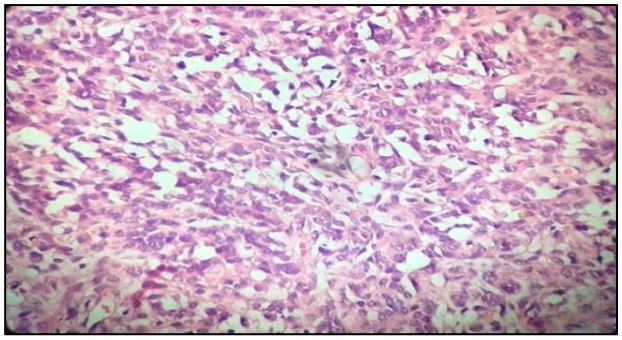

Figure 2: H/E section - tumour cells having large polygonal cells having abundant eosinophilic cytoplasm and round eccentric nucleus with prominent nucleolus.

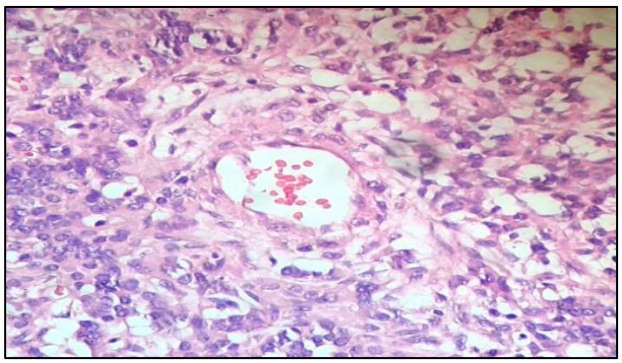

Figure 3: Pri-vascular tumour cells arrangement (H/E).

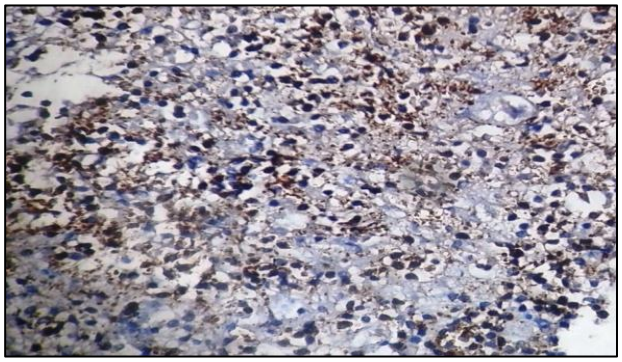

Figure 4: The cells were positive for vimentin (IHC).

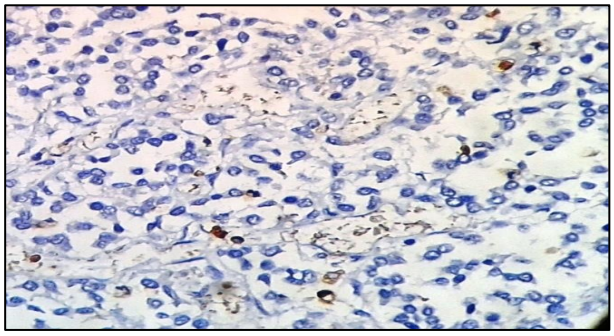

Figure 5: Tumour cells were negative for LCA (IHC).

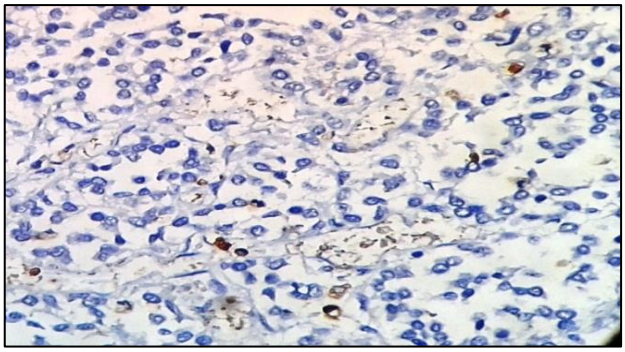

Figure 6: Tumour cells were negative for CD 68 (IHC).

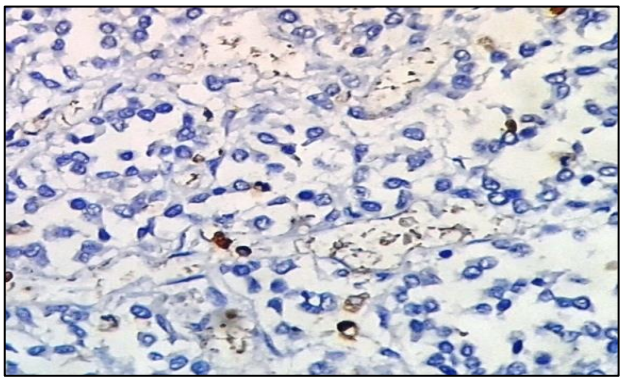

Figure 7: Tumour cells were negative for GLAP.

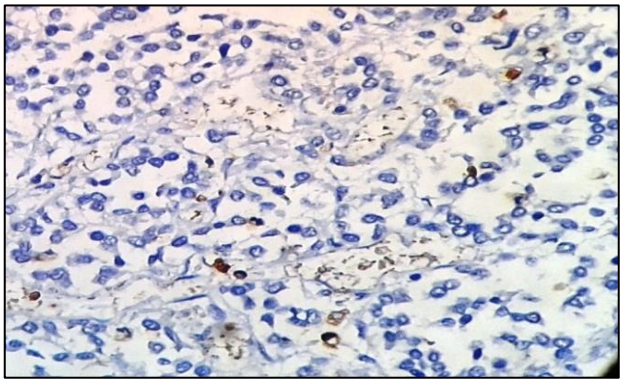

Figure 8: Tumour cells were negative for CK (IHC).

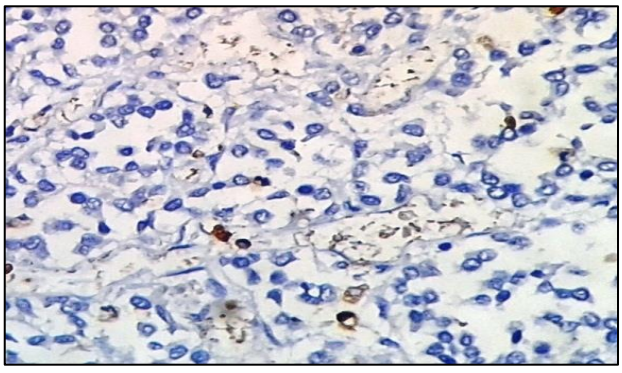

Figure 9: Tumour cells were negative for PR (IHC).

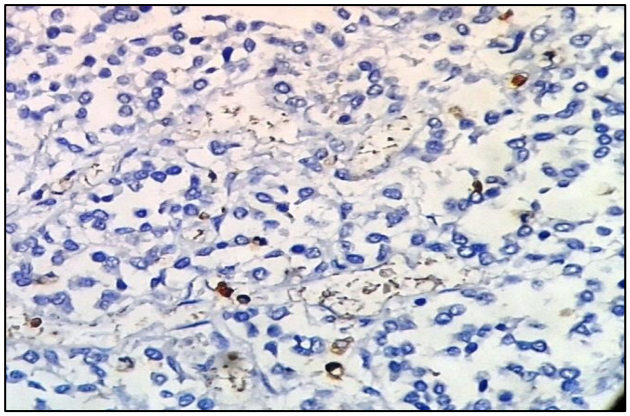

Figure 10: Tumour cells were negative for EMA (IHC).

Hence surgical resection of the mass was done and histopathological findings showed multiple friable brownish greyish tissue bits aggregating to 30 cc. On microscopy showed diffuse tumour cells arrangement infiltrating surrounding normal brain parenchyma. Some areas showed large polygonal cells having abundant eosinophilic cytoplasm and round eccentric nuclei with prominent nucleolus (Figure 2). Some of the areas showing perivascular tumour cells arrangement (Figure 3). There were large areas of necrosis and moderate infiltration of inflammatory cells noted comprising of lymphocytes, plasma cells and macrophages. Thus based on histopathological findings we kept two differential diagnosis of ATRT and PNET. IHC was performed, the cells were positive for vimentin (Figure 4). M1B labeling was 12.1% and negative for LCA (Figure 5). CD 68 (Figure 6), GFAP (Figure 7), CK (Figure 8), PR (Figure 9), EMA (Figure 10). Thus confirmed the diagnosis of atypical teratoid Rhabdoid tumor (ATRT) (WHO grade IV).

DISCUSSION

ATRT (Grade IV) is a rare and highly aggressive malignant tumour affecting infants and young children'.[1,2] They comprises 2% to 3% tumor in children in less than 18 yrs, but majority of patients are younger than 3 years It frequently affects the posterior fossa. Metastases occur early through the CSF. Other sites affected include the cerebral hemisphere, the pineal region and the spine'.[3-5] This is a case report of supratentorial ATRT in an 18 months child. Radiological (MRI) findings of PNET/ATRT are similar hence PNET is the differential diagnosis. ATRT is misdiagnosing as PNET because 70% of ATRT contain fields indistinct from classis PNET'.[2,6] The histological features can vary and can resemble medulloblastoma/PNET with small primitive looking neuronal cells with anaplasia. Rosette form can also be seen. Therefore it is therefore important to have differential diagnosis of ATRT in mind especially in younger children.'[7] In present case tumour shared the microscopic features of PNET/ ATRT. On IHC, ATRT stain positive for Vimentin and focally or weakly positive for EMA, GFAP, and SMA. Proliferative activity is high as the labeling index with Ki- 67/MIB-1. Present case showed strong positivity for Vimentin and MIB -1 is 12.1% positive. LCA, CD 68, GFAP, CK, PR, EMA were negative. Thus, confirmed the diagnosis of Atypical teratoid/ rhabdoid tumor (ATRT) (WHO grade IV).

ATRT show monosomy for chq 22 and tumour suppressor gene chq 22 .911.2' which helps to distinguish from other PNET including medulloblastom.[2,7] INI 1 gene is implicated in the intra cranial ATRT both by somatic and by germ line mutation. ATRT lack the INI-1 gene product.[8,9]

These tumours are highly aggressive and have malignant local infiltration making total excision infeasible. Prognosis is dismal with median survival of 6 months only. When associated with CSF dissemination survival in just 2.5 months.

Because of its rarity and rapid course, there has been no concern as to the optimal treatment of this tumour. However, surgery, radiotherapy and chemotherapy may all play a role.[10] Recurrent ATRT in less than 3 years of years of age are chemoresistant. Debulking followed by chemotherapy with single agent cyclophosphamide, Ifosphamide with or without radiotherapy considering the age of patient.[10]

CONCLUSION

Though atypical teratoid/ rhabdoid tumor (ATRT) is rare in supratentorial location it should be always kept in mind in less than 3 years children, due to its fatal clinical outcome and aggressive behaviour, MRI , histological features and IHC play a crucial role in the diagnosis.

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