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Research Article

RARE EMBRYONAL TUMOUR OF CENTRAL NERVOUS SYSTEM-ATYPICAL TERATOID/ RHABDOID TUMOUR

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ABSTRACT

Atypicl Teratoid/Rhabdoid Tumour is malignant embroyonal central nervous system tumour manifesting in children and composed of rhabdoid cells It may or may not be resembling a classical primitive neuroectodermal tumour, epithelial tissue and neoplastic mesenchme. A highly malignant pediatric embryonal tumor of the CNS (WHO grade IV) that exhibits rhabdoid neoplastic cells with or without non-rhabdoid components. A medulloblastoma-like and/or a primitive neuroectodermal tumor (PNET)-like component are the most common non-rhabdoid elements. A carcinomatous component or a sarcomatous component can also be present in some cases. We report two case of Atypical tertoid Rhabdoid tumour, one at the age of 3 yrs and another at the age of 9 yrs, both cases are arise from supra tentorial region.

INTRODUCTION

ATRT encountered principally in Infants and early childhood. Adult onset examples are rare. The tumour may arise in either supra tentorial or infratentorial location. This tumor is 50% more common among boys than girls (Parham DM *et al* 1994, Kleihues P *et al* 2002, Bielgel JA *et al* 2002, Dang T *et al* 2003). ATRT is a malignant embroyonal tumour with biological characteristics and histological features are similar to the malignant rhabdoid tumour of the kidney 7 The mean survival time is 3 months after only surgical intervention and 8 months with adjuvant chemo- and radiation therapy (Hilden JW *et al* 1998). MRI shows a bulky and contrast enhancing mass, cystic and hemorrhagic alterations are common. Neuroradiologic profile are usually indistinguishable from that of medulloblastoma.

Cytologic polymorphism in an embryonal neoplasm alerts the diagnostic possibility of an ATRT. The tumour usually contains rhabdoid cells with additional variable components of primitive neuro ectodermal, mesenchmal and epithelial cells (David.Ellison *et al* 2013). It behave in an extremely aggressive manner with rapid development of widespread metastases and have an extremely poor prognosis (Cossu A *et al* 1993, Satoh H *et al* 1993).

Case 1: A 9 year old male child came with complaints of gereralised fits, head ache and vomiting, he had neck pain and neck tilt. On Examination his higher functions were normal, Fundus show Bilateral papilledema, Right 6th nerve palsy, pyramidal weakness in Left face, upper & lower limbs. MRI Brain show heterogeneously enhancing necrotic lesions in right temperoparietal & perisylvian cortical, sub cortical areas with entensive edema and midline shift to left side. Under General anaesthesia, Right tempero parietal craniotomy done, the tumour was adherent to the inner surface of the dura. It was well circumscribed, lobulated, reddish soft and highly vascular. The cyst was opened deep to the solid part of the tumor, it contained colourless fluid. The cyst was thick. The solid and cystic parts of the tumor was exposed all round, coagulating the feeders. Major part of the solid tumour along with the cystic part was removed. The tumour was pale pinkish, smooth, lobulated, soft and moderately vascular. Tumor is extra axial & shows infiltration of dural collagen. No necrosis seen. Histo pathological study of show large plump cells having rhabdoid morphology arranged in cords, sheets and fascicles separated by intersecting variably sized vessels. The cells have vesicular nuclei & prominent nucleoli. The tumor cells are separated into cords by a myxoid stroma. Mitotic activity is very brisk. Immuno His to Chemistry shows GFAP positive in scattered

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tumor cells, Vimentin-diffusely positive in all cells, EMA-cytoplasmic positivity in several cells, SMA-cytoplasmic positivity in tumor cells & in blood vessels. S100-negative. INI-1 deleted in tumor cells with retained expression in vascular endothelilum & inflammatory cells. Cytokeratin and Neurofilament IHC are negative.

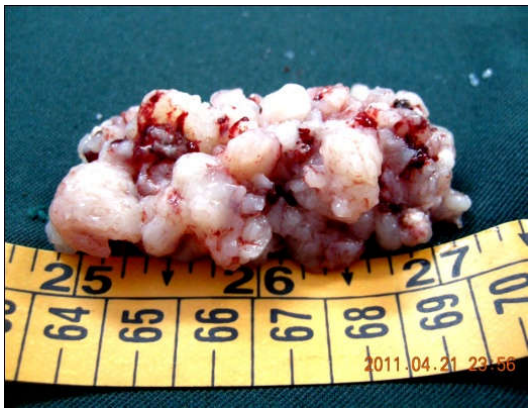

Fig No 1 show gross appearance of the tumour

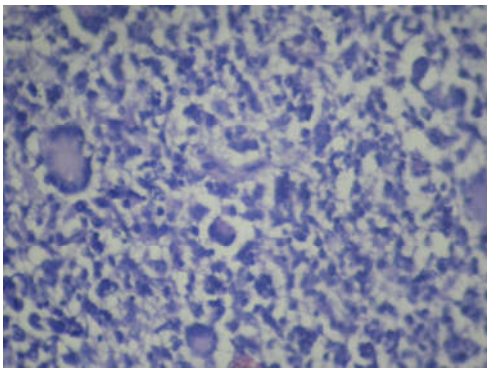

Fig No 2 show giant cells and rhabdoid cells

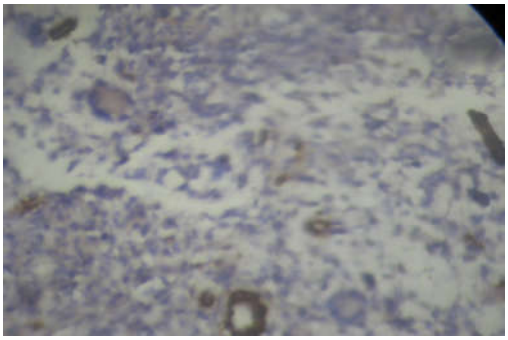

Fig No 3 Rhbdoid cells positive for EMA

Case No 2: 3 yr old male child complaints of head ache in the back of the head on the left side, he had 3 attacks of generalized seizures, 2 episodes are associated with fever, other one is non febrile.O/E: He was conscious but was found to be dull. He complained of headache and pain in the left eye. Fundi: normal. Right UMN type facial paresis. No neck stiffness. MRI study of the brain showed Mixed signal intensity lesion with cystic and solid components and enhancing nodules seen in left parieto-occipital lobe along the postero-Inferior aspects of the lesion. The lesion is measuring 5.8 x 4.9 x 4.5 cms. Moderate perilesional edema with compression of adjacent ventricular system and sulci with midline shift seen towards right side, Raised ICT with papilledema seen. Under general anesthesia with the patient in right lateral position left parieto-occipital osteoplastic craniotomy was done.

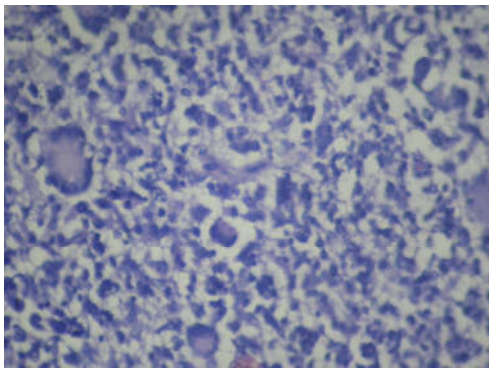

Fig No 4 show tumour cells positive for vimentin

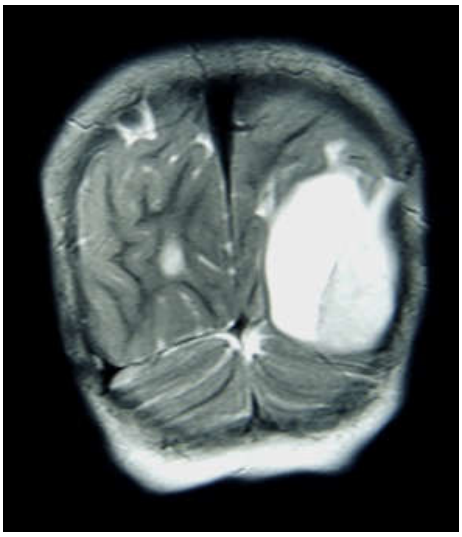

Fig No 5 show MRI brain show solid and cystic lesion in the parieto occipital lobe

The dura was opened. The cortex in the left occipital lobe was found to be thinned out; it was opened. The cyst contained slightly xanthochromic fluid. The solid parts of the tumour were found in the inferior, posterior and lateral aspects of the cyst. It was soft, grayish-pink and suckable. The tumour was found to be extending into the occipital horn of the lateral ventricle. Total excision of the solid tumour along with the cyst was done. Histopathological examination show brain parenchyma and highly cellular tumour tissue composed of round cells with pleomorphic vesicular nuclei, prominent nucleoli, clear to granular eosinophilic cytoplasm arranged in sheets, cords. Many giant cells, few rhabdoid like cells are seen. Mitotic activity is increased. Areas of increased vascularity and myxoid changes are seen. Immuno Histo Chemistry study shows tumour cells up to 70% positive with GFAP and Vimentin. Occasional Rhabdoid appearing cells Positive for EMA and SMA.

DISCUSSION

ATRT are aggressive childhood neoplasm that are located commonly in the posterior cranial fossa, Incidence show male predominance, Most patients are less than 2 yrs, However some older children and even adults with AT/RT have been described, We reported two case of male children at the age 3 yrs and 9 yrs. Clinical presentations are Recurrent Head aches,

seizures, nausea/ vomiting, progressive motor weakness, and difficulty in swallowing, Both of our case were presented with seizures, head ache. One case was presented with vomiting and neck tilt. Imaging studies, MRI show bulky, contrast enhancing mass with haemorrhage and necrosis. The histopathologic spectrum of AT/RT is broad, ranging from predominantly "small cell" with primitive morphology to tumors with large rhabdoid cells. In addition, some AT/RTs may have mesenchymal and epithelial components. Because of this morphologic variability, AT/RTs often have been misclassi ed. Cytological pleomorphism in an embroyonal neoplasm should alerts the diagonostic possibility of an ATRT. Small undifferentiated neuroepitelial cell with a high nuclear cytoplasmic ratio occupy some areas of the ATRT, but cells elsewhere often have more cytoplasm and show more nuclear pleomorphism than expected in a typical PNET. Groups of classic rhabdoid cells with abundant eosinophilic cytoplasm and eccentrically placed nuclei can sometimes be found in the ATRT& there may be large cells showing considerable nuclear pleomorphism. The mitotic count is high and necrosis is common, micorovascular proliferation in an ATRT very rarely shows the endothelial proliferation characteristic of some gliomas (David.Ellison *et al* 2013)

CONCLUSION

Atypical Teratoid Rhadoid Tumour is rare embryonal Central Nervous system tumour. We should suspectATRT when the microscopic examination show the pleomorphic cytological features and giant cells in paediatric CNS tumour. Immuno Histo Chemistry is very useful marker to diagnose ATRT

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