

## Atypical Teratoid/ Rhabdoid Tumour at Cerebello Pontine Angle- Cytopathological Features: A Rare Case Report

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**Abstract:** Atypical teratoid/ rhabdoid tumour (ATRT) is a highly malignant neoplasm primarily affecting infants and young children. The majority of tumours in the Central nervous system (CNS), approximately two-third arise in the posterior fossa. The histologic spectrum of AT/RT overlaps significantly with other forms of CNS neoplasms. The term "malignant rhabdoid tumor" was first used in reference to rare pediatric renal tumors. Neoplasms with such rhabdoid features have subsequently been identified in other locations, including the CNS with specific immunohistochemical and ultra structural features and consequently it has been called ATRT. On diagnostic imaging the tumor is most often diagnosed as medulloblastoma. The most common differential diagnosis are PNET and malignant glioma. Treatment planning depends on correct pathological diagnosis on squash cytology. FNAC or frozen section which acts as a primary method of diagnosis during intraoperative procedure later confirmed on histopathology. Herein we present a case of a 10 year old male child who initially presented to the hospital with obstructive hydrocephalus, headache, vomiting and febrile illness.

**Keywords:** Atypical teratoid / rhabdoid tumour, Medulloblastoma, Rhabdoid tumour, Squash cytology .

### INTRODUCTION

Primary CNS malignant rhabdoid tumour is a rare and fast growing tumour with an incidence of 2%. The name AT/RT was initially coined by Rorke LB *et al.* [1].

Though these tumours resemble Rhabdomyosarcomas, their immunohistochemical and ultra structural features allow a clear distinction between these two tumours. The mean age at presentation is usually 3 years with a male preponderance. However, it has also been reported to occur in older children and adults [2]. The common sites in the brain being posterior fossa (52%) supratentorial (39%), pineal (5%), spinal (2%), multifocal (2%) [3]. There are no specific imaging features for intracranial AT/RTs [4]. The cytomorphological features resemble Medulloblastoma, PNET and Giant cell Glioblastoma. Calcification, cyst formation and haemorrhage [5] are frequently seen. The cytomorphological features of AT/RT on squash cytology resemble PNET, Medulloblastoma and Malignant glioma (when extensive areas of necrosis are present). The tumour shows cytological features of epithelial (Large rhabdoid cells), primitive neuroepithelial and mesenchymal differentiation.

Although the clinicopathologic and radiologic features of AT/RT have been described previously, to our knowledge the cytomorphologic features on intraoperative squash preparation has rarely been studied, with only rare case reports appearing in cytopathology literature [6,7]. Here in we report a case of AT/RT occurring in the posterior fossa of a 10 year old boy.

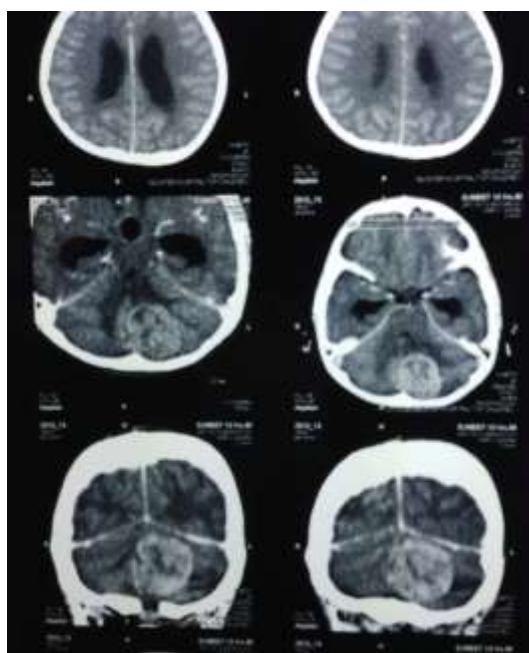
### CASE REPORT

A previously healthy 10 year old boy presented with complaints of headache, vomiting, fever, seizures and gait disturbance of 20 days duration. Headache was intermittent associated with non projectile vomiting. There were no cranial nerve palsies. On examination child was febrile and vital signs were stable. He was drowsy but arousable with Glasgow coma score of E3 V4 M6. His cranial nerves were intact and plantar reflexes were decreased. CT Brain was done with a suspicion of SOL in the brain. The results showed heterogeneously enhanced hypodense lesion with surrounding perilesional oedema noted in the cerebellar vermis and left cerebellar hemisphere causing compression over the 4th ventricle and dilatation of supratentorial ventricular system. Radiological

diagnostic possibilities of medulloblastoma and ependymoma were given.

Other routine investigations were normal. The patient underwent surgery and the tumour was sub totally excised as it infiltrated the surrounding brain tissue. Intraoperative squash cytology was requested. We received grey white, friable, moderately vascular tumour tissue H&E and toluidine blue stained squash cytosmeears showed large round to oval cells with eccentric nucleus, prominent nucleoli, distinct cell borders, along with huge bizarre and multinucleate forms admixed with sheets of round to oval cells with

scant cytoplasm and few spindle cells. Focal areas of necrosis were also seen. Squash diagnosis of AT/RT was given and the left over squash was sent for histopathological examination. CSF examination was done which was normal because children of this age group may have dissemination of this tumour at the time of diagnosis which is not seen in this case. AT/RT was confirmed on histopathological examination .On immunohistochemical studies tumour was vimentin,GFAP,Ki-67 positive, Desmin negative confirming the diagnosis. Patient was referred to a higher centre for further treatment.



**Fig. 1: CT scan shows a cerebellopontine(cp)angle tumour with heterogeneously enhancing hypodense lesion with surrounding perilesional edema noted in the cerebellar vermis and left cerebellar hemisphere causing compression over the 4th ventricle and dilatation of supratentorial ventricular system**

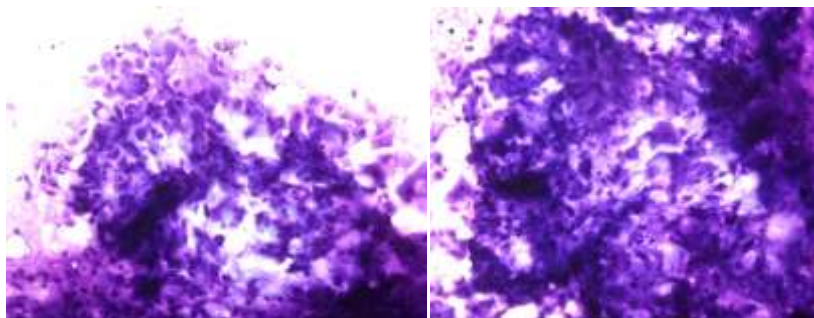
## DISSCUSSION

AT/RT is a rare primary malignant tumour of the CNS with an incidence of 2-3%.Two thirds of these cases occur in the pediatric age group with a slight male prepondance. The majority of the tumours occur in the posterior fossa. Data from AT/RT tumour registry estimates that approximately 20% of patients have dissemination at the time of presentation [8].Hence the prognosis is dismal. For an early preliminary diagnosis while the patient is still on table squash cytosmeears are an easy method to plan surgical treatment modality.

The common cytomorphological features of AT/RT on squash cytosmeears are hypercellular with tissue fragments and single cells (100%), perivascular aggregations around branching vessels or papillary

like(62%), large round and pleomorphic plasmacytoid cells(100%), rhabdoid cells with bright eosinophilic cytoplasm with or without globoid inclusions and eccentric nucleus (100%), primitive neuronal appearing cells(62%) ,pleomorphic multi nucleated giant cells, apoptosis, mitoses, necrosis [9].In the current case the cytomorphological features were corresponding with the features described above.

In our case cytologically, the smears were hypercellular with primitive appearing neoplastic cells admixed with intermediate sized rhabdoid cells in varying proportions. Therhabdoid cells showed pleomorphic eccentric nucleus, prominent nucleoli and conspicuous spherical cytoplasmic inclusions and apoptosis, mitoses, necrosis were also seen.



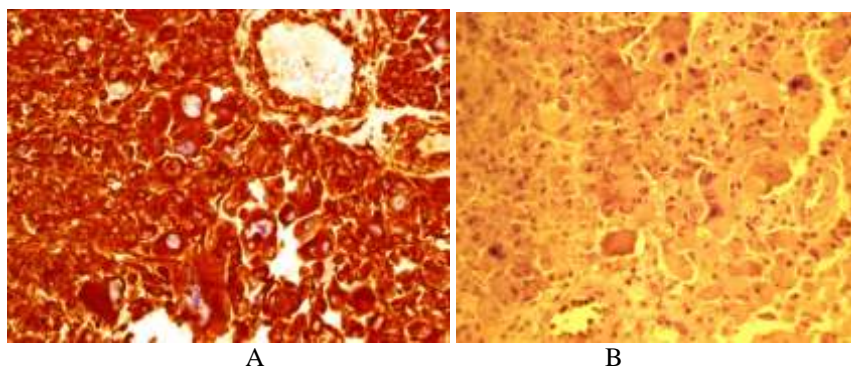
**Fig. 2: Squash cytospin smears of AT/RT (Toluidine Blue - 40x)**

Differential diagnosis entertained on cytology were PNET, Medulloblastoma and Giant Cell Glioblastoma.

Primitive neuroectodermal tumors show neuronal lineage, blastemal cells which are small to medium sized with scant perinuclear cytoplasm and hyperchromatic nuclei and abundant neutrophils in the background and true rosettes with high mitotic activity. Common sites being cerebral hemispheres but can also be seen in brainstem and cerebellopontine angle.

The other possibility being medulloblastoma which on cytology shows moderate cellularity with monolayered sheets of round to oval cells with moulded nuclei with hyperchromasia and perivascular pseudorosettes. In anaplastic variant cell wrapping i.e. one cell wrapped around another cell nucleus is seen. Medulloblastomas are negative for EMA expression.

Giant cell glioblastoma is extremely rare and occurs in younger age group. Squash smears are moderately cellular displaying malignant astrocytic tumor cells in cohesive clusters and dispersed population on a necrotic background. Most striking feature is numerous multinucleated giant cells which have more than 20 nuclei. The present case shows predominantly rhabdoid cells admixed with primitive neuronal component and few spindle cells, focal necrosis and absence of pseudopalisading. Hence a final diagnosis of AT/RT was given as cytomorphology combined with radiological finding of circumscribed mass with permeation of tumour into surrounding brain substance were in favour of it. Left over squash was processed and on histology tumor fulfilled the minimum diagnostic criteria of AT/RT i.e. presence of Rhabdoid cells with immunoreactivity with two or more of the antibodies i.e.; vimentin, EMA, GFAP, desmin.



**Fig. 3: Immunohistochemical stains: A) Vimentin: Strong Cytoplasmic Immunoreactivity  
B) Desmin: Negative**

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