



A RARE CASE OF CEREBELLOPONTINE ANGLE EMBRYONAL TUMOR

Neurosurgery

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ABSTRACT

Background: Embryonal tumors comprise a group of aggressive, poorly differentiated tumors occurring in central nervous system. Primary cerebellopontine angle (CPA) embryonal tumor is an extremely rare entity. It is important to have knowledge of this pathology and to be able to differentiate it from other commonly occurring CPA tumors, such as vestibular and trigeminal schwannomas. This distinction is essential because of the difference in the overall treatment plan and prognosis.

Case Description: This report describes a case of 4 year old girl who presented with headache and vomiting to our department. MRI findings suggested medulloblastoma and patient underwent surgery. Histopathology confirmed our diagnosis.

Conclusion: We discuss the importance of detecting this rare entity and distinguishing it from commonly occurring tumors in the CP angle region.

KEYWORDS

Cerebellopontine angle, embryonal, medulloblastoma, tumor, rare

INTRODUCTION

Embryonal tumors comprise a group of aggressive, poorly differentiated tumors occurring in central nervous system(1).They have been included in the 2016 WHO classification of tumors of the nervous system replacing the previous entity of primitive neuroectodermal tumors. They occur predominantly in the children and adolescents.

In the cerebellopontine angle (CPA) and internal acoustic meatus (IAM), these malignant tumors were shown to comprise only 0.1% of the cases.[2] The most common tumor in this location is the vestibular schwannoma (acoustic neuroma), meningioma, cavernoma, schwannomas of other cranial nerves, epidermoid and secondary deposits are also found.[3,4] Management of most CPA tumors can be planned on the basis of contrast-enhanced magnetic resonance imaging (MRI).

Here, we present a case of a young female with a CP angle tumor which turned out to be an embryonal tumor.

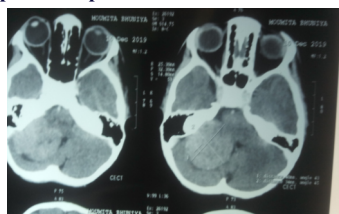
CASE HISTORY

A 4 year old girl presented to our department with complaint of headache and vomiting of 1 month duration. Neurological examination showed nothing significant.

She had a CT and MRI brain done which showed a large solid space occupying lesion in right CP angle location measuring 3.6*3.3*3.2 cm. The lesion was hyperdense on CT and MRI showed isointense lesion on T1 and T2 sequences with positive diffusion restriction. We had an MRI diagnosis of a possible medulloblastoma(MDB).



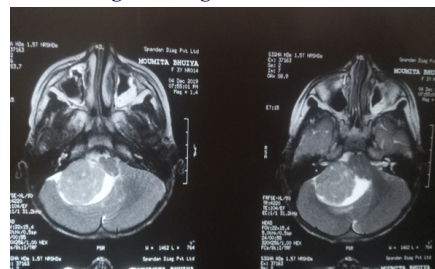
Preoperative picture of patient



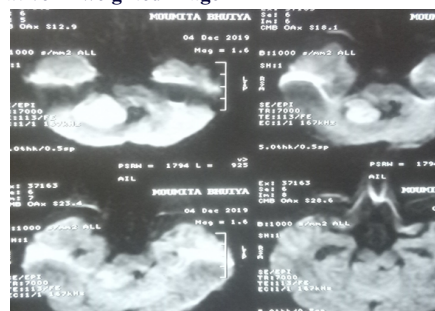
Preoperative CT Brain (plain) of the patient



Preoperative T1 weighted image



Preoperative T2 weighted image



Preoperative diffusion restriction image

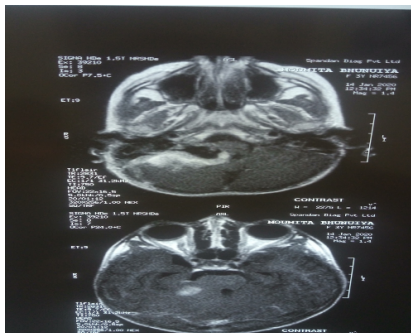
MANAGEMENT

The patient underwent a right retrosigmoid craniotomy. Intraoperatively, tumor was found to be solid, highly vascular and adherent to surrounding structures. The consistency was not conducive for removal by CUSA and was removed in piecemeal fashion. Considering the patient's age and presentation in the background of a preoperative diagnosis of MDB, we chose to do a subtotal excision. The patient recovered well with no new onset neurodeficit. Histopathology showed highly cellular lesion with small round cells

and hyperchromatic nuclei in a fibrillary background with a fair number of vascular septa suggestive of nodular variant of medulloblastoma.

Post operative MRI showed residual lesion. Spinal MRI was done to rule out drop metastasis.

Patient was referred to radiotherapy department for further management. She has been started on chemotherapy and is in regular follow up, doing well.



Postoperative MRI showing residual tumor



Postoperative picture of patient

DISCUSSION

Medulloblastoma (MDB) is the most common intracranial embryonal tumor and by definition arises exclusively in the cerebellum (5). It is a WHO grade IV tumor and is subclassified genetically into four types including WNT activated, SHH activated, group 3 and group 4 varieties.

Histologically, MDB can be classic variant, desmoplastic/nodular, extensive nodularity and large cell/anaplastic variant.

MDB's mostly arise in the midline (vermis) in children whereas involvement of hemispheres is common in adults. Extra-cerebellar sites are rare for MDB's.

Other types of embryonal tumors include

- Embryonal tumors with multilayered rosettes, C19MC-altered
- Embryonal tumors with multilayered rosettes, NOS
- Medulloepithelioma
- CNS Neuroblastoma
- CNS Ganglioneuroblastoma
- Atypical teratoid/rhabdoid tumor

In CNS, extraaxial CPA medulloblastomas and other PNETs are rare neoplasms. [6] A recent article reported two cases of CPA medulloblastomas which were preoperatively diagnosed as vestibular schwannoma and petrosal meningioma on radiological basis. These cases are comparable to ours. [7]

This rare presentation of the CPA tumor is important with regards to the differential diagnoses, due to the fact that most CPA tumors are usually considered to be vestibular or trigeminal schwannomas, and usually a gross total excision is planned for them.

However, as in our case the child clinically had no neurological findings and preoperative MRI suggested possibility of MDB, we

could change our approach to subtotal excision and remaining tumor to be managed with chemotherapy.

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