

CHOROID PLEXUS PAPILLOMA OF THE 3RD VENTRICLE- A CASE REPORTEzhil Arasi Nagamuthu¹, J. Anunayi², Syeda Iffath Tahseen³¹Professor and HOD, Upgraded Department of Pathology, Osmania Medical College, Hyderabad.²Associate Professor, Upgraded Department of Pathology, Osmania Medical College, Hyderabad.³Senior Resident, Upgraded Department of Pathology, Osmania Medical College, Hyderabad.**ABSTRACT****BACKGROUND**

Choroid Plexus Papillomas (CPPs) are rare intracranial neoplasms especially in the third ventricle. The most common site of presentation of these lesions is in the fourth ventricle in adults and lateral ventricles in children. Third ventricular lesion is uncommon, limited to a few case reports. These highly vascular tumours retain the physiological function of choroid plexus and thus lead to overproduction of Cerebrospinal Fluid (CSF) besides obstructing the pathway resulting in hydrocephalus. CT and MRI are the investigations of choice and are diagnostic. Surgical management vary according to the site of tumour and aim is complete excision of tumour. We present an interesting report of a 5 months old infant who presented with symptoms of raised intracranial pressure whose CT revealed third ventricular CPP. After ventriculoperitoneal shunt, tumour was excised. Pathological examination revealed choroid plexus papilloma.

KEYWORDS

Choroid Plexus Papilloma, Hydrocephalus, Third Ventricular Tumours.

HOW TO CITE THIS ARTICLE: Nagamuthu EA, Anunayi J, Syeda Iffath T. Choroid plexus papilloma of the 3rd ventricle: a case report. J. Evid. Based Med. Healthc. 2016; 3(87), 4777-4778. DOI: 10.18410/jebmh/2016/1005

BACKGROUND**Case Report**

A 5 months old male presented with increased head circumference and vomiting for 1 week. On general examination, the child was alert with downward gaze. CT and MRI demonstrated gross ventriculomegaly and enhancing mass lesion in third ventricle. The child underwent a ventriculoperitoneal shunt procedure followed by total excision of tumour. The histopathology of the resected tumour revealed tumour cells arranged in multiple papillae with fine fibrovascular core confirming the diagnosis of choroid plexus papilloma.

Choroid plexus tumours are rare intraventricular tumours accounting for less than 1% of all intracranial tumours and 2-4% of all brain tumours in children.^(1,2) Most of these tumours occurs in patients less than 2 years of age. Most common location is lateral ventricle followed by fourth ventricle. Sporadic case reports have described third ventricle choroid plexus papilloma.^(3,4,5,6) Diagnosis rests on imaging studies that show that mass in the location of third ventricle with typical imaging features on sonography, Computed Tomography (CT), Magnetic Resonance Imaging [MRI]. Treatment is complete surgical excision of tumour.

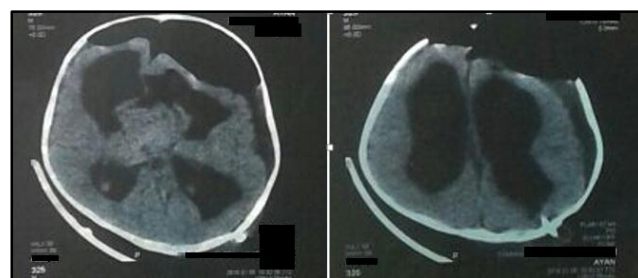


Figure 1. Axial Computed Tomography (CT) Images Reveal an Enhancing Lobulated Mass in the Third Ventricle (a) with Dilatation of the Lateral Ventricles (b)

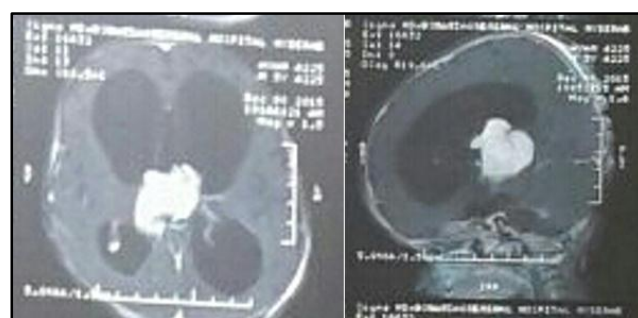


Figure 2. Axial Magnetic Resonance (MR) Images Contrast-Enhanced Axial (c) and Sagittal (d) Images Reveal Avid Enhancement within the Tumour and Extension into Fourth Ventricle

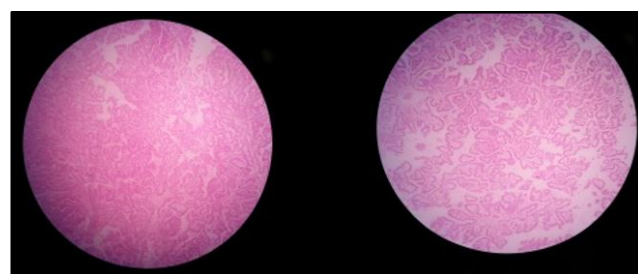


Figure 3. Low Power (a) and High Power (b) Photomicrograph Reveal Tumour Cells Arranged in Multiple Papillae with Fine Fibrovascular Core

Financial or Other, Competing Interest: None.
Submission 12-09-2016, Peer Review 18-09-2016,
Acceptance 15-10-2016, Published 29-10-2016.

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DOI: 10.18410/jebmh/2016/1005



Imaging plays a vital role in differentiating between obstructive (noncommunicating) and nonobstructive (communicating) hydrocephalus. Ultrasonography (USG) is the modality of choice in utero, neonatal and early infancy. CPPs appear as echogenic masses with lobulated outline. Their echogenicity and other morphological features closely match the normal choroid plexus. CT plays an important role in older children where the acoustic window for USG is lost. CECT depicts these lesions as avidly enhancing lobulated masses. The degree of heterogeneity is variable depending on the size of tumour, and if marked, should lead to a suspicion of CPC, particularly if there is adjacent invasion. MRI is the modality of choice in older children for central nervous system imaging in general and intracranial tumours in particular. This is related to the ability of MRI to provide multiparametric imaging affording a preoperative typing and grading of tumour, planning of biopsy and surgery.

DISCUSSION

CPP is a rare tumour of neuroectodermal origin accounting for less than 1% of all intracranial neoplasm.^[1] CPP is one of the neoplasm frequently observed to occur primarily within the ventricular system, but it rarely involves the third ventricle. In childhood, 80% of CPPs arises in lateral ventricle, 16% in the fourth ventricle and 4% in third ventricle. The incidence of third ventricular CPP is higher in the first decade of life with a female predominance, although overall, these tumours are more common in male subjects.⁽³⁾

The tumours typically present in neonatal or early childhood period pointing to their congenital origin. These lesions typically manifest with increasing head size, lethargy, decreased activity and poor psychomotor milestones in infants. As the child grows older, visual impairment and gait disturbances are noted related to raised Intracranial Pressure (ICP) and direct pressure caused by the tumour. Hydrocephalus and raised ICP in these cases caused by two mechanisms: Direct obstruction of CSF pathway and overproduction of CSF by tumour cells that tend to maintain function of choroid plexus. CPPs are histologically benign neoplasms derived from neuroectoderm assigned a WHO grade 1, grade 2 designation is reserved for atypical CPPs. Resection of these tumours is often curative with little chance of recurrence following gross total resection.

These tumours present special management challenges due to several unique characteristics including young age at presentation potential for CSF overproduction and the inherent vascularity of these lesions.^(2,7,8,9,10) Due to the rich vascular supply of these tumours, endovascular embolisation as a preoperative adjunct has been used in an attempt to reduce blood supply intraoperatively.^(9,11,12)

CONCLUSION

Management usually comprises of diversion procedure prior to definitive tumour excision. Complete removal of tumour is advocated and various approaches to third ventricular tumours include transcallosal, transfrontal, transforaminal routes and supracerebellar infratentorial.^[13,14]

In individual cases, the surgical approach would depend on location, size, vascularity and extension of tumour through foramen of Monro. Recently, microsurgical techniques have also been described to improve the outcome. Another innovative therapy is radiosurgery that aims at avascular necrosis and shrinkage of tumour and is best as an adjunct to surgery. CPPs have an excellent long-term survival after only gross total resection ranging from 90% to 100%.

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