

Case Report

Choroid Plexus Papilloma with Cytologic Differential Diagnosis – A Case Report

Pant I*, Chaturvedi S**, Suri V#, Dua R##

Abstract

The cytopathologic features of choroid plexus papilloma observed in a one year-old male child are reported and compared with other paediatric central nervous system neoplasms. The cytologic features of choroid plexus papilloma are similar to those of normal choroid plexus and may be difficult to distinguish from those of a well differentiated papillary ependymoma and other tumours with papillae formation. However, the salient cytologic features of choroid plexus papilloma include well defined papillary clusters, monolayered sheets and isolated scattered tumour cells, comprising of cuboidal cells with uniform round or oval nuclei, evenly dispersed chromatin and moderate amount of cytoplasm. The utility of crush cytology in the rapid diagnosis of central nervous system tumours along with the differential diagnoses are highlighted in this report.

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Introduction

Choroid plexus papillomas (CPP) are relatively rare tumours of the central nervous system. They occur most often in children, particularly during the first decade of life and constitute 3.9% of cerebral tumours in infancy and 2.3% of primary intracranial neoplasms of childhood.¹ Choroid plexus papillomas are confined to areas where choroid plexus is normally found, i.e. the lateral (50%), third (5%) and fourth (40%) cerebral ventricles, with two or three ventricles being involved in 5% of cases. The lesion tends to block cerebrospinal fluid pathways and patients accordingly present with signs of hydrocephalus (in infants increased circumference of the head) and raised intracranial pressure.

Intraoperative diagnosis of CNS lesions greatly aids in the surgical approach. Diagnosis of CPP on crush smears is, however, a challenge and very few cases have been reported. Cytologic features and

differentials in our patient are reported.

Case Report

A one-year-old boy presented with a five days history of irritability, increasing lethargy and vomiting. One day prior to admission the child had fallen asleep and was unarousable. The parents complained of a large head and delayed milestones. Physical examination revealed increased circumference of the head. Computed tomographic scan of the head revealed a hyperdense, contrast enhancing mass located in the cerebellum extending from the midline to the right side. The third and lateral ventricles were dilated and fourth ventricle was not identified separately. A provisional diagnosis of primitive neuroectodermal tumour was considered. Peroperatively a circumscribed mass was identified in the inferior cerebellar region, which was removed piecemeal.

On gross inspection, the specimen consisted of multiple reddish, granular, cauliflower like tissue bits together measuring 5.5x1.8x0.2 cm which were subjected for crush cytology and paraffin section study. For crush smears a tumour fragment about 1 mm³ in size was placed at one end of a glass slide and then divided into smaller bits, each of it was transferred to a new slide. The tissue was lightly crushed by another slide and was drawn smoothly along the length of the first slide. No resistance was felt while making the

* Lecturer, Department of Pathology, Advanced Medical and Dental Institute, Universiti Sains Malaysia; ** Associate Professor, #Assistant Professor, Department of Pathology, Institute of Human Behaviour and Allied Sciences, New Delhi; ##Neurosurgeon, Department of Neurosurgery, Guru Teg Bahadur Hospital, New Delhi.

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Corresponding Author : Dr Ishita Pant, Advanced Medical and Dental Institute, Universiti Sains Malaysia, No.29, Lorong Bertam Indah 4-9, Taman Bertam Indah, 13200 Kepala Batas, Seberang Perai Utara, Pulau Pinang, Malaysia.

E-mail: drishitapant@amdi.usm.edu.my

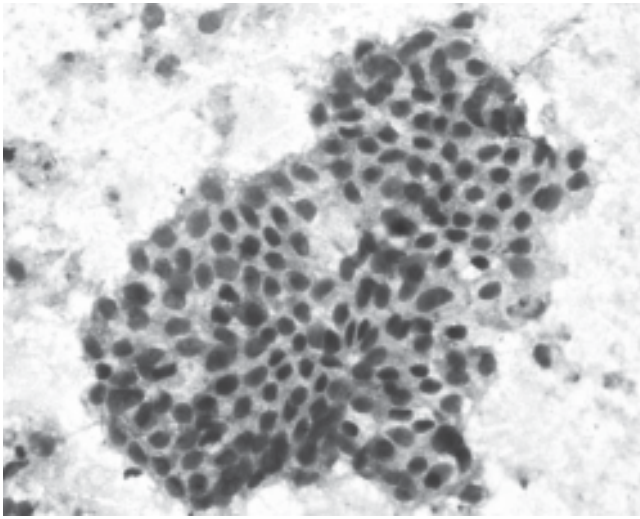


Fig. 1 : Crush cytology: salient cellular features of choroid plexus papilloma, showing round to oval uniform nuclei, with fine, evenly dispersed chromatin and ample cytoplasm (Toluidine blue, x 400).

smears. The smears were stained by the rapid toluidine blue method. The remaining tissue was fixed in 10% buffered formalin and routinely processed. Haematoxylin and eosin stained paraffin sections were observed under the light microscope.

Two toluidine blue smears were examined. Each smear was characterized by the presence of several papillary clusters, monolayered sheets and singly scattered tumour cells. The papillary clusters and monolayered sheets were composed of cuboidal cells with regular, round to oval nuclei with evenly dispersed chromatin and moderate amount of cytoplasm (Fig 1). There was no evidence of cellular atypia or mitosis. A diagnosis of choroid plexus papilloma (WHO grade I) was made independent of histology. Histopathology confirmed the diagnosis of choroid plexus papilloma (Fig 2).

Discussion

Choroid plexus papillomas develop in the ventricular system: in the fourth, lateral and third ventricles in that order of frequency. In children these tumours tend to develop most often in the lateral ventricles, whilst in adults the fourth ventricle is most commonly involved.

Histologically CPP is composed of delicate fibrovascular connective tissue fronds covered by a single layer of uniform cuboidal to columnar epithelial cells with round to oval basally situated monomorphic nuclei. Conspicuous mitotic activity, brain invasion and necrosis are absent.

A wide variety of lesions are relevant in the cytologic differential diagnoses of choroid plexus papillomas. Cells from normal choroid plexus may be indistinguishable from those seen in association with a choroid plexus papillomas.^{2,3} However, the papillary clusters and single cells are usually much more numerous in association with a CPP than expected in a smear from normal choroid plexus.

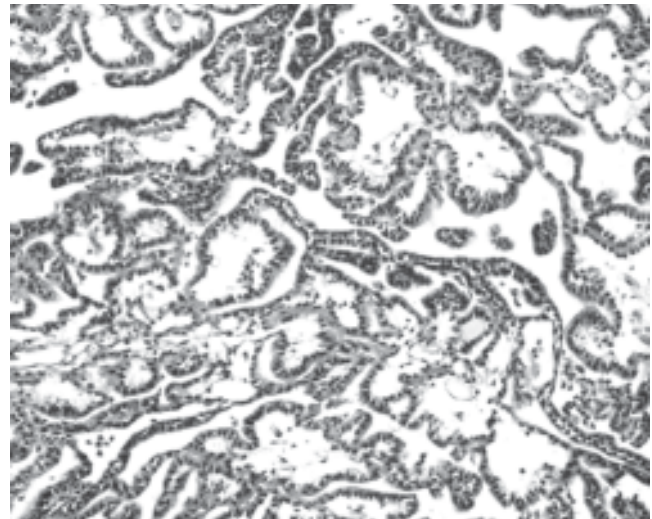


Fig. 2 : Histopathology of choroid plexus papilloma: numerous papillae having a thin, fibrovascular core covered with a single layer of benign cuboidal epithelium (H&E, x 100).

Villous hypertrophy is a diffuse enlargement of the choroid plexus in both lateral ventricles with normal histological appearances, often associated with hydrocephalus. Cytologically, it may be confused for CPP.⁴ Correlation with radiological features is advised when considering a diagnosis of choroid plexus papilloma.

Cells from a papillary ependymoma may be difficult to distinguish from those of a CPP. Generally both show relatively bland nuclei and abundant cytoplasm. The cell clusters of a papillary ependymoma may have a multilayered arrangement and frequently exhibit nuclear moulding in contrast to those of a choroid plexus papillomas.⁵ The histologic features of a neurofibrillary core in the papillary ependymoma is usually not identifiable on cytologic examination, however, if it is, it is a clinching diagnostic feature.

Primitive neuroectodermal tumours (PNET) also occur in this age group. While the cells in these tumours may be of a size similar to those seen in CPP, they typically reveal much more hyperchromatic nuclei with coarse chromatin. Nuclear moulding is another striking feature of cell clusters originating with PNET.

Finally choroid plexus carcinoma must be considered in the differential diagnosis. While rarer than CPP, two thirds of choroid plexus carcinoma occurs in infants, with a mean age of 2.5 years. Differential features include various sized nuclei with a high nuclear cytoplasmic ratio. Nuclear indentations and lobulations are often striking.⁶ Single or multiple micro-nucleoli are frequent. The cytoplasm is scanty, pale and granular.

To conclude, though the cytologic features of these paediatric tumours overlap, certain distinctive features always help to clinch the diagnosis, therefore it is of utmost importance to take into account the clinical and radiologic features of a case as well as the cytologic morphology.

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