

Choroid Plexus Papilloma of the Cerebello-pontine Angle

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Abstract

Choroid plexus papillomas are intraventricular papillary neoplasms, most commonly located in the lateral ventricles. We report a rare case of a choroid plexus papilloma of the cerebello-pontine angle in a 37 year-old lady who presented with a six-month history of worsening headache and blurring of vision, associated with cerebellar signs. Magnetic resonance imaging showed a tumour located at the right cerebello-pontine angle associated with obstructive hydrocephalus. The tumour was partially resected and an external ventricular drain inserted. Histopathological examination revealed delicate papillomatous fibrovascular fronds covered by a single layer of uniform columnar epithelial cells, confirming a choroid plexus papilloma. The postoperative period was complicated by intracerebral haemorrhage, left hemiparesis, hypertension and sepsis. She also suffered from persistent hydrocephalus attributed to presence of tumour remnants obstructing CSF flow. This rare case of choroid plexus papilloma of the cerebello-pontine angle is presented with a review of the literature.

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tumour was removed and an external ventricular drain placed.

INTRODUCTION

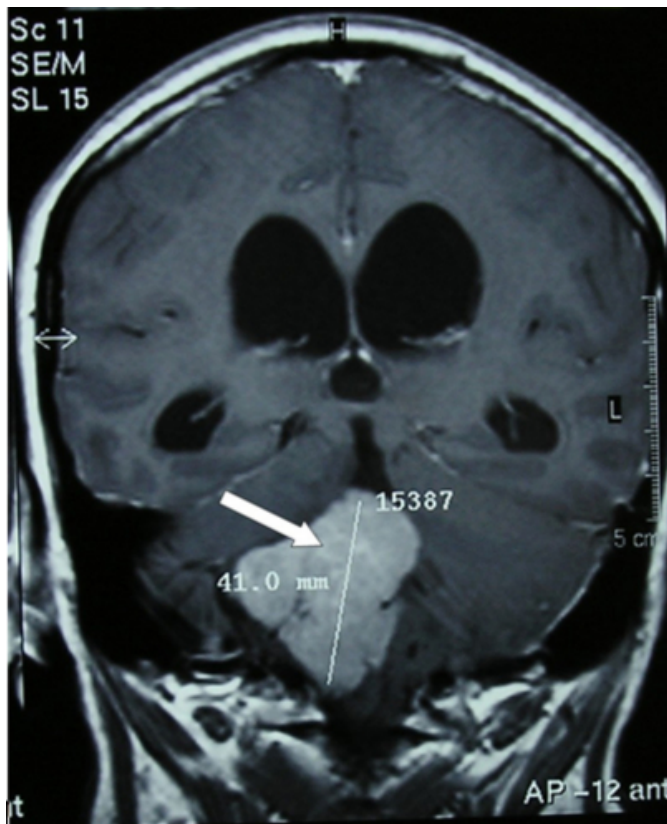
Choroid plexus tumours are intraventricular papillary neoplasms derived from choroid plexus epithelium and account for 0.4 – 0.6% of all brain tumours. Choroid plexus papillomas (CPP) outnumber choroid plexus carcinomas (CPC) by a ratio of at least 5:1. Choroid plexus papillomas are confined to lateral (50%), third (5%), and fourth (40%) ventricles with two or three ventricles involved in 5% of cases¹. Choroid plexus papilloma of the cerebello-pontine angle is rare and this is the first reported case in Malaysia.

CASE REPORT

A 37 year-old lady presented with a six-month history of worsening headache and blurring of vision. On examination she had cerebellar signs but no evidence of cranial nerve deficit. Magnetic resonance imaging (MRI) showed a tumour located at the right cerebello-pontine angle and measured 3.0 x 3.9 x 4.1 cm, associated with obstructive hydrocephalus (Figure 1). A provisional diagnosis of acoustic schwannoma was made. An atlanto-suboccipital craniotomy was performed and the tumour approached via the midline. The tumour was vascular and the medial part was adherent to blood vessels. Approximately 70% of the

Figure 1

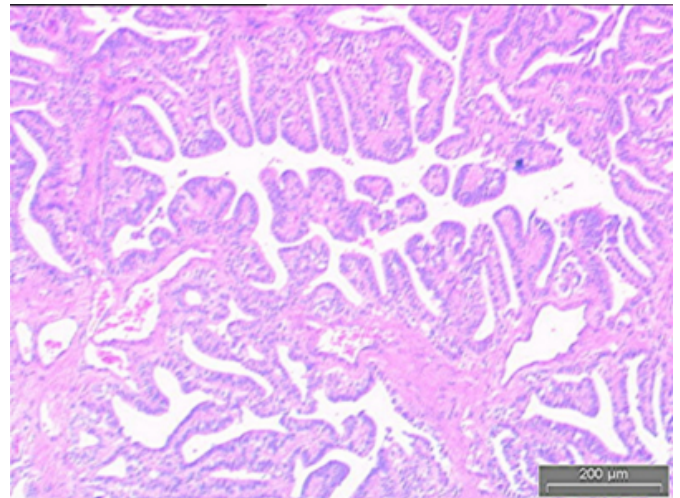
Figure 1. MRI - Coronal view of T1 weighted image with gadolinium infusion, shows a tumour at the cerebello-pontine angle.



On macroscopy the tumour consisted of multiple brownish tissue fragments, and measured 35 mm in aggregate diameter. Histopathological examination revealed delicate papillomatous fibrovascular fronds covered by a single layer of uniform columnar epithelial cells with oval basally located nuclei. Mitotic figures, cellular atypia and necrosis were absent (Figure 2). Immunohistochemically, cytokeratin, vimentin and S-100 were expressed while epithelial membrane antigen (EMA) and glial fibrillary acidic protein (GFAP) were negative.

Figure 2

Figure 2: Histopathology of choroid plexus papilloma exhibiting delicate papillary fronds, H&E stain, x 40.



About a month postoperatively the patient suffered an episode of intracerebral haemorrhage with acute stroke, left hemiparesis and uncontrolled hypertension. She was managed accordingly and her condition stabilised. She was discharged from the ward a month later, with a ventriculo-peritoneal shunt implant, but was subsequently lost to follow-up.

DISCUSSION

Choroid plexus papilloma (CPP) arising in the cerebello-pontine angle is a rare entity. CPPs of the cerebellopontine angle are thought to originate from the choroid plexus of the fourth ventricle that extends through the foramen of Luschka, known as Bochdalek's basket. Most literature is in the form of single case reports though a few case series have been published. Through a pubmed search, this is the first reported case of choroid plexus papilloma of the cerebello-pontine angle in Malaysia.

In a meta-analysis study of 566 cases of choroid plexus tumours in Canada, Wolff et al. reported that in children, tumours were most often localized to the supratentorial region, whereas the most common sites in adults were the fourth ventricle and cerebello-pontine angle. Cerebello-pontine angle tumours were also noted to be more common in the female gender and had an associated benign histology

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Symptoms and signs of raised intracranial pressure and those of typical cerebello-pontine angle (CPA) syndrome^{3, 4} represent the most common presentation, as the tumour blocks cerebrospinal fluid flow and causes hydrocephalus, as

was noted in our patient. The average duration of symptoms before presentation was reported to be around 21 months in a 12 patient series ³. With regards to clinical signs, the most frequent clinical signs of CPPs located in the cerebello-pontine angle include VIIth and VIIIth cranial nerve deficits, cerebellar signs, gait ataxia and visual disturbances. Our patient had cerebellar signs but no evidence of cranial nerve deficits.

Patients are often misdiagnosed clinically as having acoustic schwannoma, meningioma or ependymoma, as the associated clinical features and radiology appear nonspecific. Pre-operative neuroimaging investigations such as computed tomography (CT), magnetic resonance imaging (MRI) and angiography may provide clues to the diagnosis ^{3, 5, 6, 7}. CPPs typically appear as an isodense homogenous enhancing tumour on CT ⁶. On T₁-weighted MR images, CPPs are described as hypointense or isointense with gray matter, whereas on T₂-weighted images, they have variable signal ^{6, 7}. MRI with gadolinium contrast produces intense enhancement of the tumour, corresponding to its vascular nature ^{3,9}, as is clearly noted in our patient.

Surgical resection of the tumour with histopathological examination is necessary for diagnosis. A midline approach via the cerebellomedullary fissure or retromastoid routes are two options mentioned in literature though neither are described to be superior to the other ³. Choroid plexus papillomas exhibit a delicate villous architecture attached to the wall of the ventricle that it arises from. Haemorrhage, necrosis and brain invasion are often associated with carcinomas. One of the problems commonly encountered during surgery is tumour bleeding and hence, various procedures such as microembolization have been proposed to reduce the vascularity of CPPs preoperatively. Adherence of the tumour to the brainstem and cranial nerves is another hindrance to complete removal of tumour tissue.

Histopathologic examination of the resected tumour was typical of CPP which corresponds to WHO grade I tumours of the nervous system. Papillary fronds lined by bland columnar epithelium are characteristic features. The absence of mitotic activity, nuclear pleomorphism and necrosis distinguishes it clearly from atypical choroid plexus papilloma and carcinoma ¹. Villous hypertrophy can be excluded in view that it is a non-neoplastic, diffuse enlargement of choroid plexus in both lateral ventricles and often manifests as hypersecretory hydrocephalus.

Expression of cytokeratin, vimentin and S-100 are well

documented in CPP. The absence of EMA and GFAP further support its diagnosis. It is interesting to note that there is variable expression of S-100, vimentin and GFAP in CPP compared to CPC. There is more S-100 and transthyretin (TTR) tumour positive cells in CPP compared to less frequent staining in CPCs. On the other hand GFAP may be focally seen in some cases of CPP but positive in about 20% of CPC. It has been reported that fourth ventricle tumours express more S-100 than lateral ventricle tumours and older patients (above 20 years) express more GFAP and TTR than younger patients ⁸. Immunohistochemical expression of p53 expression was reported to be consistently positive in choroid plexus carcinoma but undetected in the majority of CPPs ⁹, proving the role of gene mutation in cancer genesis.

Chromosomal aberrations have been reported in CPP though it was not done in our case. Rickert et al. reported that 32 of 34 choroid plexus papillomas were associated most commonly with gains on chromosomes 5, 6, 7, 9, 15 and 18. These aberrations differed from choroid plexus carcinoma as well as between paediatric and adult choroid plexus papillomas, supporting the notion of different genetic pathways ¹⁰. Chromosomal analysis may also aid in prognostication of patients.

Choroid plexus papillomas are associated with favourable long-term prognosis. Factors that correlate with prognosis are often related to tumour size, preoperative symptoms and the difficult surgical intervention related to its anatomical location.

Radical tumour resection is the main modality of treatment for CPP. Lesions that are removed entirely carry a good prognosis though this is not achievable in all cases due to the factors described above. Wolff et al. reported a 10-year survival rate of 85% in CPP patients who had complete tumour removal ². Most clinicians advocate a 'wait and see' approach after gross total resection of CPP. Improvement in surgical techniques and surgery after care has improved the prognosis of patients. Recurrences are occasionally seen and rare cases of metastases to suprasellar and even craniospinal seeding have been reported ¹¹. While some patients with residual tumour mass or recurrence may show response to conventional or stereotactic radiotherapy, its effectiveness is yet to be established ^{2, 3, 4}.

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