

Case Report

Adult-onset fourth ventricular choroid plexus papilloma: Telovelar approach and post-operative outcome

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ABSTRACT

Choroid plexus papillomas (CPPs) are rare, benign intraventricular tumours, predominantly affecting children. Adult-onset CPPs are uncommon but usually arise in the fourth ventricle and present with features of obstructive hydrocephalus and cerebellar signs. We report a case of a 42-year-old male with progressive truncal ataxia, raised intracranial pressure-type headache, and visual decline. Imaging revealed a posterior fossa mass causing obstructive hydrocephalus. Differential diagnoses included ependymoma, medulloblastoma and dermoid cyst. The patient underwent midline suboccipital craniotomy with near-total tumour excision, with intraoperative external ventricular drainage, followed by ventriculoperitoneal shunt placement later. Histopathology confirmed CPP. Postoperative complications included lower cranial nerve dysfunction requiring tracheostomy. The patient gradually improved and was successfully decannulated. At 6-month follow-up, he had a stable small residual tumour with no progression. Although rare in adults, CPP should be considered in the differential diagnosis of posterior fossa tumours presenting with cerebellar signs and raised ICP. Surgical excision remains the mainstay of treatment, with the extent of resection being a key prognostic factor. Near-total resection may be appropriate when tumour adherence to critical structures risks significant morbidity, and continued radiological follow-up is essential to monitor for recurrence.

Keywords: Adult onset, Cerebellar signs, Choroid plexus papilloma, Hydrocephalus, Posterior fossa tumor

INTRODUCTION

Choroid plexus papillomas (CPPs) are benign epithelial neoplasms arising from the choroid plexus epithelium, accounting for approximately 0.4–0.6% of all intracranial tumors.^[1] They are most frequently encountered in children, with the lateral ventricles being the commonest location in pediatric cases, whereas in adults, the fourth ventricle predominates.^[2,3] Adult-onset CPPs are uncommon, but when present, they typically manifest with symptoms of obstructive hydrocephalus, such as headache, vomiting, papilledema, and cerebellar signs.^[2-4] Neuroimaging often reveals a vividly enhancing intraventricular mass, and the diagnosis is confirmed histopathologically. The mainstay of treatment is surgical excision, with gross total resection (GTR) offering the best long-term control.^[5] We present a rare case of adult-onset CPP of the fourth ventricle, highlighting surgical management and post-operative course, along with a review of relevant literature.

CASE REPORT

Presentation

A 42-year-old male presented with a 4-month history of progressive headache, gait instability, and visual blurring. The

headache was dull, diffuse, and associated with intermittent vomiting. Neurological examination revealed truncal ataxia, mild limb dysmetria, and bilateral papilledema. No focal motor or sensory deficits were noted.

Investigations

Contrast computed tomography head [Figure 1a] displays a lobulated posterior fossa mass with heterogeneous enhancement, and contrast magnetic resonance imaging (MRI) brain [Figure 1b-d] revealed a well-circumscribed, lobulated mass in the fourth ventricle displaying iso-hyperintense signal on T2-weighted images, with heterogeneous contrast enhancement and a patchy area of diffusion restriction.

The lesion caused obstructive hydrocephalus with upstream dilatation of the ventricles and transependymal seepage. Differential diagnoses included ependymoma, medulloblastoma, and dermoid cyst.

Surgical management

The patient underwent midline suboccipital craniotomy via a telovelar approach. Intraoperative findings revealed a highly

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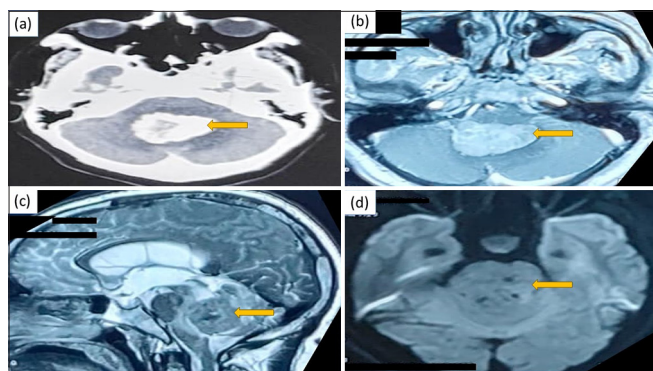


Figure 1: (a) Contrast-enhanced computed tomography showing a lobulated posterior fossa mass (yellow arrow) with heterogeneous enhancement. (b) Gadolinium-enhanced magnetic resonance imaging brain (axial view) showing a well-defined lobulated mass in the fourth ventricle with heterogeneous enhancement (yellow arrow). (c) T2-weighted imaging (sagittal view) showing the mass displaying iso- to hyperintense signal (yellow arrow). (d) Diffusion-weighted imaging sequence showing the tumour exhibiting patchy diffusion restriction (yellow arrow).

vascular, friable tumor arising from the choroid plexus of the fourth ventricle, closely adherent to the floor. Near-total resection (NTR) was achieved, leaving a small fragment adherent to the brainstem to avoid neurological morbidity. An external ventricular drain was placed intraoperatively.

Histopathological examination [Figure 2] revealed papillary fronds lined by a single layer of uniform cuboidal epithelium overlying fibrovascular cores, consistent with CPP (World Health Organization [WHO] Grade I). Immunohistochemistry showed positivity for cytokeratin and transthyretin.

Postoperatively, the patient developed lower cranial nerve palsies (IX, X, XII), causing dysphagia and aspiration, necessitating tracheostomy. A ventriculoperitoneal shunt was placed on post-operative day 10 due to persistent hydrocephalus.

Outcome and follow-up

The patient gradually improved with physiotherapy and swallowing rehabilitation. The tracheostomy was successfully decannulated at 3 months. At 6-month follow-up, MRI revealed a stable residual tumor with no progression, and the patient remained functionally independent with mild residual cranial nerve deficits.

DISCUSSION

CPPs are rare, benign intraventricular neoplasms (WHO Grade I) that constitute approximately 0.4–0.6% of all brain tumors and 2–4% of pediatric intracranial tumors, but fewer than 10% occur in adults.^[1,2] Adult-onset CPPs arising in

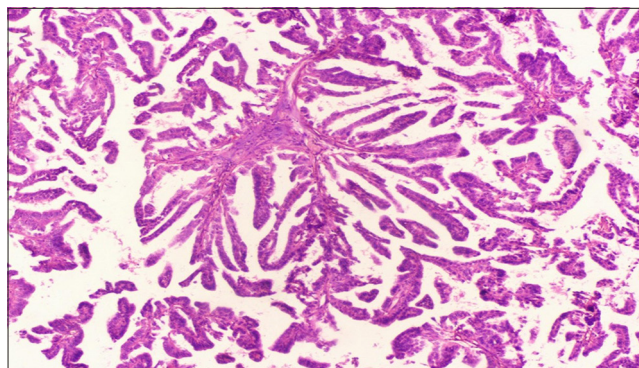


Figure 2: Histopathology: papillary arrangement of tumour cells with fibrovascular core, minimal atypia (Hematoxylin and Eosin, $\times 100$).

the fourth ventricle are particularly uncommon and can mimic other posterior fossa tumors such as ependymoma or medulloblastoma, making pre-operative diagnosis challenging.^[3,4]

In our patient, the clinical presentation with features of raised intracranial pressure and cerebellar signs was consistent with a posterior fossa mass obstructing the cerebrospinal fluid pathway. Neuroimaging revealed a fourth ventricular lesion with obstructive hydrocephalus. While CPPs in children more commonly arise in the lateral ventricles, adult CPPs have a predilection for the fourth ventricle.^[5,6]

Surgical considerations

Complete microsurgical excision remains the treatment of choice for CPPs.^[7] In the fourth ventricle, two principal intradural approaches following a midline suboccipital craniotomy are the telovelar and transvermian routes. The telovelar approach, which we used, provides wide exposure without splitting the vermis, thus reducing the risk of post-operative cerebellar mutism and truncal ataxia.^[8] In our case, an NTR was performed due to dense adherence of the tumor to the floor of the fourth ventricle, where aggressive dissection would have risked significant neurological morbidity. Literature supports maximal safe resection over GTR in such situations.^[9,10] In our setting as a tertiary care center, where some patients may face individual socioeconomic or logistical barriers to prolonged post-operative intensive neuro-rehabilitation, the decision to perform NTR rather than pursue aggressive dissection was aimed at minimizing the risk of additional lower cranial nerve injury. Despite this precaution, a post-operative lower cranial nerve deficit occurred, likely attributable to the tumor's dense adherence to the brainstem. This approach reflects a pragmatic balance between oncological control and functional preservation, prioritising patient quality of life and regular post-operative surveillance over immediate radiological clearance.

Table 1: Summary of reported adult fourth ventricular choroid plexus papilloma cases, including surgical approach, extent of resection, and clinical outcomes.

| Author, Year | Age/Sex | Location | Surgical approach | Extent of resection | Follow-up (months) | Outcome |
|--|---------|------------------|--|---------------------|--------------------|--|
| Adib <i>et al.</i> , 2000 ^[5] | 44/F | Fourth ventricle | Midline suboccipital craniotomy (intradural route not specified) | GTR | 12 | No recurrence |
| McEvoy <i>et al.</i> , 2000 ^[6] | 37/M | Fourth ventricle | Midline suboccipital craniotomy (intradural route not specified) | GTR | 24 | No recurrence |
| Present case | 42/M | Fourth ventricle | Midline suboccipital craniotomy (Telovelar) | NTR | 6 | Stable residual; postop lower CN deficit |

CPP: Choroid plexus papilloma, GTR: Gross total resection, NTR: Near-total resection, CN: Cranial nerve, F: Female, M: Male

Recurrence risk and follow-up

Residual CPP carries a measurable risk of recurrence, though generally slow-growing. Recurrence rates for incompletely resected CPPs have been reported between 10% and 29%.^[11] Close imaging surveillance is therefore recommended. Our patient is scheduled for an MRI every 6 months for the first 2 years and annually thereafter, in line with published follow-up protocols.^[11] Importantly, recurrence after NTR in benign CPPs is often slow-growing and, when detected early on surveillance imaging, can be addressed with delayed re-operation or alternative treatments, minimizing the risk of adverse outcomes over time.

Post-operative morbidity

Lower cranial nerve palsy is a recognised complication of fourth ventricular tumor surgery, particularly when the tumor is densely adherent to the brainstem. In our patient, post-operative cranial nerve IX–X dysfunction caused significant swallowing impairment with microaspiration detected on bedside evaluation. To prevent progression to full-blown aspiration pneumonia, a prophylactic tracheostomy was performed early in the post-operative period. The patient received intensive swallowing and speech rehabilitation, with gradual improvement permitting decannulation at 3 months. At 6-month follow-up, swallowing was adequate for oral intake with only occasional cough on liquids, and voice quality had returned close to baseline. In comparative series, the telovelar approach has been associated with a lower incidence of post-operative lower cranial nerve deficits compared to the traditional transvermian approach, with rates reported between 5% and 15% versus 15–30%, respectively.^[12] Our patient's deficit likely reflects the tumor's adherence to the brainstem rather than the exposure route itself, reinforcing that complication risk is multifactorial.

Prognosis in adult CPP

The prognosis for CPP is generally excellent after maximal safe resection, with 5-year survival rates exceeding 90%.^[13] Recurrence risk increases with subtotal resection and in atypical CPP (WHO Grade II).^[14] Although our case involved a benign CPP, the residual tumor mandates long-term vigilance to detect regrowth early and consider re-intervention if needed.

Adjuvant therapy, such as radiotherapy or chemotherapy, is generally not indicated for WHO Grade I CPPs after adequate surgical resection, but may be considered in atypical CPPs or when resection is incomplete and recurrence is documented.^[15]

Table 1 summarises reported adult fourth ventricular CPP cases, including the present case along with the extent of resection and outcomes. Our patient underwent NTR with stable residual at 6-month follow-up and minimal residual deficits.

Two of the cited cases documented only “midline suboccipital craniotomy” without specifying the intradural route. Based on operative descriptions, these were likely transvermian approaches; however, as the route was not explicitly stated, they are marked as “not specified” to avoid misrepresentation.

CONCLUSION

Adult-onset CPPs of the fourth ventricle are rare and can mimic other posterior fossa tumors. Surgical excision remains the mainstay of treatment, with the extent of resection being the most significant prognostic factor. NTR may be appropriate when adherence to critical structures is present. Continued radiological follow-up is essential to monitor for recurrence or progression.

Ethical approval: Institutional Review Board approval is not required.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed

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