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Suboccipital Craniotomy and C1 Laminectomy for Atypical Choroid Plexus Papilloma

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Abstract

Atypical choroid plexus papilloma is a rare pediatric brain tumor that has distinct clinical and pathologic features. In this case, we highlight the diagnosis and management of this rare disease. The details of case positioning and execution are discussed. The case review is utilized as an overview of histopathologic findings, to discuss clinical features of the disease, and to highlight areas warranting further investigation. In particular, we provide insight into the typical clinical course post-treatment.

Keywords

Atypical Choroid Plexus Tumor; Surgical resection; Follow up

Background

Choroid plexus tumors (CPTs) are rare tumors of neuroectodermal origin, accounting for only 0.4–0.6% of all pediatric intracranial neoplasms(1). While CPT can occur in any age group, the median age of diagnosis is 3.5 years(2). CPT in children is typically supratentorial within the lateral ventricles whereas in adults, the fourth ventricle is the most common location(2). While gross total resection is the treatment of choice with cure rates approaching 100%(2), radiosurgery(3), bevacizumab(4), and adjuvant chemotherapy(5) and radiation are also possible treatment options in certain clinical scenarios. In this case report, we highlight a unique CPT in the fourth ventricle of a 2-year-old.

Case Presentation

A 2-year-old previously healthy male presented with two months of progressively worsening non-bilious, non-bloody emesis. The episodes occurred daily in the morning and after meals and were associated with increased fussiness, gait abnormalities, anorexia, and a five-pound weight loss. He was first evaluated by a pediatric gastroenterologist. Due to concerns about his associated behavioral changes and the positional nature of his symptoms, an MRI was

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ordered. Shortly thereafter, the patient then presented to the ED for intractable vomiting. The ordered MRI was completed at that time which revealed a large fourth ventricular tumor with obstructive hydrocephalus, most concerning for a medulloblastoma (Figure 1). Neurosurgery was consulted and initial assessment revealed no neurologic deficits. He was then transferred to the pediatric ICU for hourly neuro-checks and started on decadron. After discussion with family, they elected to undergo tumor resection.

Approach and Operation

The patient was positioned prone on a Mayfield gel horseshoe. A linear suboccipital incision was made from theinion to the spinous process of C2 and carried deep using Bovie electrocautery down to the suboccipital skull and the posterior ring of C1 until there was sufficient visualization to perform the bony opening. A pair of burr holes was made and a craniotome bit was used to perform the lateral craniotomy down to the foramen magnum and across the bottom of the skull. This ultimately created a suboccipital boneflap that was then removed. A C1 laminectomy was performed to complete the bony opening and dura was incised in a standard Y-shaped fashion. Operating microscope was used for the entire tumor resection. The tumor was hollowed out using a Sonopet on 40/50/8 and the adherent tumor capsule was carefully dissected from the cerebellar tonsils, vermis, and floor of the fourth ventricle. Several specimens were taken for pathologic evaluation. Progress of debulking was monitored using the Aloka ultrasound. The cerebral aqueduct was uncovered which restored normal CSF flow. Despite focally poor planes with regards to the brainstem and areas of the lateral walls of the fourth ventricle, a gross total resection was achieved with gradual and careful dissection. Excellent hemostasis was achieved, cavity was irrigated, dura was closed, bone flap was replaced, and the wound was closed in layers. The skin was closed with a running 5-0 Monocryl.

Post-Operative

The patient remained intubated post-operatively. No EVD or drains were required. He was sedated for post-operative MRI which revealed improved hydrocephalus and some enhancement adjacent to the resection cavity likely related to postsurgical reactive changes rather than residual tumor. He was discharged without difficulty 3 days post-surgery.

Pathology

On gross examination, the tumor appeared as a large, soft, friable, pink mass. Microscopic examination revealed a neoplasm with predominantly papillary (Figure 2) and focal cribriform (Figure 3) architecture. Immunohistochemical features of the tumor were consistent with a diagnosis of CPT (Figure 4). The tumor demonstrated up to 4 mitoses per 10 high power fields and an elevated ki-67 proliferation index (Figure 5) consistent with atypical choroid plexus papilloma, WHO Grade II.

Outcome

Three- and six-month follow-up imaging revealed no new lesions, but there was a small area of slightly enlarging enhancement noted at the fourth ventricular roof representing scar

tissue versus residual tumor which is being followed closely with serial imaging (Figure 6). Clinically, the patient has improved significantly without seizures or neurological deficits. No further episodes of emesis and no signs of hydrocephalus.

Discussion

Historically, choroid plexus tumors (CPT) have been classified by the World Health Organization (WHO) as either being benign or malignant based on certain histological criteria. Choroid plexus carcinoma (CPC) has high mitotic activity, high degree of cellularity, necrosis, and often invasion into the brain parenchyma while the benign choroid plexus papilloma (CPP) lacks such frank features of malignancy. Differentiating between CPC and CPP has proven challenging in some cases which prompted the creation of a third category which represents a small subset of CPT—atypical choroid plexus papilloma(6).

This case is unique as it represents a rare subtype of an already rare tumor in a location that is uncommon in the pediatric patient population. Despite the radiographic evidence favoring medulloblastoma, a malignant tumor with a guarded prognosis, pathology confirmed that this patient actually had a much more benign, albeit rare, diagnosis of an atypical choroid plexus papilloma. Although CPT are typically supratentorial in children, our patient's tumor was located infratentorial within the fourth ventricle. Pathologic hallmarks of choroid plexus papillomas include papillary fronds lined by bland columnar epithelium; positive staining for cytokeratin, transthyretin, and GFAP; and absence of nuclear pleomorphism and necrosis. These were all features displayed in our patient's tumor specimens. Choroid plexus papillomas with greater than two but fewer than five mitotic figures per high power field are characterized as atypical, as was seen in our patient. Further research is warranted in regards to follow up chemotherapy and surveillance needed for these patients.

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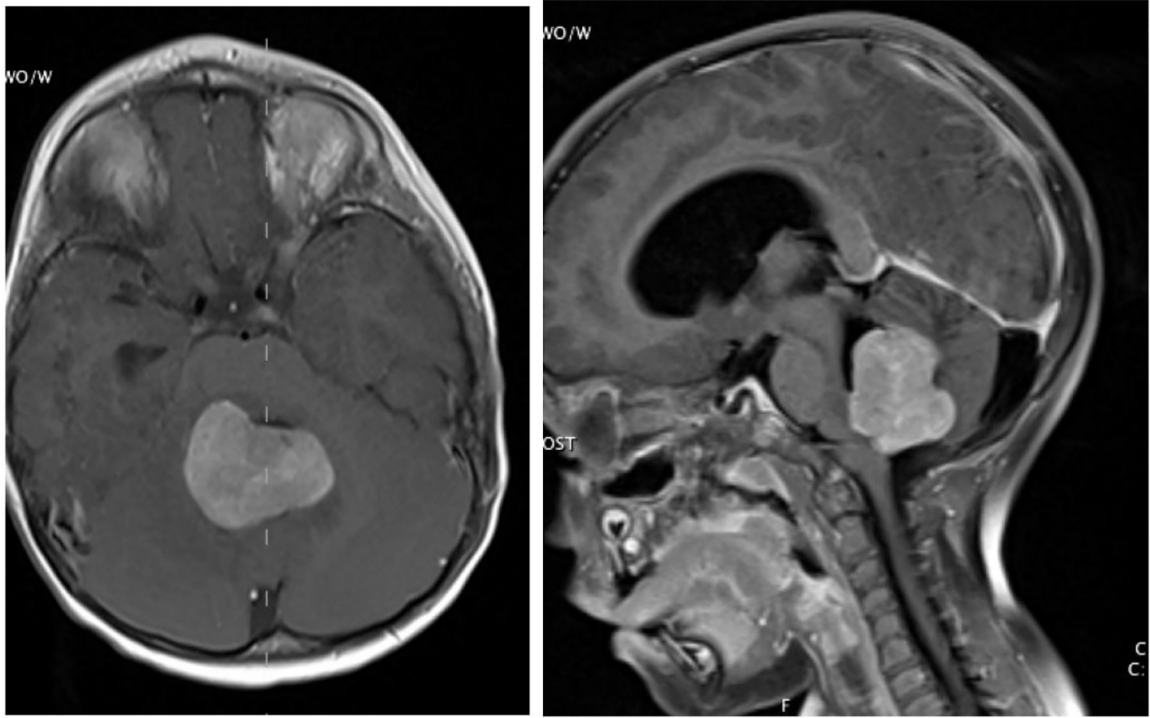


Figure 1:
Contrast enhancing mass in posterior fossa on MRI. **A.** Axial view, **B.** Sagittal view

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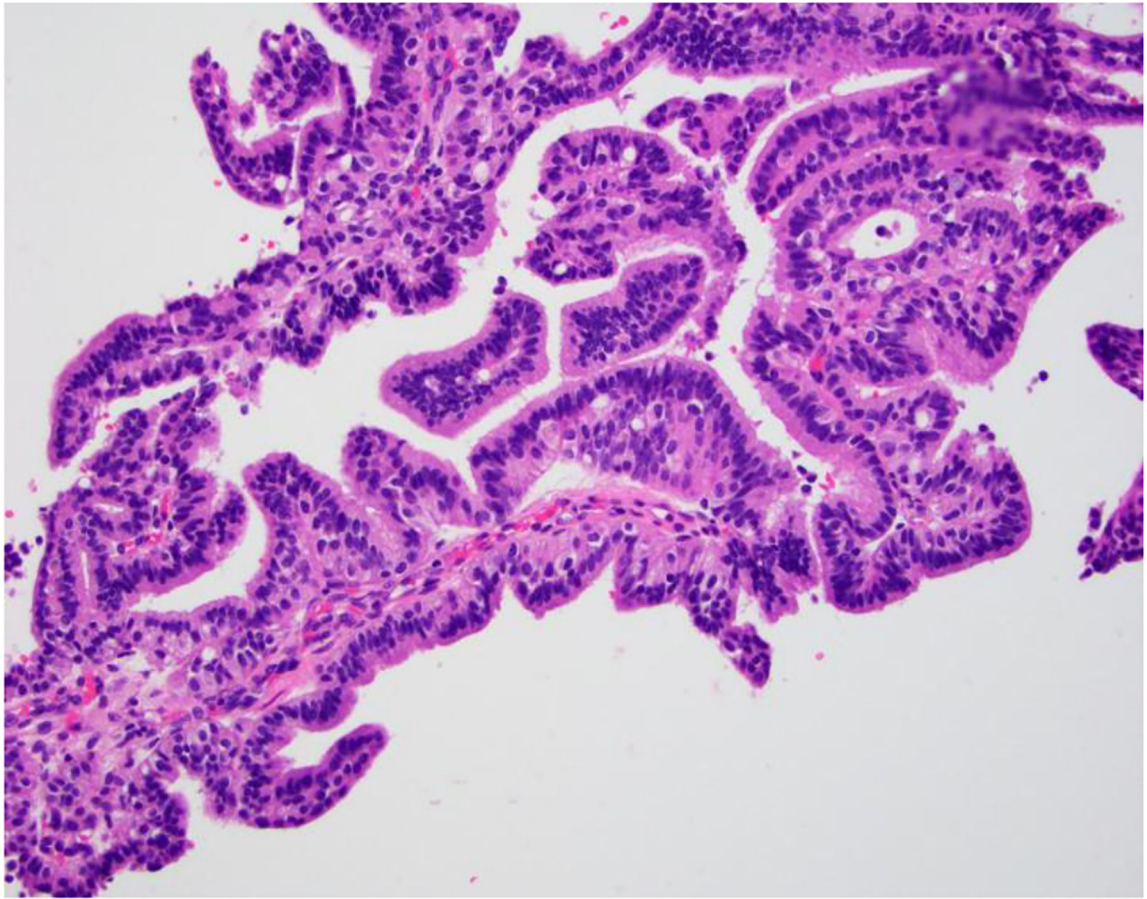


Figure 2:
H&E staining of tumor sections revealed a papillary neoplasm consisting of fibrovascular cores covered by a single layer of columnar epithelial cells showing little to no nuclear pleomorphism.

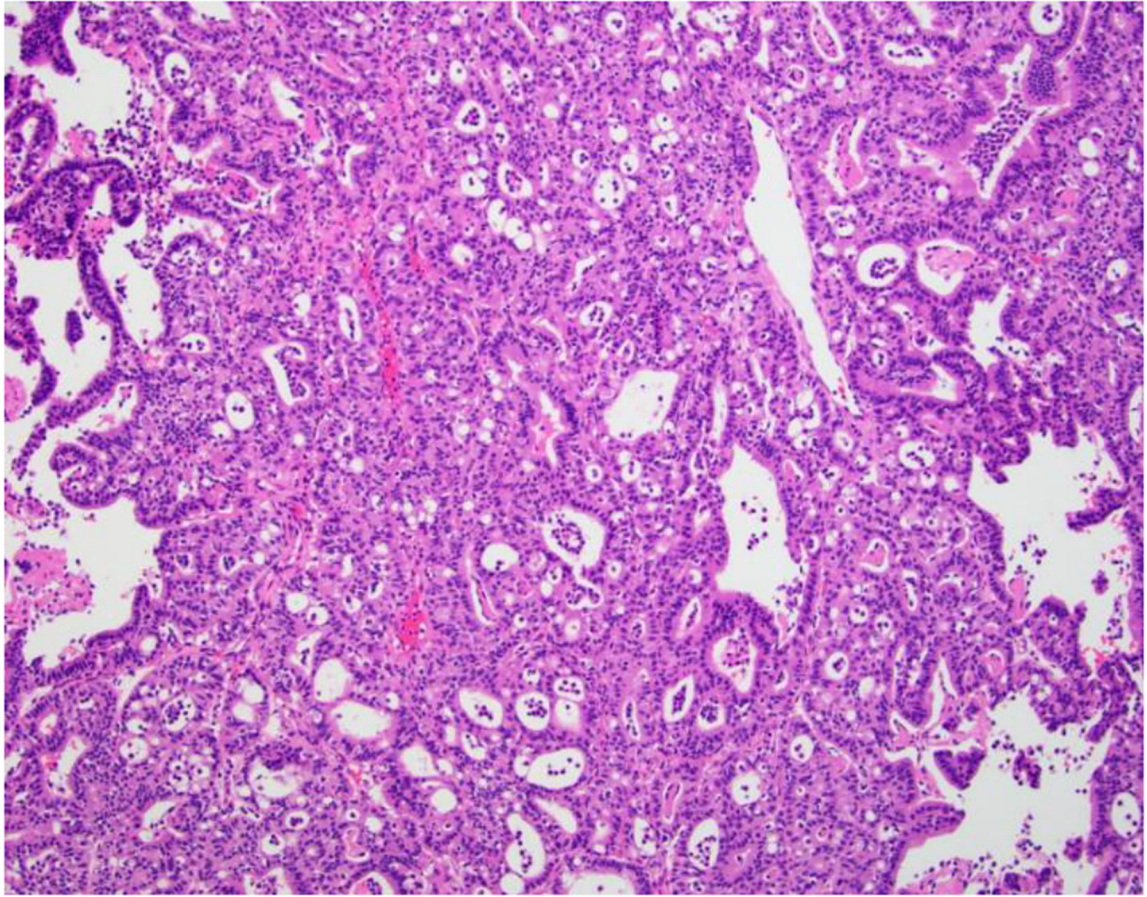


Figure 3:
Focal areas of the tumor showed loss of the papillary architecture with a cribriform pattern.

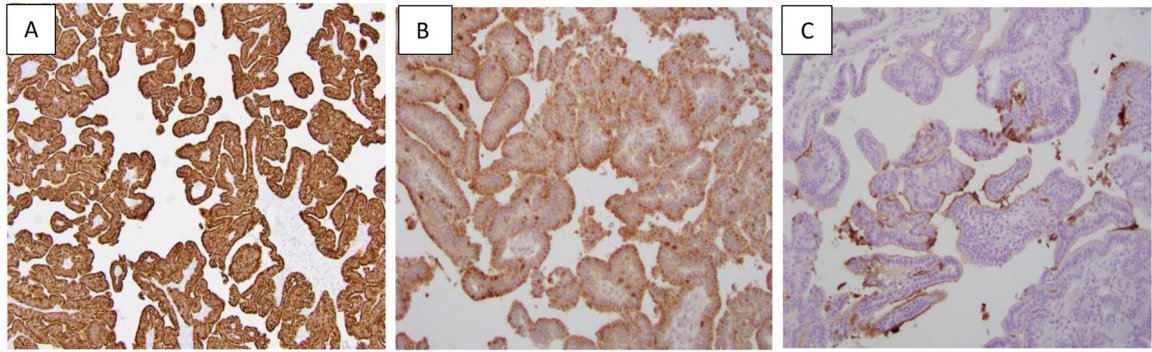


Figure 4:
The tumor was diffusely positive for A. cytokeratin and B. transthyretin and showed focal positivity for C. GFAP.

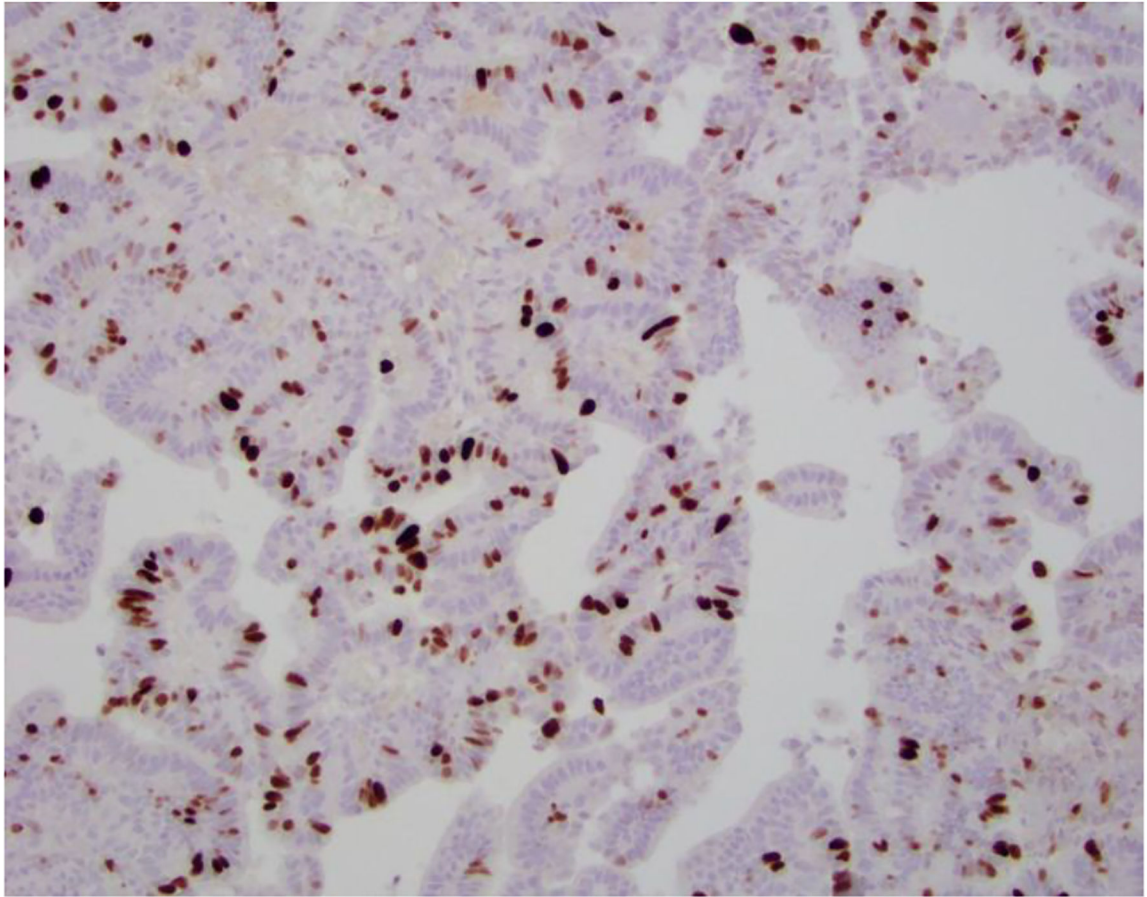


Figure 5:
Ki-67 immunostudy revealed an elevated proliferative index (10–15%).

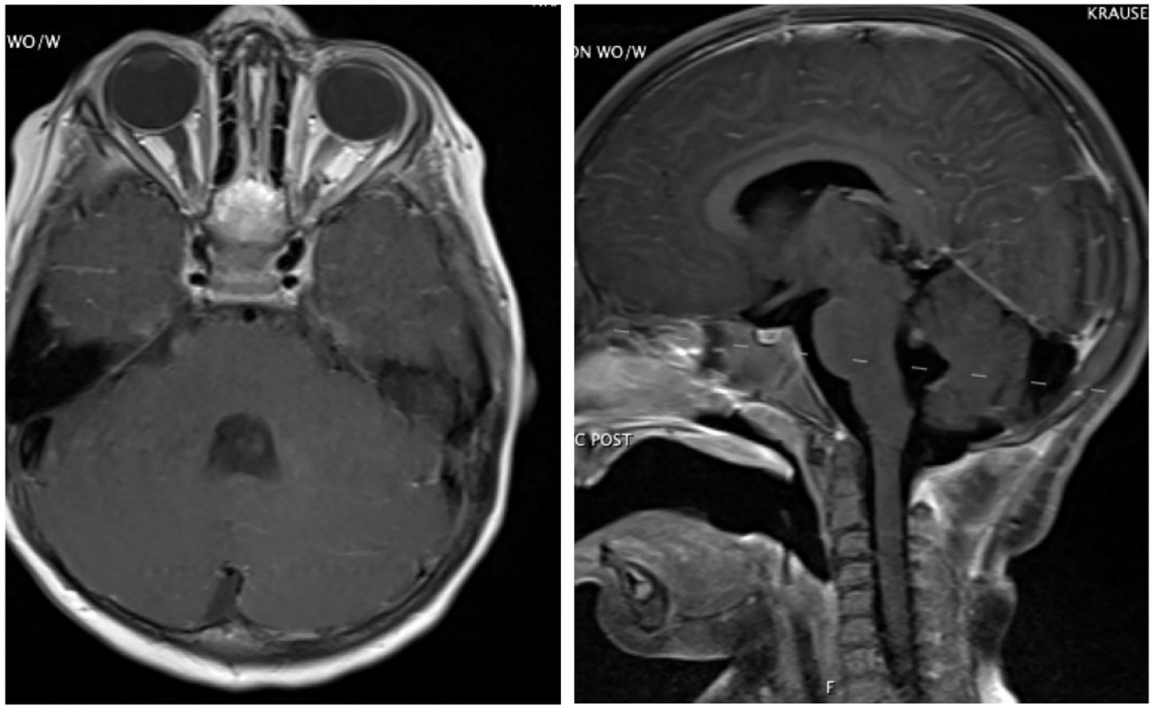


Figure 6:
Six-month follow up showing no tumors residual. A. Axial, B. Sagittal.

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