

Atypical Choroid Plexus Papilloma an Uncommon Entity: A Case Report

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Abstract Choroid plexus tumors (CPTs) account for 2-4% of all brain neoplasms in the pediatric age group, and 14% of brain tumors occurring in the 1st year of life. On the basis of their histological features, as per 2016 WHO classification of tumors of the central nervous system, these tumors are classified as choroid plexus papilloma (CPP; WHO Grade I), atypical CPP (ACPP; WHO Grade II), and choroid plexus carcinoma (CPC; WHO Grade III). Atypical CPP was first recognized as a distinct entity in the 2007 WHO classification of tumors of the central nervous system. They were characterized by increased mitotic activity and a higher probability of recurrence as compared with CPP. The prognostic features of and clinical outcome rates for ACPP are between those displayed by CPP and CPC. Choroid plexus tumors can metastasize as solid nodules or as sub-arachnoid seeding, especially to the spine cord in patients with posterior fossa tumors. Metastases from CPP are rare and few cases have been reported. Whereas ACPP and CPC metastasize with greater frequency. We herewith report one child with ACPP managed by surgical excision and is currently under follow up with oncology.

Keywords: atypical choroid plexus papilloma

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classified as grade II by the WHO in 2007.

1. Introduction

Choroid plexus tumors (CPT) are rare central nervous system neoplasms originating from the choroid plexus epithelium and therefore typically located in the ventricles. They account for 2–4% of all pediatric brain tumors and represent 14% of pediatric brain tumors that occur in the 1st year of life. [1] These tumors were traditionally classified by the World Health Organization (WHO) on the basis of histological criteria as either benign choroid plexus papilloma (CPP, WHO grade I) or malignant choroid plexus carcinoma (CPC, WHO grade III) with frank signs of malignancy including brisk mitotic activity, nuclear pleomorphism, high cellularity, blurring of the papillary growth pattern, necrosis, and often diffuse brain invasion. [1] Distinguishing between these two histologies has been difficult in some cases and prompted the coining of the term atypical choroid plexus papilloma to describe the tumors with an intermediate histology. In 2006 Jeibmann et al. reported that mitotic activity was the only atypical histological feature independently associated with recurrence based on a retrospective series of CPP patients. [2] Based on this data, atypical choroid plexus papilloma (APP) was defined in terms of mitotic activity and

2. Case Report

A 2-year-old child presented to the neurosurgery clinic with complaints of headache and vomiting. On examination the patient was noted to have papilledema. Brain imaging, including CT and MRI, revealed a large interventricular tumor with hydrocephalus on right side (Figure 1).

After explaining to the parents all possible risks and benefits of surgery, the patient underwent emergency insertion of a right ventriculoperitoneal shunt, and once stabilized, he underwent right parietal craniotomy and microsurgical excision of the interventricular lesion. The postoperative period was uneventful, and the patient was discharged home. Histopathological diagnosis of the tumor was Atypical choroid plexus papilloma, WHO grade II (Figure 2). Patient was referred to pediatric oncology and is currently under their follow up. Patient was later readmitted, and shunt was exteriorized and clamped, and once patient tolerated the clamping for few days, shunt was removed. Patient is also currently under follow up with neurosurgery OPD and the last post-operative MRI showed post-surgical changes and no residual lesion (Figure 3).

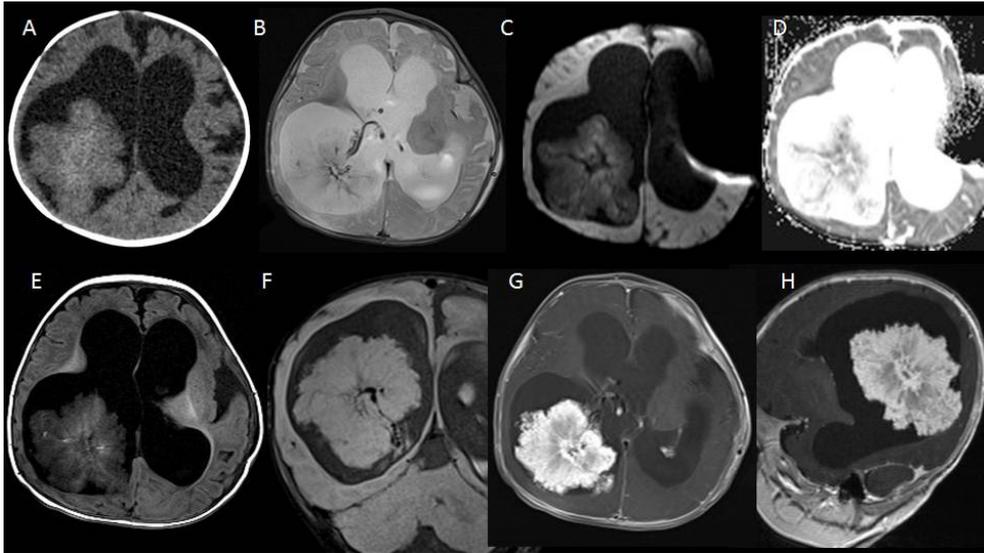


Figure 1. Axial Plain CT (A), MRI T2WI (B), DWI (C), ADC (D), T1WI (E), Coronal FLAIR (F), Axial T1WI+C (G) and Sag T1WI +C (H) shows a large lobulated avidly enhancing intra-ventricular lesion (70x5.7x7cm-TRxAPxCC) localized to the right posterior choroid plexus within trigone - posterior body region of right lateral ventricle. The lesion revealed signal characteristics hyperintense on T2WI/FLAIR and isointense on T1WI, without restricted diffusion. No other enhancing foci or deposits were seen in rest of the brain and on spine screening

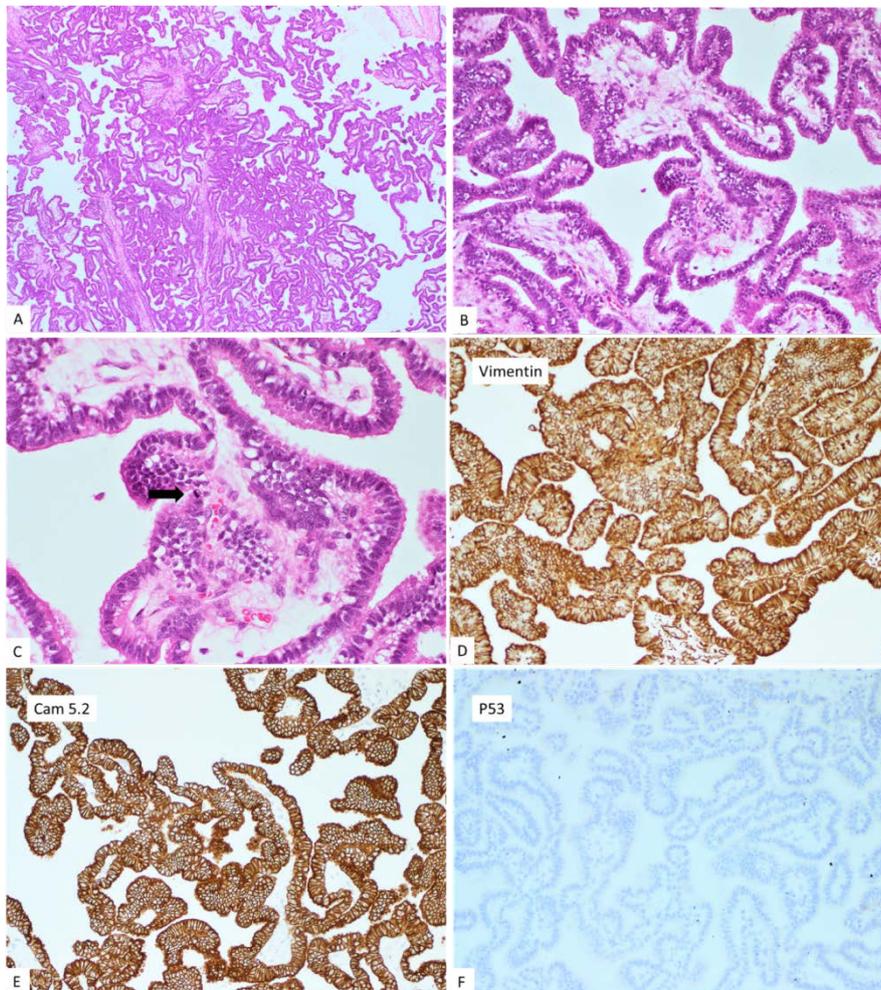


Figure 2. A, B, C. Atypical Choroid Plexus Papilloma (10x, 20x, 40x magnification). Papillary neoplasm composed of papillary fronds, lined by single to multiple layers of cuboidal epithelium, with fibrovascular cores. Neoplasm showed increased cellularity, mild nuclear pleomorphism and increased mitosis (11/10 HPFs) (C- black arrow-mitotic figure). No blurring of architecture, sheets of neoplastic cells or necrosis was seen. D. Vimentin, immunohistochemical stain, showed positive staining of the neoplastic cells (20x magnification). E. Cytokeratin Cam 5.2 immunohistochemical stain showed positive staining in the neoplastic cells (20x magnification). F. P53 immunohistochemical stain was negative in the neoplastic cells (20x magnification). Other positive immunohistochemical stains included: AE1/AE3, CK7, S100 and E-cadherin, and negative immunohistochemical stains included: GFAP, CEA and TTF-1. Ki-67 proliferation index was around 15-20%

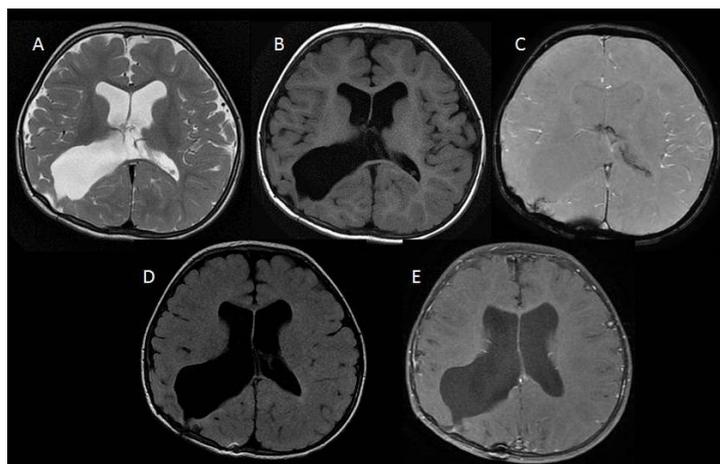


Figure 3. Follow up imaging at 4 months. Axial MRI T2WI (A), T1WI (B), SWAN (C), FLAIR (D), Axial T1WI+C (E) with post surgical changes and no residual lesion

3. Discussion

Atypical choroid plexus papilloma (ACPP) has an intermediate position between choroid plexus papilloma (CPP) and choroid plexus carcinoma (CPC) which is supported by clinical data. [3] Atypical choroid plexus papilloma have rarely shown spontaneous resolution of diffuse leptomeningeal contrast enhancement after primary tumor removal in pediatric cases. [4] ACPP term came into prominence in 2007, when it was first reported that a small percentage of choroid plexus papilloma showed recurrent tumor growth despite gross-total resection, but lacked malignant progression to choroid plexus carcinoma. [2] Recommended treatment of all choroid plexus tumors is to start with radical surgical resection, followed by adjuvant treatment in case of choroid plexus carcinoma, and a "wait and see" approach in case of choroid plexus papilloma. [5]

The prognosis for children with papilloma is excellent, and a 5-year survival rate for children with choroid plexus carcinoma is 50%, and all deaths usually occur within 7 months of operation. [6] The patients with choroid plexus carcinomas who underwent complete gross resection of the tumor had a better prognosis than did those with subtotal resection. [6]

Prognostically, high histological proliferation MIB-1 labelling indices by immunohistochemistry are associated with less a favorable clinical post-operative outcome in choroid plexus carcinomas, and in papillomas with atypical histology. However, most tumors with atypical histological features do not have distinctive MIB-1 labelling indices. [7] Thus analysis of growth fraction by MIB-1 immunohistochemistry may prove a useful ancillary method for assessing the malignant potential of choroid plexus neoplasms.

4. Conclusion

Atypical choroid plexus papilloma remains an uncommon entity. Papillomas and carcinomas of the choroid plexus are frequently seen, but atypical papillomas which have histological features in-between the two are seldom seen. Our case is one of these uncommonly seen lesions, which was managed by surgical excision and is under currently follow up in neurosurgery OPD and pediatric oncology with serial MRIs.

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