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Case Report

Choroid plexus papilloma originating from the cerebrum parenchyma

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Abstract

Background: Choroid plexus papilloma (CPP) can develop at a primary intraparenchymal location completely unrelated to the ventricular system. Here, we present a case of CPP that was difficult to diagnose preoperatively.

Case Description: Preoperative imaging and operative findings showed that the tumor originated entirely within the cerebrum parenchyma, with no connections between the tumor and the ventricular system. Histopathological examination of the tumor revealed a papillary structure with a single layer of well-differentiated columnar epithelium in the lesion. Furthermore, part of this lesion had infiltrated the cerebral parenchyma. Therefore, the tumor was diagnosed as CPP, and the diagnosis was confirmed by immunohistological examination.

Conclusions: CPP originating as intraparenchymal growths are extremely rare. Origins of extraventricular CCP are discussed in the context of the literature.

Key Words: Choroid plexus papilloma, intraparenchymal origin, magnetic resonance imaging

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INTRODUCTION

Choroid plexus papillomas (CPPs) are comparatively rare neuroectodermal tumors that develop from choroid plexus epithelial cells and account for 0.4–0.6% of all primary brain tumors.[13] The most common site of occurrence is the lateral ventricle in children and the fourth ventricle in adults. Occurrence in the third ventricle is rare.

CPP is usually restricted to within the trigone of the lateral ventricle and the fourth ventricle, shows a clear border, and ordinarily displays no intraparenchymal infiltration.[17] In adults, almost all CPPs develop in the fourth ventricle, but in rare cases they can occur at the cerebellopontine angle[9,10] or in the suprasellar region,[7,15] both of which are outside the ventricles. According to a previous report, extraventricular CPP has often been diagnosed preoperatively as meningioma of the cerebellopontine region, [9,10] pituitary adenoma or meningioma of the suprasellar region on the basis of imaging findings.[7,15] So, this rare tumor, particularly the one located in the extraventricular region, is difficult to diagnose based on preoperative imaging findings. In the present case, we treated a patient with CPP that had originated in and was limited to the cerebrum intraparenchymal region.

CASE REPORT

A 42-year-old man presented with a 13-year history of intractable tonic seizures that were indicative of right parietal lobe brain tumor. Plain computed tomography (CT) of the head revealed a tumor 40 mm in diameter

and consisting of calcification and cyst formation located in the right parietal region. Magnetic resonance imaging (MRI) revealed an area of marked signal hypointensity around the solid component on T_2 and T_2*-weighted imaging. These findings were thought to represent the calcification seen on CT, or perhaps hemosiderin deposition due to old hemorrhage. The tumor was located adjacent to the lateral ventricle, and the posterior part of the lateral ventricle was slightly enlarged toward the tumor. Normal choroid plexus of the right lateral ventricle was located in the normal position, and continuity of the normal choroid plexus to the tumor was not confirmed [Figure 1]. Cerebral angiography did not show any tumor staining or vascular abnormalities. The provisional diagnosis was cerebral cavernous angioma with hemorrhagic episode.

A right parietal craniotomy was performed using a navigation system and motor-evoked potentials. A yellowish, granulomatous, moderately hard, slightly lobulated avascular tumor was located in the right parietal lobe, with scant hemosiderin deposition identified within the lesion. The margin of the tumor was covered with predominant gliosis. At the deepest part of the tumor, the tumor was firmly adhered to the subependymal layer of the lateral ventricle. During dissection of the adhered area, the ventricular ependyma was penetrated and the body of the lateral ventricle was visualized through the cavity of the removed tumor. The normal-appearing choroid plexus was placed within the posterior part of the lateral ventricle and continuity of the normal choroid plexus and the tumor was not confirmed. The lateral wall on the lateral ventricle showed a normal appearance and continuous coverage with ependymal [Figure 2]. The tumor thus originated completely in the cerebrum parenchyma and was firmly adhered to the wall of the lateral ventricle.

Pathological examination of the tumor revealed a papillary structure with a single layer of well-differentiated columnar epithelium in the lesion. Part of this lesion growth had infiltrated the cerebral parenchyma. In addition, activated macrophages were prominent around the cerebral parenchyma and were considered to represent a reactive lesion related to an old hemorrhage. Immunohistochemical examination was accomplished with the antibodies detailed in Table 1. Vimentin was strongly immunoreactive. Neural Cell Adhesion Molecule (N-CAM, CD56), Epithelial Membrane Antigen (EM) and Cytokeratin 7 (CK7) exhibited focal immunoreactivity. Cytokeratin 20 (CK20) and prealbumin were not immunoreactive. Glial Fibrillary Acidic Protein (GFAP) was strongly immunoreactive. Podoplanin exhibited focal immunoreactivity in a few reactive cells [Figure 3]. Mindbomb Homolog 1 (MIB1) labeling index (MIB1-LI) was 0.4%. Based on these findings, histological diagnosis was CPP with hemorrhagic episode.

The patient showed no postoperative neurological

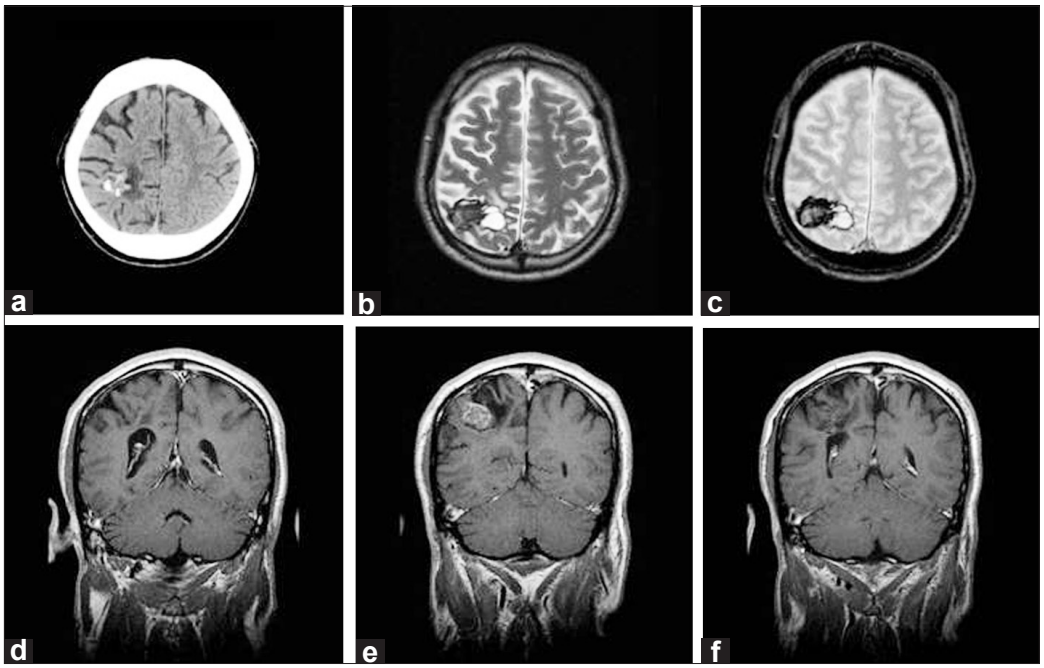

Figure 1: (a) Plain computed tomography (CT) of the head revealing a 40-mm-diameter tumor with calcification and cyst formation in the right parietal lobe. (b, c) Axial images are shown for magnetic resonance imaging (MRI). T2 and T2*-weighted imaging shows signal hyperintensity. T2 and T2*-weighted imaging reveals an area of marked hypointensity around the solid component, which may represent the calcification seen on CT or hemosiderin deposition due to an old hemorrhage. (d–f) Coronal images are shown for MRI. T1 gadolinium enhancement shows a heterogeneous contrast effect. Note: The normal-appearing choroid plexus was placed within the lateral ventricle and continuity of the normal choroid plexus and the lesion was not confirmed

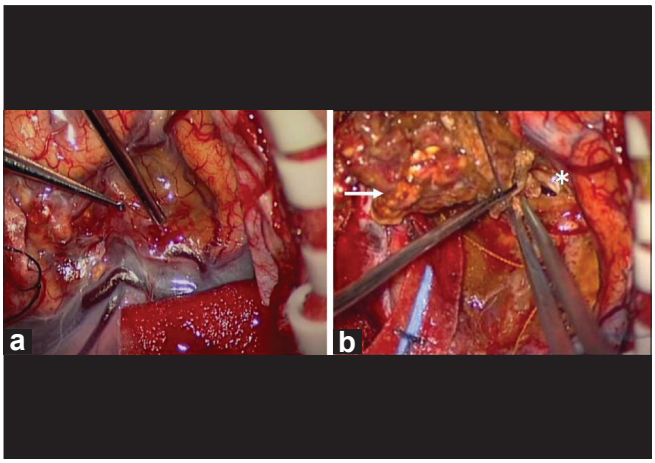

Figure 2: (a) Intraoperative microscopic view. The yellowish, granulomatous, slightly hard tumor was located in the right parietal lobe. (b) The tumor was firmly adhered to the cerebral ventricular wall, and the lateral ventricle was opened during resection of the deepest part of the tumor. Arrow (←) indicates the tumor margin. Asterisk (*) indicates the opening in the lateral ventricle

Table 1: The antibodies detailed of Immunohistochemical examination

Antibody	Company	Clone	Dilution magnification
CK 7	DAKO	monoclonal OV-TL 12/30	×25
EMA	DAKO	monoclonal E29	×100
GFAP	DAKO	monoclonal 6F-2	×50
Podoplanin	DAKO	monoclonal D2-40	×50
Prealbumin	DAKO	Rabbit polyclonal	×200
N-CAM(CD56)	Novocastra	monoclonal 1B6	×30
Vimentin	DAKO	monoclonal V9	×100

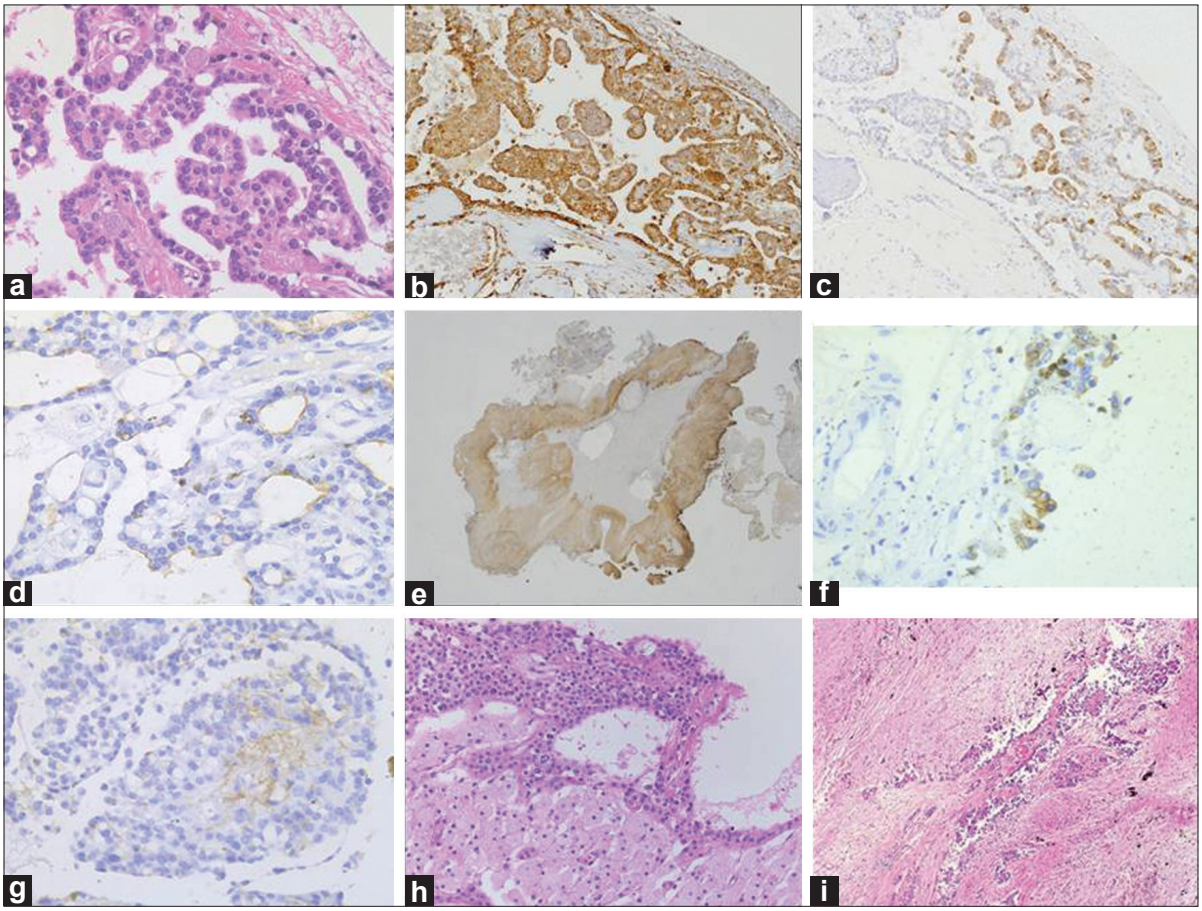

Figure 3: (a) The tumor tissue consists mainly of a fibrous component with crystal and hemosiderin deposition and macrophages. Densely proliferating cells with ovoid nuclei and resembling choroid plexus epithelium are observed in the peripheral area, showing a papillary architecture. (b) Vimentin was strongly immunoreactive. (c) N-CAM (CD56) exhibited focal immunoreactivity. (d) EMA exhibited focal immunoreactivity. (e) CK7 exhibited focal immunoreactivity. (f) GFAP was strongly immunoreactive. (g) Podoplanin exhibited focal immunoreactivity, contained a few reactive cells, if any. (h) In addition, activated macrophages were prominent around the cerebral parenchyma and were considered to represent a reactive lesion in relation to an old hemorrhage. (i) Papillary architecture was observed, extending further into the cerebral parenchyma (×100 for a–c, h, i; ×200 for d–g)

deficits, and cranial MRI confirmed complete removal of the tumor. Postoperatively, seizures were well controlled using antiepileptic drugs.

DISCUSSION

CPP is a comparatively rare neuroectodermal tumor that develops from choroid plexus epithelial cells, accounting for 0.4–0.6% of all primary brain tumors.[13] Based on imaging findings for the present case, primary brain tumor localized to the right parietal lobe was suspected. Preoperative diagnosis had been cavernous angioma with intratumoral bleeding. Operative findings revealed no findings indicative of CPP, such as hypervascular structure (e.g., reddish color) or a cauliflower-shaped appearance. The tumor was located entirely within the cerebrum parenchyma, adhered to the ventricle wall, but was entirely separate from the ventricular system. At the conclusion of the surgery, no definitive evidence of CPP was observed prior to histopathological diagnosis.

Several reports have described CPP located outside the ventricular system. Cases involving the cerebellopontine angle or foramen magnum reportedly originated from the choroid plexus through the cerebello-medullary fissure. Kimura *et al*.,[7] Ma *et al*.,[8] and Samejima *et al*.[15] have reported CPP in the suprasellar region, completely unrelated to the third ventricle. In those case reports, CPP was located within the subarachnoid space. To the best of our knowledge, intraparenchymal CPP is extremely rare. In 2005, Dwarakanath *et al*. reported CPP with cyst in the cervico-thoracic intramedullary region,[3] while Greene in 1951[4] and Robinson in 1955[14] independently reported cases of cerebellar CPP, and Pillai *et al*. reported a case of intrinsic brainstem CPP in 2004.[11] In 1981, Azzam and Timperley described a cystic tumor in the right frontotemporal cortex.[2] Azzam and Timperley suggested two hypotheses for the origins of that lesion: first, that a primitive ectopic choroid plexus had already been present in the cerebral parenchyma and second, that migration of epithelial tissue had occurred in the parenchyma during brain development. Accordingly, the present case might have originated due to ectopic development or metaplasia of an ependymal cyst that became established and then differentiated into CPP in the cerebral parenchyma.

Histological findings show that CPP has a structure generally characterized by a layer of cast epithelial cells, an inner stroma comprising connective tissue rich in blood vessels, and papillary growth. These findings revealed clear papillary architecture in part of the CPP in the present patient. Vimentin, CK7, GFAP, EMA and podoplanin are expressed by virtually all CPPs.[5] Approximately 70% of CPPs are positive for prealbumin,[6] but results were negative in this case. However, in some studies, a significant proportion of tumors failed to stain with antibodies to prealbumin, and, more importantly, metastatic lesions such as ovarian teratoma may be positive when stained with this marker.[1] These findings limit the utility of prealbumin as a single marker in the differential diagnosis of CPP. These histological findings supported the diagnosis of CPP in the present case.

As differential diagnoses, histological examination focused on papillary ependymoma and metastatic brain tumor. However, papillary ependymoma exhibits growths with fingerlike projections lined by a single layer of cuboidal tumor cells with smooth contiguous surfaces and with GFAP-positive tumor cell processes; therefore, this diagnosis was ruled out in the present case. In contrast, CPP and metastatic carcinomas form bumpy, hobnail cellular surfaces that do not feature extensive GFAP-positivity. In addition, primary malignant tumor was excluded in the present case.

Another characteristic finding in this patient was the infiltration of the cerebral parenchyma by tumor growth. We observed a clear papillary architecture in part of the CPP in the present patient. Also, focal immunoreactivity of podoplanin might indicate a characteristic of ectopic CPP. CPP usually presents with well-defined, smooth or lobulated margins, related to the choroid plexus in neuroradiological examination. Our imaging studies have shown that a tumor located within the cerebrum parenchyma was unrelated to the choroid plexus, with no uniform enhancement and with unclear margins from the surrounding structures. Dense calcification and cyst were noted on CT. These findings are infrequent in CPP, with patchy calcification in 24% and cyst in 20% of CPPs.[16] Our histological findings showed calcification and multiple cystic cavities around the tumor, and these were attributed to chronic post-hemorrhagic changes.[12] Arriving at a clear assessment to diagnose CPP was difficult based on preoperative imaging findings, given the tumor location and the possibility of reactive lesions forming in relation to an old hemorrhage. Finally, we reached the diagnosis as CPP of the intraparenchymal region with previous history of hemorrhage around the tumor.

CONCLUSIONS

One factor explaining why CPP was located in the extraventricular system is very difficult to distinguish on the basis of diagnostic imaging findings alone. This is the second case report in the literature of CPP originating in the cerebrum parenchyma.

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