Intraparenchymal Choroid Plexus Papilloma: A Case Report and Literature Review

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Abstract
Choroid plexus papilloma (CPP) is a rare tumor of the central nervous system, which is usually located in the ventricular system and can metastasize to cisterns along the cerebrospinal fluid circulation. CPP is only rarely located in the intraparenchymal. Here, we describe an unusual case of such a tumor in a 25-year-old man. He initially experienced a sudden, generalized tonic-clonic seizure; further investigation showed no electrolyte imbalance and ruled out the possibility that the seizure was induced by medication. A sharply circumscribed regular mass lesion was identified on a magnetic resonance image in the medial aspect of the left temporal lobe together with invasion of the cortex. The patient underwent stereotaxic biopsy for a pathological diagnosis, followed by craniotomy with total resection of the mass 1 week later. The histopathology showed that the lesion was a CPP. The pathophysiology, radiology, classification, and treatment strategy of this rare tumor are discussed in the report.

INTRODUCTION

The choroid plexus, which produces cerebrospinal fluid (CSF), is a fibro vascular tissue composed of neuroepithelial-lined papillae. Choroid plexus papillomas (CPPs) are rare and histologically benign intracranial neoplasms of the choroid plexus and are normally located in the ventricular system. Nevertheless, extra ventricular sites have been described in a few reports. It can disseminate throughout the neuroaxis, including the suprasellar region, cerebellopontine angle, pineal region, posterior fossa and spinal canal. To the best of our knowledge, only two cases of ectopically intraparenchymal CPP have been reported. Here, we describe a rare case of intraparenchymal CPP, which presented as two general seizure attacks within five hours of each other, and we review the relevant literature.

CASE PRESENTATION

This 25-year-old man with no significant medical history firstly presented with a generalized tonic-clonic seizure attack that lasted 2 minutes. This occurred in the afternoon, whilst he was in his office. During the episode, it was observed that his neck turned to the right, and that he experienced lockjaw, loss of consciousness and generalized convulsions. He recovered consciousness 5 minutes later and he was immediately transferred to our emergency department. The anti-epileptic drug, Valproate acid, was initially administered as an 800mg intravenous drip. Five hours later, whilst still in the emergency department and having just finished Valproate acid infusion, he suffered a second epileptic episode. This lasted for 3 minutes and lorazepam 2mg infusion was administered for sedation. After 1 hour, the patient recovered consciousness, and subsequently had no further seizures.

Physical and neurological examination, including all 12 cranial nerves, yielded no unusual findings, and laboratory blood tests results, including electrolyte and hormone levels, were all within the normal range. A computed tomography (CT) scan of the brain without contrast demonstrated a focal hypodense lesion in the medial aspect of the left temporal lobe, with extensive invasion of the cortex (Figure 1A). This was a sharply circumscribed regular mass, initially suggestive of tumor growth. Blood tumor markers (AFP, CEA, CA125, and CA19-9) were all within the normal range. Magnetic resonance imaging (MRI) of the brain (Figure 1B, 1D) revealed an ill-defined, expansible lesion with partial cystic change in the medial temporal lobe and extensive involvement of the cortex without perifocal edema. The hypointense part of the mass on the T1-weighted image was slightly enhanced by contrast (Gd-DTPA) (Figure 1C), and contrast images provide no evidence that the tumor was attached to the normal choroid plexus. Therefore, the differential diagnosis included dysembryoplastic neuroepithelial tumor (DNET), low-grade glioma, and ganglioglioma.

Based on these imaging findings and the repeated seizures suffered by the patient, we initially performed a stereotaxic biopsy using Navigator (Brain Lab) instead of total resection, because the lesion was close to the optic tract and a number of large

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- Intraparenchymal
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blood vessels. If the pathologic findings show that a lesion can be treated using an alternative, non-invasive method (e.g. Gamma-knife, radiotherapy, or chemotherapy), this would be the first choice of treatment. Finally, the pathological analysis of the biopsy sample revealed a papillary neoplasm characterized by marked nuclear atypia, stratification with glandular complexity, and branching papillary fronds. Histologically, it was necessary to distinguish between CPP, metastatic adenocarcinoma from the lung, or thyroid, and papillary glioneuronal tumors. Immunohistochemical staining of the biopsy sample was negative for CK7, CK20, CK, TTF-1, and GFAP, but strongly positive for synaptophysin, suggesting that the tumor was a CPP.

Based on previously reported cases, en-bloc resection was the optimal treatment. The patient underwent a craniotomy, which was performed using Navigator, in order to totally remove this lesion because CPP has a poor response to chemotherapy. After the operation, the patient’s condition was good and no obvious neurologic deficit was apparent over the following two months. We also follow brain MRI and it revealed a small, poor enhancing residual cystic lesion in the left medial temporal lobe (Figure 4).

**DISCUSSION**

CPPs are rare neoplasms and represent only 0.3 -0.6% of all intracranial tumors. Their location is strongly associated with age; over 80% of supratentorial CPPs are found in patients younger than 20 years [1]. The clinical features and signs at diagnosis include hydrocephalus, papilledema, gait impairment, seizure and psychomotor retardation.
There are 3 grades of choroid plexus tumor, CPP (WHO grade I), atypical CPP (WHO grade II) and choroid plexus carcinoma (WHO grade III). CPPs exhibit a well-formed and continuous basement membrane and very low mitotic activity. Macroscopically, they are pedunculated, pink-grey, and cauliflower-like masses and are usually well circumscribed from normal brain.

In radiological imaging, most CPPs are well vascularized and can be enhanced with contrast on brain CT. On MRI, papilloma is typically iso- or hypointense on T1- and T2-weighted imaging. It may also demonstrate a heterogeneous hyperintensity on T2-weighted imaging and be enhanced after contrast injection unless the tumor is highly calcified.

Although over 75% of choroid plexus neoplasms are benign, this is not reflected by their histological appearance. A number of literature reviews have stated that CPPs can occur ectopically (as: metastasis or dissemination) along the neuraxis from the supratentorial compartment to the sacrum [2,3-6]. In some reports, patients with a CPP who exhibited a loss of the normal architecture or evidence of parenchymal invasion had a good outcome after surgical resection [1,2].

McCall et al. reviewed 14 cases of disseminated CPP and only one of these involved intraparenchymal CPP of the temporal lobe with hydrocephalus [3,7]. Another intraparenchymal case with a right parietal mass resulting in an intractable seizure was reported by Masaaki et al. in 2011 [2]. To the best of our knowledge, the case we report here is the first CPP to be located in the medial temporal lobe, leading to repeated seizures. The initial brain CT and MRI with contrast showed no obvious calcification in this case, and only slight enhancement at the CPP lesion, which made diagnosis more difficult. This intraparenchymal CPP was not connected to the normal-appearing choroid plexus and was present instead within the ventricles, and may have undergone calcification.

**CONCLUSION**

The best treatment strategy for CPPs is maximum safe surgical resection [1]. McCall et al. reported that chemotherapy with temozolomide combined with radiotherapy could be used to successfully treat two metastatic CPPs, but more data is needed to allow a definitive recommendation [3].

**REFERENCES**