Choroid Plexus Papilloma of Foramen of Luschka

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ABSTRACT

We reported a rare case of choroid plexus papilloma arising at the foramen of Luschka of the fourth ventricle. The features of the tumor, as observed by computed tomography, magnetic resonance imaging and digital subtraction angiography are described, along with a review of the literature.

CASE REPORT

A 24-year-old male patient presented with intermittent headache and dizziness, which had persisted for approximately 1 month following a traffic accident. He also had dysfunction of taste, such as intolerance of salty food and increasing tolerance of spicy food compared to his usual taste preference; an intolerance of strong light while driving was also found. Physical and laboratory examinations disclosed no abnormal findings except double visions.

Computed tomography (CT) showed a slightly hyperdense solid mass lesion sized approximately 1.0 × 1.0 cm in diameter at the cerebellomedullary cistern of the right hemisphere. Magnetic resonance imaging (MRI) showed isointense on T1-weighted image (T1WI), slightly hyperintense on T2-weighted image (T2WI) and moderate, slightly heterogeneous enhancement on an enhanced T1WI with gadolinium (Fig. 1). Cerebral digital subtraction angiography (DSA) showed no evidence of vascular abnormalities such as tumor vessels, early arteriovenous shunting, stain or mass effect.

Considering a differential diagnosis of ependymoma, lower cranial neuroma, lymphoma, metastasis and CPP, we performed a craniotomy via the right suboccipital approach followed by partial resection of C1 and subtotal excision of the mass. The surgical findings suggested that the tumor had originated in the choroid plexus located at the foramen Luschka and extended into the cerebellomedullary cistern. Gray-white soft tissue fragments, measuring up to 0.9 × 0.8 × 0.4 cm in size were harvested, and histological examination showed that the tumor was a CPP (Fig. 2). The postoperative course was uneventful.

DISCUSSION

CPPs account for approximately 3-5% of all intracranial neoplasms in children and 0.5% in adults [1]. Essentially, these are tumors of childhood and most commonly affect...
Figure 1. A mass lesion of 1 cm diameter located within the cerebellomedullary cistern on the right. The lesion was seen as a slightly hyperdense on plain CT image a, isointense on plain T1WI b, slightly hyperintense on T2WI c, and moderate, somewhat heterogeneous enhancement on axial contrast-enhanced T1WI d.
Choroid plexus papilloma of Foramen of luschka

Figure 2

Figure 2. Photomicrograph of the tumor specimen revealed a papillary growth consisting of a single layer of columnar epithelium, indicating a choroid plexus papilloma (H&E, x400)

children under the age of 5 years. Most of CPPs are found in the atrium of the lateral ventricle. Fourth ventricular CPPs are more prevalent in adults [2]. CPPs located primarily in the extraventricular region are uncommon; however the most frequent primary extraventricular location is the cerebellopontine angle. Rovit et al. reported that the commonest sites of CPPs (in order of frequency) were the lateral ventricle (43%), fourth ventricle (39%), third ventricle (10%), and cerebellopontine angle (CPA) (9%) across all groups [1]. The cerebellomedullary cistern is a rare site for CPPs [3-6]. A few cases of primary intraparenchymal CPP and a rare case of primary CPP in the suprasellar region have also been reported [3]. We speculate that extraventricular CPPs originate either from direct extension of a fourth ventricular papilloma, from distant cerebrospinal fluid (CSF) seeding, or from a primary CPA lesion, possibly arising de novo from embryonic choroidal remnants [2, 3, 7, 8].

CPPs of the CPA are slow-growing tumors and usually develop between the second and fifth decades of life [8, 9]. Clinically, patients with CPP of the CPA typically present with trigeminal, facial or acoustic nerve palsies or ataxia. On CT, CPPs are usually homogeneous in children but inhomogeneous in adults, often related to cystic degeneration [10]. Tumoral calcification has been reported in 4.1% to 20% of intraventricular papillomas, but is more frequent in the rarely reported CPA papillomas [1, 7]. An MRI of CPPs usually discloses homogeneously hypointense or isointense signal on T1WI, heterogeneously hyperintense signal on T2WI, and a characteristic intense, heterogeneous enhancement with Gd-DTPA [7, 9, 11, 12].

Hydrocephalus is often encountered in CPPs but is comparatively less common in patients with CPA-CPPs. Internal hydrocephalus develops due to excessive formation of CSF by the papilloma. Another contributing factor is that the tumors are apt to spontaneous bleeding. This may result in a low-grade arachnoiditis with consequent fibrosis leading to disturbance in CSF return [1]. Talacchi et al. hypothesized that this difference could be attributed to the inability of the extraventricular CPPs to over produce CSF because of having less blood supply than the intraventricular CPPs [13].

A CPP situated in the fourth ventricle may extend into the CPA or into the cerebellomedullary cistern. Nomura et al. reported a CPP located in the cerebellomedullary cistern and attached to the choroid plexus of the foramen of Magendie [3]. Morello et al. described a CPP that developed from the normal choroid tuft outside the foramen of Luschka and extended into the cerebellomedullary cistern [8]. In the presented case, a primary CPP of the foramen of Luschka CPP which arose from the choroid tuft remnants in the foramen and extended into the cerebellomedullary cistern. The mass was 1 cm in diameter and located in the right cerebellomedullary cistern below the CPA, the tumor not being large enough to extend into the CPA.

The images of the mass were slightly hyperdense on plain CT, isointense on plain T1WI, slightly hyperintense on T2WI, and showed moderate and slightly heterogeneous enhancement on enhanced T1WI (Fig. 1). These image manifestations were consistent with those of previously reported cases. Cerebral DSA showed no evidence of
abnormal tumor vessels or stain. The differential diagnosis for this lesion on MRI includes meningioma, neurinoma, hemangioblastoma, ependymoma, choroid plexus papilloma and metastatic tumor [11-14]. The average ages for the peak incidence of CPP, ependymoma, and medulloblastoma are 6 months, 2.5 years, and 6 years, respectively. An ependymoma usually arises from the lower cerebellar vermis and extends through the outlets of the fourth ventricle. It is an intra-axial tumor and usually has more irregular margins and surrounding edema [11, 13]. A meningioma must have an attachment to the dura or tentorium at the posterior fossa, and may show a persistent stain on DSA. A meningioma may show isointense on T1WI and T2WI as well as strong homogeneous enhancement; it may also present glutamine and glutamate levels of around 2.1 to 2.5 ppm, as well as a constantly observed tarurine level of 3.3 ppm in medulloblastomas in magnetic resonance spectroscopy studies. Hemangioblastomas most often occur infratentorially as intra-axial masses and may give multiple variable patterns, such as solid or cystic and solitary or multiple mass (es). Hemangioblastomas are slightly hyperdense with good enhancement of the soft tissue part on CT and hypointense with very good enhancement on T1WI, and hyperintense with flow-void features observed as engorged vessels on T2WI on MRI. DSA is the most suitable method for hemangioblastomas, as it reveals a highly vascular tumor nodule in the mid-arterial phase and dense, prolonged tumor blush with prominent arteriovenous shunting in the capillary phase [15]. Most metastases to the CPA are associated with additional brain metastases and meningeal involvement [12]. Diffusion-weighted imaging and apparent diffusion coefficient maps can enable a differential diagnosis between an arachnoid cyst and an epidermoid tumor.

CONCLUSION

The case we have presented is rare in terms of its location and lack of typical image features of CPPs. No obvious intraventricular extensions, lack of hydrocephalus, and homogeneous enhancement on MRI made the diagnosis more difficult. However, it is necessary to consider that CPP is a potential diagnosis of cerebellomedullary cistern tumor and that its image presentation may be different from that of intraventricular CPPs.

REFERENCES