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Choroid Plexus Papilloma in an Adult

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Choroid plexus papillomas are rare intraventricular tumors and present infrequently in adults. Clinical presentation is typically associated with symptoms and signs of increased intracranial pressure secondary to hydrocephalus. Total tumor resection is reported to be curative for choroid plexus papillomas. Herein, we report a 39-year-old woman who complained of intermittent dizziness, nausea, and unsteadiness for nearly one month before admission. Magnetic resonance images revealed a mass about 3 cm in maximal diameter located in the left exit of the foramina of Luschka at the cerebellomedullary angle, with compression of the lower pons and medullary regions and obstructive hydrocephalus. The patient underwent a suboccipital craniotomy with total removal of the tumor with the assistance of a surgical navigator system. Pathology confirmed a diagnosis of choroid plexus papilloma. A ventriculo—peritoneal shunt was performed seven days later because of hydrocephalus. The patient was discharged on day 18 of hospitalization and no obvious neurological deficits were noted.

Key words: choroid plexus, brain neoplasm, papilloma, hydrocephalus

INTRODUCTION

Choroid plexus papillomas (CPPs) are uncommon benign neuroectodermal tumors that account for 0.4 to 1% of all intracranial neoplasms[1]. Most CPPs arise within the ventricles and affects children less than two years of age[2]. However, CPPs presenting in adults are rarely reported in the literature and remains a challenge for clinical physicians. Here we report a case of a CPP in an adult and review the relevant literature.

CASE REPORT

A previously healthy 39-year-old woman complained of a progressive spinning sensation with nausea and vomiting, headache, tinnitus, numbness in the left facial region, and unsteadiness for nearly one month before admission. She did not experience blurred vision or hearing impairment concurrently. She also complained of disorientation in time and place and impaired short-term memory. Neurological examinations revealed left facial palsy and paresthesia over the left V_2 dermatome. Nystagmus during left-sided gazing and left dysmetric finger-to-nose movements were also noted. Otherwise, the patient showed no gait ataxia, no other abnormal cerebellar signs, no papilloedema in the fundus, no decrease in muscle power, no limited eyeball movement, and absence of Babinski's signs or Hoffmann's signs. Cognitive function tests revealed fluctuating impairments in short-term memory and disorientation in time and place. Magnetic resonance (MR) images revealed a mass about 3 cm in maximal diameter located in the left exit of the foramina of Luschka at the cerebellomedullary angle, with compression of the lower pons and medullary regions and obstructive hydrocephalus. (Fig. 1A-C) The tumor displayed homogenous enhancement after injection of gadolinium. As there was no dural tail sign for the tumor on MR, a vestibular schwannoma was the initial clinical diagnosis. The patient underwent a suboccipital craniotomy with total removal of the tumor using the assistance of a surgical navigator system. At surgery, the tumor was yellowish to grey in color and easily detachable. Microscopically, it appeared with delicate papillary stalks composed of fibrovascular fronds, with a lining of columnar epithelium (Fig. 2) Pathology confirmed the diagnosis of CPP. A ventriculo — peritoneal shunt was inserted seven days later because of persistent hydrocephalus. The patient was discharged without obvious neurological deficits on Day 18 of hospitalization.

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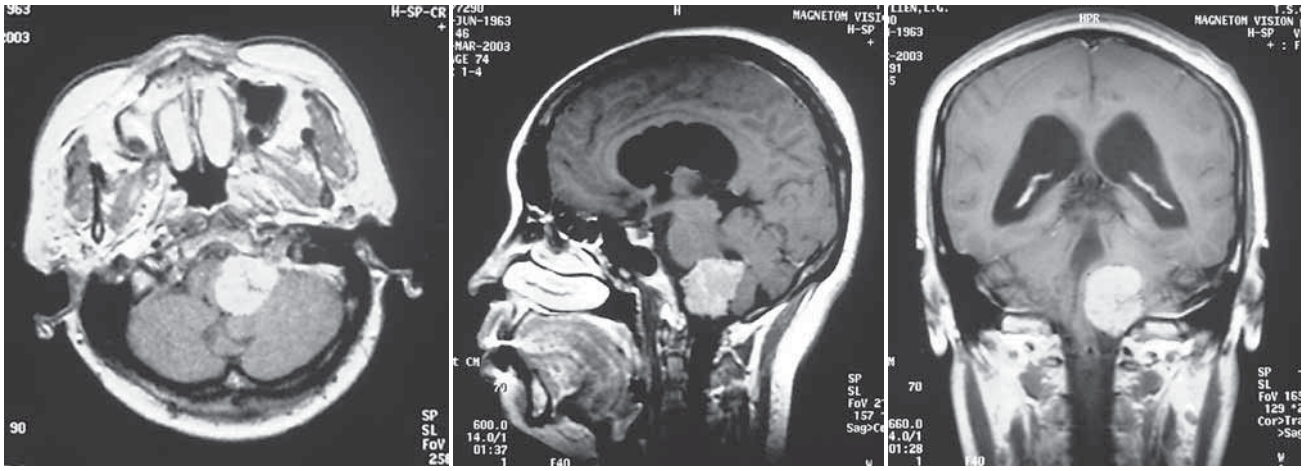

Fig. 1 T1-weighted magnetic resonance images of brain after injection of gadolinium. (A) Axial image revealed an enhanced mass over left cerebellopontine angle, compressing the medulla. (B) Sagittal image revealed an enhanced mass over the foramen of Luschka with extraventricular extension. (C) Coronal images revealed an enhanced mass over left cerebellopontine angle, with obstructive hydrocephalus.

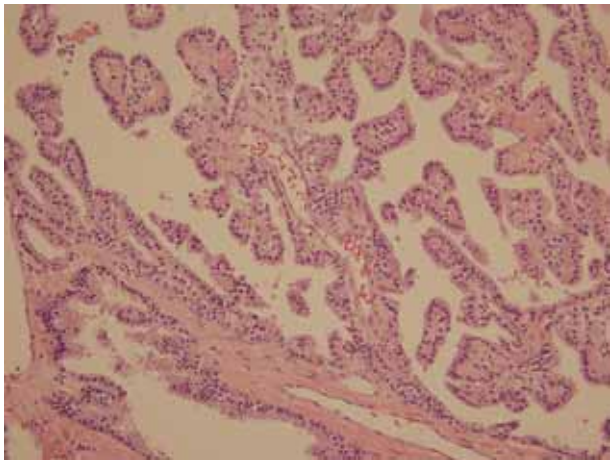

Fig. 2 Microscopically, the tumor showed delicate papillary stalks composed of fibrovascular fronds, with the lining of columnar epithelium. (HE×400)

Postoperatively, no cerebrospinal fluid (CSF) leakage or other progressive symptoms were noted. During the regular follow-up with neurological examinations and MR images of the brain, no recurrence was noted. Otherwise, the patient's previous symptoms such as horizontal nystagmus and trunk ataxia disappeared; only dizziness persisted.

DISCUSSION

CPPs are rare intraventricular tumors of neuroectodermal origin and represent less than 1% of all brain tumors in adults[3]. Lateral ventricles, followed by the fourth and third ventricles in order of decreasing frequency are the most common locations in all populations[4]. However, in adults the most common location is the fourth ventricle, whereas in children the trigone of the lateral ventricle with dominance for the left side is most frequent[1,5]. CPPs in the posterior cranial fossa also tend to arise at the foramen of Luschka with extraventricular extension[4]. The etiologies of CPPs remain obscure and associations with Li — Fraumeni cancer syndrome, Aicardi syndrome, and Von Hippel — Lindau syndrome have been proposed[6].

Most patients who have CPP present with symptoms and signs of increased intracranial pressure secondary to hydrocephalus[3,6]. In a surgical series of 33 cases, Tacconi et al. found headaches to be the most common symptoms; with papilloedema being the main clinical sign in adults (63%) and unsteady gait in children (71%)[5]. The time-course of CPPs of the lateral ventricle in children is often short and presents with progressive hydrocephalus. On the contrary, in adults with CPPs located in the lateral ventricle, a long history of headache, paresis, cranial nerve palsies, and progressive visual loss is typical. Otherwise, with CPPs in the fourth ventricle or cerebellopontine angle, signs and symptoms of hydrocephalus accompanied by ataxia, lower cranial nerve palsies, and visual disturbances are the most common presentations in all age groups[7]. The pathophysiology may be related to an obstruction to CSF flow by the tumor or the higher production (four to five times) of CSF by the stimulated tumor cells[2,4].

The diagnosis of CPP is based on histopathology. These tumors consist of a single layer of regular columnar epithelium resting on a delicate papillary stroma composed of fibrovascular fronds[3]. In neuroimaging studies, a CPP usually presents as a well-margined, smooth or lobulated,

calcified, isodense or hyperdense mass on precontrast computed tomography (CT) and shows dense and usually homogeneous enhancement on postcontrast CT[4,8]. On MR imaging, these tumors show homogenous hypointense or isointense signals on T1-weighted images, heterogeneous hyperintense signals on T2-weighted images, and characteristic intense, heterogeneous enhancement[4]. Proton MR spectroscopy of CPPs typically reveals high levels of choline-containing compounds and complete absence of the creatine and neuronal/axonal marker, N-acetyl aspartate[9]. On angiography, CPPs in the posterior cranial fossa are supplied mostly by the anterior or posterior inferior cerebellar arteries[8]. The differential diagnosis for similarly located lesions includes a meningioma, neurinoma, hemangioblastoma, ependymoma, or metastatic tumor[4].

Total tumor resection of CPPs is generally accepted as the optimal management and adjuvant therapy including radiotherapy or chemotherapy is not recommended for these cases[1,2]. Up to 100% 5-year survival has been reported after a complete surgical resection[4]. In a meta-analysis of surgery for these tumors, the 10-year survival rate for patients with complete resection was estimated as 85%[10]. In a study of 25 patients with CPP, reported by McEvoy et al., 39% suffered from developmental delay, 17% from severe behavioral problems, and 48% from epilepsy[2]. However, a longer prognosis depended partly on patient age and was only indirectly linked with the tumor site. The overall prognosis was better for adults with CPPs in the fourth ventricle than among children, with a survival time of 9-13 years. Severe hydrocephalus, mental handicap, or retardation are the most severe forms of morbidity for children with CPPs treated with surgery[7]. With the advance of microsurgical techniques such as neuroendoscopy, both morbidity and mortality have decreased dramatically[5,11].

In this case, the patient complained of a progressive spinning sensation with nausea and vomiting, headache, tinnitus, numbness in the left facial region and unsteadiness. These symptoms were related to the hydrocephalus and to the compression of cranial nerves by the tumor. Neuroimaging showed a solitary, huge (approximately 32 mm long), homogenously enhanced mass in the left exit of the foramina of Luschka at the cerebellomedullary angle, with compression of the lower pons and medullary regions and obstructive hydrocephalus. However, CCPs present only rarely in the cerebellomedullary angle in adults and it is hard to predict the diagnosis of CPP from a preoperative assessment, so this case contributed to our clinical experience.

Thus, when an adult presents with posterior fossa tumors, even in the ventricle, an extraventricular extension of a CPP should be considered.

In conclusion, a CPP in the adult is rare and presents diagnostic and therapeutic challenges for clinical physicians because of its deep intraventricular location and marked vascularity. MR imaging, angiography, and CT are helpful for the differential diagnosis.

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