

Case Report

Choroid Plexus Papilloma of the Cerebellopontine Angle

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Abstract

The cerebellopontine angle is an extremely rare site for the growth of a choroid plexus papilloma. The clinicoradiological diagnosis of this tumor in the cerebellopontine angle is difficult because of its rarity in addition to a nonspecific clinical presentation and radiological features. Herein, we report the case of a 49-year-old woman with complaints of headache and features of raised intracranial pressure, whose computerized tomography (CT) and magnetic resonance imaging (MRI) were suggestive of acoustic neuroma or meningioma with hydrocephalus. Histology revealed multiple arborizing papillae with a central fibrovascular core, lined by cubo-columnar cells. The cells showed diffuse immunoreactivity for pancytokeratin, S100 protein, synaptophysin, and vimentin, as well as focal expression of glial fibrillary acidic protein, epithelial membrane antigen, and cytokeratin 7. English medical literature is also reviewed.

Keywords: central nervous system, cerebellopontine angle, choroid plexus papilloma

Introduction

Choroid plexus papillomas (CPPs) are benign and slow growing tumors (grade 1 according to the WHO classification).¹ Both sexes and different age groups can be affected, but they are more frequently seen during childhood.² Among all intracranial neoplasms of all age groups, the incidence is reported to be between 0.4 and 0.6%.³⁻⁷ In children, however, it is 1.5 to 4% with a peak in the first two years of life.^{3,4,7,8} Regardless of age, the lateral ventricle is the most common site (50%), followed by the fourth (40%) and third (5%) ventricles. In 5% of cases, two or three ventricles are

involved.^{1,3,9} CPPs arising in the third ventricle and cerebellopontine angle (CPA) are extremely rare.^{3,6,7,10,11} We present a rare case of CPP arising in the CPA which extended to the cerebellar tonsil with adhesion to the lower cranial nerves. Differential diagnosis and immunohistochemical profile are also discussed.

Case report

A 49-year-old woman presented with a one-year history of headache and vomiting which had increased in frequency and intensity during the last two months. She also developed unsteady gait, swaying to the right side and right hearing loss. Neurological examination revealed mild bilateral papilledema with right gait ataxia and a diminished gag reflex on the right side. Audiometry showed mild right hearing loss. Routine investigations were within normal limits. Non-enhanced computerized tomography (CT) showed a homogenous isodense mass lesion in the

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Accepted for publication: 19 May 2010

right CPA. After contrast injection, intense and diffuse enhancement of the tumor was noted (Figure 1) which was surrounded by hypodense edematous tissue. The fourth ventricle was compressed and shifted to the left. Hydrocephalus was present and the tumor extended down to the cerebellar tonsil with adhesion to the lower cranial nerves as seen by magnetic resonance imaging (MRI).

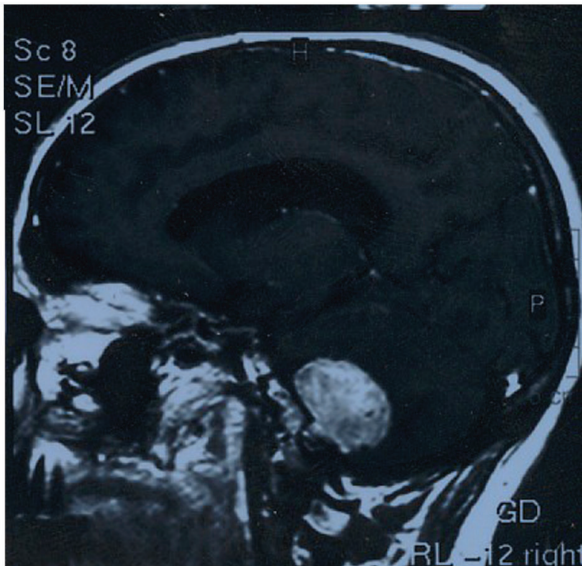


Figure 1. CT revealing a well-defined contrast enhanced mass at the right CPA

Craniotomy was performed with a provisional diagnosis of CPA meningioma or acoustic neuroma. On exploration, there was an encapsulated, white, soft, and focally hard mass with attachment to the cerebel-

lar tonsil and lower cranial nerves which were preserved. Histological examination revealed branching papilla with a fibrovascular core lined by a single layer of mildly pleomorphic columnar cells (Figure 2A). Foci of calcification, epithelial oncocyctic and signet ring changes were also present. Mitotic figures were very rare and there was no necrosis. Immunohistochemical studies of the tumor cells showed diffuse and strong immunoreactivity for pancytokeratin (CK), S100 protein, synaptophysin, and vimentin (Figure 2B). CK7 was focally expressed and CK20 was negative. Glial fibrillary acidic protein (GFAP) and epithelial membrane antigen (EMA) were focally positive, carcinoembryonic antigen (CEA) was weakly expressed and transthyretin (TTF-1) was negative. Less than 5% of cells revealed Ki67 positivity. A diagnosis of choroid plexus papilloma was made. Recovery was uneventful and the patient was discharged without any newly developed deficit.

Discussion

Choroid plexus papillomas that originate from the neuroectoderm are rare.^{7,10} The most frequent location in children is the lateral ventricle,^{2,3,6,8} whereas in adults it is the fourth ventricle.^{2,4,12} In 66.6% of adults CPP happens infratentorially while in 57.1% of children the tumor has been reported in the lateral ventricle.⁶ They rarely arise in the CPA, which represent about 9% of all CPPs, and are almost always found in adults.^{3,6,10,11,13} Direct extension of tumor through the foramen of Luschka, direct extension from the fourth ventricle or through CSF dissemination, are the pos-

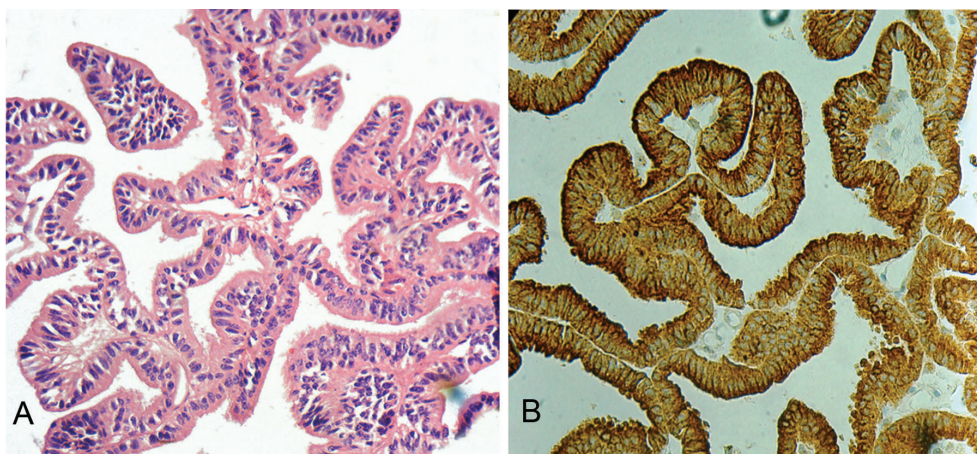


Figure 2. Microscopic view: A) papillary architecture (H&E, 250×). B) Synaptophysin expression (250×)

sible ways of CPA involvement.^{3,4} In this case, the tumor was situated in the right CPA with adhesion to the cerebellar tonsil and lower cranial nerves, as well as significant fourth ventricle compression and shifting toward the left side.

Overproduction of CSF by tumor or obstruction of the CSF pathway is responsible for the clinical manifestation, which is usually hydrocephalus (especially when the tumor is located in the 3rd or 4th ventricle).^{2,4,13} It is also suggested that spontaneous micro-hemorrhages with secondarily thickening of basal arachnoid and ependymitis can cause bilateral ventricular dilatation despite a unilateral lesion, or persistence of hydrocephalus even after complete removal of the tumor.⁶ Different symptoms are reported in different age groups. Headache is the most common symptom in both children and adults, with papilledema being the main clinical sign in adults (63.1%) and unsteady gait in children (71.4%) in Tacconi's study.⁶ Infratentorial tumors may present with sign and symptoms of cerebellar or brain stem dysfunction. The case presented with features of raised intracranial pressure (headache, vomiting, and papilledema) and ataxia.

By CT scan and MRI, CPP is a well defined lobulated lesion with irregular borders, a hypo and isodense lesion with marked enhancement on contrast. Calcification is reported in 4 to 10% of cases.^{1,5,12}

CPPs are soft, friable, gray-pink tumors on gross examination, with a well defined capsule⁴ or have a villiform or bosselated surface.² Gritty consistency or hardness is due to different degrees of calcification.² From a histological point of view, they resemble the normal choroid plexus with minimal nuclear atypia and few or no mitotic activity.^{1,2} At immunohistochemical studies, strong diffuse positivity for pancytokeratin (CK), CK7, S100 protein, synaptophysin, and vimentin, as well as focal expression of GFAP and EMA are reported. CK20 and CEA are not expressed.^{2,3} Our findings included strong diffuse positivity for CK, S100, synaptophysin and vimentin, focal expression of GFAP, EMA, and CK7, as well as negative CK20 and TTF1. CEA was weakly expressed.

The microscopic differential diagnosis includes papillary ependymoma and metastatic carcinoma, including papillary carcinoma of the thyroid. Ependymomas diffusely express GFAP but contain few, if any, CK positive cells and are negative for synap-

tochrome. Metastatic carcinomas more often show immunoreactivity for EMA, are less often S100 protein positive and express no GFAP (rarely GFAP positivity is reported). Papillary carcinoma is TTF1 and thyroglobulin reactive; whereas, thyroglobulin is negative and TTF1 is variable in CPP.^{2,14-16}

During the last decade, the morbidity and mortality of surgical treatment of CPP has significantly decreased. This is due to improvement of microsurgical and anesthetic techniques as well as better neuroimaging.⁶ The prognosis of CPP is very good.

Acknowledgement

We thank Ms. Mahnaz Khadem Huseini for typing this manuscript.

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