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Case Report

The cerebellar peduncle as localization of a recurrent atypical plexus papilloma: A case report

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ABSTRACT

Background: Choroid plexus papillomas (CPPs) are infrequently encountered brain tumors with the majority originating in the ventricular system. Rarely, CPP occurs outside of the ventricles.

Case Description: We report the case of a recurrent CPP that initially originated within the fourth ventricle, though years later it recurred in the left middle cerebellar peduncle.

Conclusion: Patients with cerebellar plexus papilloma need long-term follow-up comprising regular magnetic resonance imagings since, in patients with a history of CPP, any new mild symptomatology, even years after the initial presentation, may be an early sign of tumor recurrence.

Keywords: Brain tumors, choroid plexus papilloma, Nystagmus

INTRODUCTION

Choroid plexus papillomas (CPPs) are rather rare brain tumors originating from the epithelium of the CP. Some of the CP lies along the velum medullare inferius of the fourth ventricle and is responsible for the production of the cerebrospinal fluid (CSF). The tumor classification of the World Health Organization (WHO) differentiates between benign forms, CPP (Grade I), atypical forms (Grade II), and malignant forms, such as the CP carcinoma, CPC (Grade III).[5,14] CPPs constitute 0.3–0.6% of all brain tumors[6] with a male-to-female ratio of 2:1.[10] They represent 1.5–4% of pediatric intracranial tumors and 0.5% of intracranial brain tumors in adults.[8] Fifty percent of all CPPs originate from the lateral ventricle, 40% from the fourth ventricle, and 5% are either found in the third ventricle or in multiple ventricles.[8,11] Symptomatology differs according to tumor location: third ventricle CPPs were described with seizures, vision disturbances, and hemiparesis, while fourth ventricle CPPs were associated with headache, ataxia, vomitus, and visual symptoms.[11] CPPs occur only rarely outside of the ventricles with a few case reports

describing CPPs at the cerebellopontine angle, the suprasellar region, and the posterior fossa.[13]

Here, we report a case of a recurrent CPP that originated from the fourth ventricle, while years later, it reoccurred in the left middle cerebellar peduncle (MCP). To the best of our knowledge, this is the first description of a CPP localized in the MCP.

CASE REPORT

In 2010, a 52-year-old female patient complained of nausea present for more than 6 months, vomitus, intermittent vertigo, loss of appetite with B-symptoms, and weight loss of 10 kg. The initial diagnostic workup consisted of gastrointestinal imaging and a thorax X-ray, which were normal. However, a cranial magnetic resonance imaging (MRI) showed a tumor mass within the fourth ventricle (3 cm × 2 cm of size [Figures 1 and 2]); the differential diagnosis included an ependymoma, a plexus papilloma, and an intraventricular meningioma. A suboccipital craniectomy with tumor resection was performed at the University Hospital Basel with histopathological evidence of an atypical CPP WHO Grad II. One day postoperatively, the patient developed a hydrocephalus due to the fourth ventricle outflow obstruction secondary to a blood clot. For immediate treatment of the elevated intracranial pressure, a ventricular drainage was placed. The hematoma was subsequently removed to restore the physiological dynamics of the CSF. A few days later, the patient presented with diplopia and paralysis of the left-sided glossopharyngeal nerve and the hypoglossal nerve requiring an emergent tracheotomy. Nine days after the initial tumor resection (i.e., 7 days after external ventricular drain placement and evacuation of the hematoma), the patient developed double vision and nystagmus. A computed tomography (CT) head showed a new epidural hematoma at the site of the suboccipital craniotomy. The hematoma was subsequently removed with resolution of the symptoms. At this point of time, the ventricular drainage was still in place. CSF analysis revealed an elevated cell count, and after consultation with the infectious disease team, IV antibiotic treatment with rifampicin and ceftriaxone was initiated. Due to the absence of bacteria in the CSF (negative gram stain and cultures) and normal white blood cell and CRP findings, the antibiotic treatment was stopped after 5 days. Three days postoperatively, the patient developed a left-sided hemiparesis due to ischemia in the right parietal lobe as diagnosed by MRI scan. Since subsequent cardiologic workup (transesophageal echocardiography with contrast agent), an extra- and intracranial Doppler ultrasound, and blood chemistry were unremarkable, the diagnosis of a cryptogenic stroke was made. The left-sided hemiparesis persisted. After a period of inpatient surveillance, the patient was referred to our rehabilitation center and underwent an intense neurorehabilitation program. After several weeks, she was able to be discharged from our rehabilitation center, walking independently. Brain imaging during the 2 following years did not show any new pathological findings.

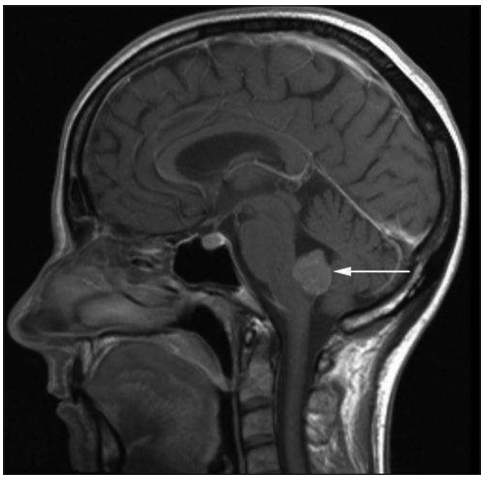

Figure 1: Image of the primary tumor, an intraventricular atypical choroid plexus papilloma. T1-weighted sagittal image with contrast shows a homogeneously enhancing lesion in the inferior fourth ventricle (arrow).

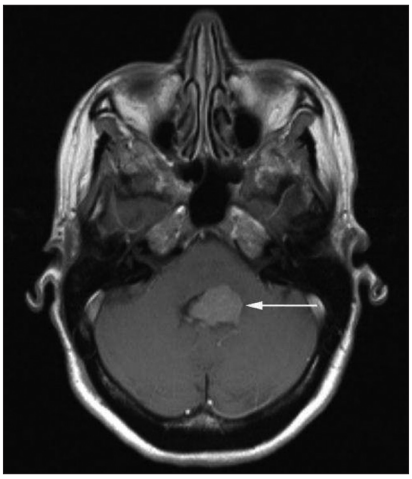

Figure 2: Image of the primary tumor, an intraventricular atypical choroid plexus papilloma. T1-weighted axial image with contrast shows a homogeneously enhancing lesion in the fourth ventricle with contact to and impression on the left middle cerebellar peduncle (arrow).

Seven years later (2017), the patient complained again of two episodes of paroxysmal vertigo. Ophthalmological examination revealed a bilateral upbeat nystagmus. In addition, the patient reported a mild headache and bilateral pulse-synchronous tinnitus. Neurological examination showed a mild left-sided limb ataxia. She continued to ambulate without a walking aid. In September 2017, a cranial MRI showed a localized, inhomogeneous contrast-enhancing tumor mass in the middle left cerebellar peduncle (circa 17 mm × 17 mm) [Figure 3]. On a follow-up MRI in February 2018, a further increase in size to 22 mm was noted, without any evidence of obstruction of the ventricular system. There were no signs of a metastatic disease detectable on the thoracoabdominal CT scan. After detection of the new lesion on the MRI, the patient initially refused surgical intervention and opted for MR follow-up. After 3 months, with

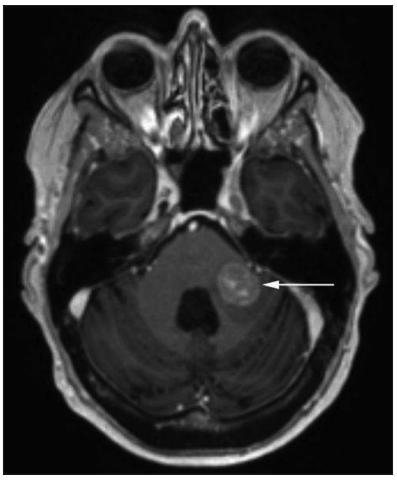

Figure 3: Image of the recurrent atypical choroid plexus papilloma in the middle cerebellar peduncle. T1-weighted axial image with contrast shows an enhancing intraparenchymal lesion with surrounding edema (arrow).

increasing symptoms and further growing of the new lesion on the follow-up MRI, she decided for surgical treatment. In March 2018, a left retrosigmoid craniectomy with tumor resection was performed. Histopathological examination revealed an atypical CPP (WHO Grade II). Post-operative irradiation was recommended. Due to the patient's wish (i.e., fear of worsening of neurological deficits), post-operative irradiation was not performed and is planned in case of progression of the remaining contrast-enhancing area on MRI. After the surgical intervention, the patient re-entered our rehabilitation clinic for the 2nd time. The follow-up examination in June 2018 showed a satisfactory course. Improvement was seen in coordination, trunk-stability, balance, and gait, resulting positively in a reduced tendency to fall; only for outside walks, she had still to use a stick. The clinical improvement translated equally into objective assessment scales. The Functional Independence Measure test improved from 96 to 112 points (maximal score possible 126), and on the Mini-Balance Evaluation Systems Test, she scored 22 of 28 possible points. On the Functional Ambulation Categories test, the patient scored 5 out of 5 points. In the 6-min walk test, she achieved 451 m (reference 700–800 m).

DISCUSSION

Neuroanatomy

The cerebellum is structurally connected to the brainstem through six symmetrically positioned peduncles. The two inferior cerebellar peduncles project on the medulla oblongata, the MCPs adjoin it to the pons, while the superior CPs represent the link to the midbrain.[7] A lesion of the MCPs causes ipsilateral symptoms.[7]

Typical symptoms after MCP lesion

Typically, MCP lesions manifest with a vestibular syndrome (oculomotor symptoms, vertigo, and vomitus) or motor symptoms. At the time of tumor recurrence, our patient presented with an acute vestibular syndrome characterized by an upbeat nystagmus. Distinct eye findings appear to be common with MCP lesions. Kim and Kim[4] described an acute vestibular syndrome in 23 patients with acute strokes of the unilateral MCPs with distinct abnormal eye movements. The authors hypothesized that the MCP lesions may cause damage to neural circuits involved in eye position stabilization. Damage of the medial longitudinal fasciculus (MLF) and ventral tegmental tract are discussed as potential causes of an upbeat nystagmus.[3,9]

Complementary observations were reported in other clinical settings: Hall *et al*.[1] described a 61-year-old man with an acute infarct of the left MCP who presented with a new gaze-evoked nystagmus that persisted when looking horizontally to the left.[1] Sharma *et al*.[12] described a 70-year-old male patient with acute onset of vertigo associated with two episodes of vomitus, truncal ataxia, and dysarthria. Neurological examination showed a bilateral horizontal nystagmus. Imaging evidenced lesions in both MCPs and a small right pontine lesion. Jang and Kwon[2] reported a patient who presented with ataxia and tremor arising from a cortico-ponto-cerebellar tract (CPCT) lesion following mild traumatic brain injury.

CONCLUSION

Recurrence of CPP is rare, especially in extraventricular regions. However, in patients with a history of CPP, even years after the initial presentation, one should keep in mind that a mild symptomatology, such as vomitus or vertigo (even without eye symptoms), may already be early signs pointing to tumor recurrence. Therefore, patients with cerebellar plexus papilloma need long-term follow-up with sequential MRIs.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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