CASO CLÍNICO/CASE REPORT

Fourth Ventricle Cystic Choroid Plexus Papilloma: An Unusual Presentation

Variante Quística do Papiloma do Plexo Coroideu no Quarto Ventrículo: Uma Apresentação Invulgar

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Abstract

Choroid plexus papilloma (CpP) is a rare, mostly paediatric tumour. Despite occurring predominantly in intraventricular supratentorial compartment, in adults, CpP is mostly infratentorially (70% IV-ventricle). Usually, CpP is a solid tumour. Cystic forms are uncommon. CpP highly enhances with gadolinium in magnetic resonance imaging, even in the cystic form. Hydrocephalus is common in infratentorial CpP.

A 26-year-old healthy female progressively developed intracranial hypertension symptoms, cerebellar signs (ataxia, nystagmus) and right-side facial paresis. Head magnetic resonance imaging revealed hydrocephalus and a fourth ventricle cyst, with mass effect, without gadolinium enhancement. A craniotomy with cyst fenestration and a reddish mass resection were performed. Histology revealed a CpP. Patient gradually improved with complete symptom resolution.

We present a rare presentation of a rare tumour. Cystic lesions in posterior fossa are vast and we must consider CpP as a possible diagnosis. Treatment is surgical and requires complete tumour resection. Hydrocephalus might require shunting.

Resumo

O papiloma do plexo coroideu (CpP) é um tumor raro, maioritariamente pediátrico. Apesar de ocorrer predominantemente no compartimento intraventricular supratentorial, em adultos, o CpP é principalmente infratentorial (70% no IV ventrículo). Geralmente, o CpP é um tumor sólido. Apresentações císticas são incomuns. O CpP capta avidamente gadolínio na ressonância magnética, mesmo na forma cística. A hidrocefalia é comum no CpP infratentorial.

Uma mulher saudável de 26 anos desenvolveu progressivamente sintomas de hipertensão intracraniana, sinais cerebelosos (ataxia, nistagmo) e paresia facial do lado direito. A ressonância magnética craniana revelou hidrocefalia e uma lesão cística no quarto ventrículo, com efeito de massa, sem realce por gadolínio. Foi realizada uma craniotomia com fenestração do cisto e ressecção de uma massa avermelhada. A histologia revelou um CpP. O paciente melhorou gradualmente com a resolução completa dos sintomas.

Apresentamos uma apresentação rara de um tumor raro. As lesões císticas na fossa posterior são vastas e devemos considerar a CpP como um possível diagnóstico. O tratamento é cirúrgico e requer ressecção completa do tumor. Para a hidrocefalia pode ser necessária derivação de líquor.

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Introduction

As choroid plexus papilloma (CpP) are rare tumours counting for 05% -0.6% of all brain tumours. CpP are more frequent in the paediatric ages accounting for 1.5% to 6% of all Central Nervous tumours in this group - 70% of CpP occur before 2 years of age being more commonly found in the supratentorial compartment.[1,2] CpP are slightly more common in males (1,2:1).[3] In a 1979 review of 209 CpP, it was found that these are mainly intraventricular tumours being more common on lateral ventricles (78%), followed by fourth and third ventricles (16% and 5.7% respectively).[4] In adults, CpP are more common on the infratentorial compartment with 70% occurring in the fourth ventricle.[2] CpP are usually solid tumours macroscopically and on imaging – on head computed tomography (CT) scanning, CpP are a round or lobulated mass, isodense or hyperdense, with punctate calcification and strongly enhances with intravenous contrast.[5] Hydrocephalus is highly common, and many times dominates the clinical presentation. In magnetic resonance imaging (MRI) CpP are isointense to brain tissue on T1 weighted images (hyperintense areas suggest haemorrhage and/or necrosis) and avidly enhances after gadolinium administration.[6]

Pure cystic CpP are extremely rare and makes the diagnosis challenging, especially in adult patients.[7-9] Very few cases have been reported so far and differential diagnosis with morphological identical lesions on the same location is difficult.

A case of a young female with a cystic fourth ventricle CpP is presented and a review on the subject is performed.

Case Report

A 26-year-old female, without relevant past medical history, presented to the Emergency Department complaining of headache, gait imbalance, nausea and vomit, rotatory vertigo and phonophobia for a week. She reported a progressive symptom worsening. She was evaluated by Otorhinolaryngology which documented a left-side nystagmus, unstable Romberg test without dysmetria or ataxia. She was hospitalized with the diagnosis of labyrinthitis. Later, she worsened from headache (mostly in decubitus), nausea and vomiting and developed vertical nystagmus, diplopia, right side facial central paresis and a marked gait ataxia. Eye fundoscopic examination revealed bilateral papilledema.

Patient was studied with cranial CT which revealed tetraventricular ventriculomegaly, mainly pronounced on the fourth ventricle. Patient was then studied with a cranial magnetic resonance imaging (MRI) which revealed a tetraventricular hydrocephalus with transependymal transudation and a cystic formation on the fourth ventricle with mass effect over brainstem anteriorly and over cerebellar vermis posteriorly. After gadolinium administration, no abnormal enhancement was observed **(Fig. 1)**.

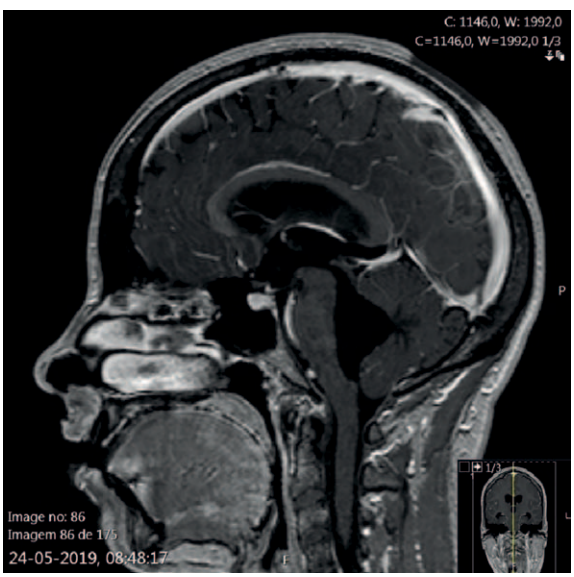

Figure 1. T1 sequence, Sagittal cut MRI with gadolinium. Tetraventricular hydrocephalus. A cystic formation on fourth ventricle with mass effect over brainstem anteriorly and over cerebellar vermis posteriorly. No abnormal gadolinium enhancement.

A suboccipital median craniotomy was performed after placing an external ventricular drainage (EVD) on the right-side Frazier's point **(Fig. 2)**. A thick arachnoid membrane was found over Magendie foramen. After its

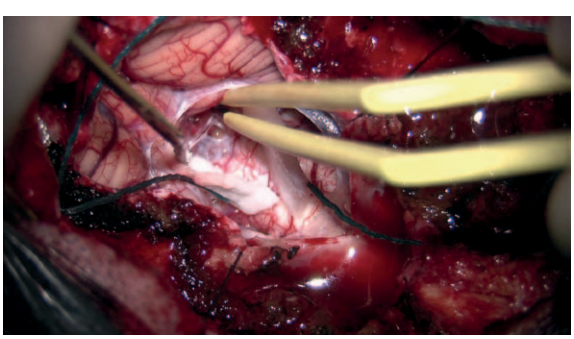

Figure 2. After cystic membrane fenestration, a reddish mass, distinct from choroid plexus, was completely resected.

aperture, the cystic clear fluid (similar to cerebrospinal fluid) collection was evacuated and in the anterior surface of the lower vermis (uvula), a reddish mass, distinct from choroid plexus, was observed and completely removed **(Fig. 2)**.

Histological examination revealed a World Health Organization grade I CpP **(Fig. 3)**. Cytological cerebrospinal fluid evaluation was negative for neoplastic cells.

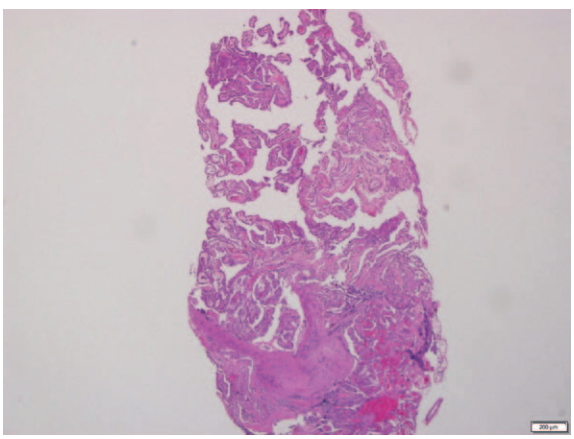

Figure 3. A papillary architectural pattern neoplasm constituted by eosinophilic columnar cells with round nuclei and granular chromatin. No mitotic figures were observed. These findings are compatible with a Grade I choroid plexus papilloma.

After surgery, EVD was kept closed and removed on fourth post-operative day. Patient immediately improved from headache, nausea and vomiting after surgery. Gradually, an improvement was observed on nystagmus, diplopia and on facial paresis over the next 3-4 days. She started rehabilitation with progressive resolution of the gait ataxia over the following weeks. Post operative MRI, performed 3 months after surgery, revealed a reduction on fourth ventricle size and on the compression over brainstem and cerebellar vermis **(Fig. 4)**. Active hydrocephalus was resolved and, as in pre operative MRI, no gadolinium enhancement was observed. Patient continues under clinical and periodical imaging follow up. Nine months after surgery, patient remained asymptomatic.

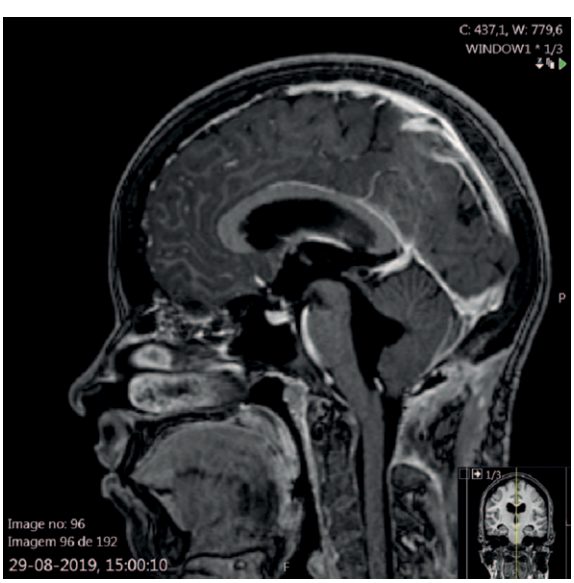

Figure 4. Post-operative MRI. Ventriculomegaly reduction. Decrease on fourth ventricle size and on compression over brainstem and cerebellar vermis.

Discussion

Primary CpP are rare tumours and presents mainly in paediatric ages. Usually, these choroid plexus tumours are benign lesions but malignant rates have been reported in about 10% - 20%.[10] Choroid plexus tumours are classified according to World Health Organization (WHO) classification in choroid plexus papilloma (WHO grade I), atypical choroid plexus papilloma (WHO grade II), and choroid plexus carcinoma (WHO grade III).[11]

Posterior fossa is the most common location on adult patients, mostly intraventricularly but cerebellopontine lesions had been described.[12] Mean age reported for CpP diagnosis in fourth ventricle is 22.5 years[13] which is consistent with the reported case.

The clinical presentation is usually gradual. In supratentorial lesions, symptoms might be seizures, cognitive changes and focal motor deficit while in posterior fossa (as observed in the reported case), manifestations include headache, nystagmus, ataxia, dizziness, papilledema with loss of vision, vomiting, and diplopia.[14] Similarly to colloid cysts, sudden death has been reported from ventricular blockage, particularly in third ventricle lesions.[15] Hydrocephalus is highly frequent and might result from a combination of several mechanisms – cerebrospinal fluid (CSF) overproduction, circulation blockage or impaired absorption.[16] In cystic CpP variant, CSF overproduction appears to be the main mechanism and acute hydrocephalus is less common.[17]

CT and MRI usually present a solid mass with intense contrast enhancement.[18] CpP with cystic presentation is very rare and other diagnostic possibilities shall be considered including malformative, infectious and neoplastic pathologies.[19] Considering malformative congenital pathologies, arachnoid/ependymal cysts might present as intraventricular lesions. Fourth ventricle entrapment

can occur after chronic shunting from lateral ventricles and after infections such as ventriculitis – neither was experienced by the reported patient. These cystic lesions do not enhance after contrast administration. Infectious conditions such as neurocysticercosis must be considered since opening this infectious cyst may lead to a severe chemical meningitis.[8,19] CpP, similarly to hemangioblastoma and pilocytic astrocytoma, may present a cyst with one (rarely more) mural nodules with contrast enhancement, however mural nodule absence with only capsular enhancement might be seen.[7,9] In the reported patient, no contrast enhancement was observed due to mural nodules absence nor capsular enhancement. This case report highlights that no contrast enhancement may occur in cystic CpP.

Macroscopically, CpP are digitiform friable reddish, high vascularized soft masses such was observed during surgery **(Fig. 4)**. Histologically, it characterizes by papillary ferns lined by a columnar epithelium. The visualization of nuclear atypia, mitotic figures and necrosis is suggestive of a more malignant form – atypical CpP or papillary carcinoma.[11,20]

CpP treatment is surgical and aims for a complete resection. Due to the risk of acute obstructive hydrocephalus, in some occasions an external ventricular drainage or shunting is required, even after complete resections. If clinical suspicion for neurocysticercosis is high, a craniotomy shall be employed to avoid cyst rupture.[8] In some cases, endoscopic surgery might allow a satisfactory resection in supratentorial lesions. It is important to excise all the enhancement portions of the tumour to avoid relapse. We advise EVD placement during the initial surgical stages in order to avoid herniation during dural opening or an acute hydrocephalus in the post operative period.

CpP presents a good long-term prognosis and complete surgical removal is usually curative. Patients require clinical and periodic MRI surveillance since tumour recurrence might occur. Despite infrequent, CSF metastases have been reported.[21] ■

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