Clinical Case Reports

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Infratentorial choroid plexus tumors in children: Case report and review of literature

Amanda de Oliveira López[1], Patricia Dastoli[2], João Gabriel Ribeiro Gomes[1], Jardel Mendonça Nicácio[2], Marcos Devanir Silva da Costa[2], Sergio Cavalheiro[2]

[1] Departament of Pediatric Neurosurgery, Real Hospital Português, Recife, Pernambuco, Brasil

[2] Department of Neurology and Neurosurgery, Universidade Federal de São Paulo, São Paulo- SP, Brazil

✉ Amanda de Oliveira Lopez, MD

e-mail: lopezamanda1@gmail.com

Available at:
http://www.archpedneurosurg.com.br/

Background: Infratentorial choroid plexus tumors are rare lesions in the pediatric population. At diagnosis, they most often present with signs of intracranial hypertension and cranial nerve deficits. The main objective of the surgery is total resection and the prognosis is favorable in most cases.
Case Report: A 16-year-old boy with gait disturbance, headaches and left side hypoacusia had a diagnosis of posterior fossa tumor with hypertensive hydrocephalus. He underwent urgent placement of a ventriculoperitoneal shunt and subsequently microsurgery with gross total resection. At follow up, the patient was asymptomatic and without deficits.
Conclusion: Choroid plexus tumors should be included in the differential diagnosis of posterior fossa tumors in childhood. Preoperative planning, choosing the best surgical approach and interruption of the lesion's vascular supply as the initial step of surgery are fundamental strategies for successful treatment.

Keywords: Choroid Plexus Tumors, Posterior Fossa, Pediatric

INTRODUCTION

Choroid plexus tumors (CPT) are uncommon pathologies located preferentially in the supratentorial compartment in the pediatric population and in the posterior fossa in adults. We present a case of an infratentorial choroid plexus tumor in a 16-year-old child with complete resection. There are few reports in the literature of cases similar to this one. In this article we will discuss the main aspects concerning the clinical presentation, imaging, treatment and prognosis of choroid plexus tumors of the posterior fossa in the pediatric population.

CASE REPORT

Boy, 16 years old, with history of gait disturbance, headaches and left hypoacusia that started a few months before hospitalization. There was an acute worsening of the symptoms, which led the patient to emergency department. On admission exam, he presented with: disorientation, gait ataxia and left-sided dysmetria / dysdiadochokinesia. Fundoscopy revealed bilateral papillary edema.

A CT scan showed hypertensive hydrocephalus and a lesion with contrast enhancement, multilobulated, occupying most of the fourth ventricle, extending to the left cistern of the cerebellopontine angle (CPA). (figure 1)

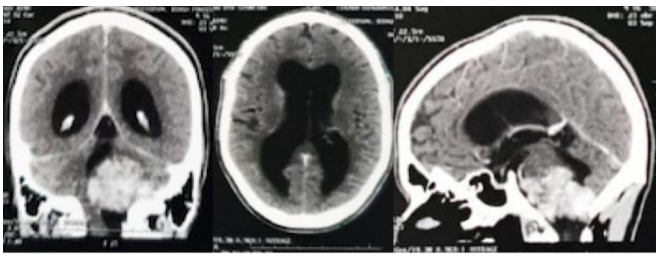

Figure 1- CT scan showing an enhanced contrast lesion centered in the fourth ventricle and occupying the left CPA region with associated hydrocephalus.

An urgent placement of a ventriculoperitoneal shunt was performed for the hydrocephalus.

The patient evolved well postoperatively with resolution of the intracranial hypertension symptoms. He remained with left hypoacusia and coordination deficit.

A brain MRI showed a large Hypo-isointense tumor on T1 with contrast enhancement and Hyper signal on T2, associated with brainstem compression. (figure 2)

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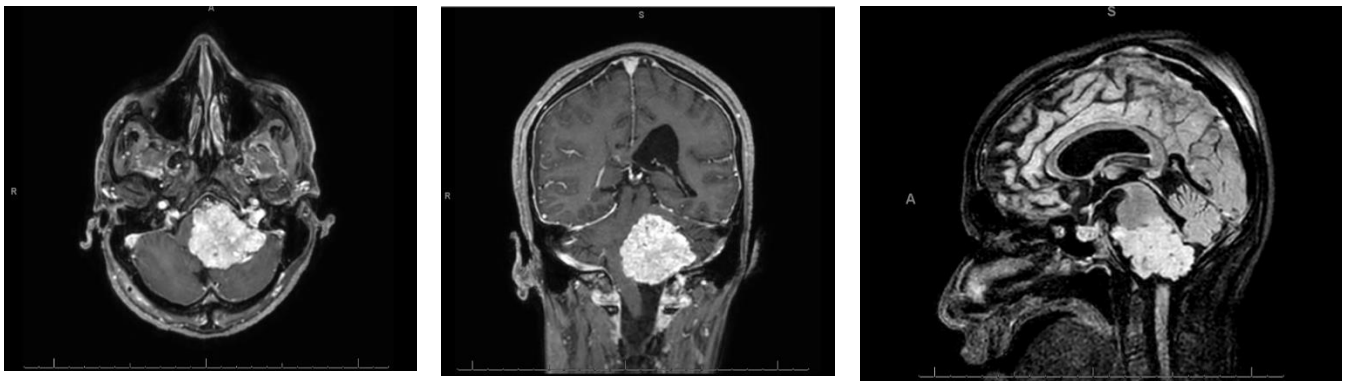

Figure 2- Axial and sagittal magnetic resonance images showing intense, homogeneous contrast enhancement of the lesion. Important extension to the left cerebellopontine angle can be noted (a and b).Sagittal FLAIR image showing brainstem compression

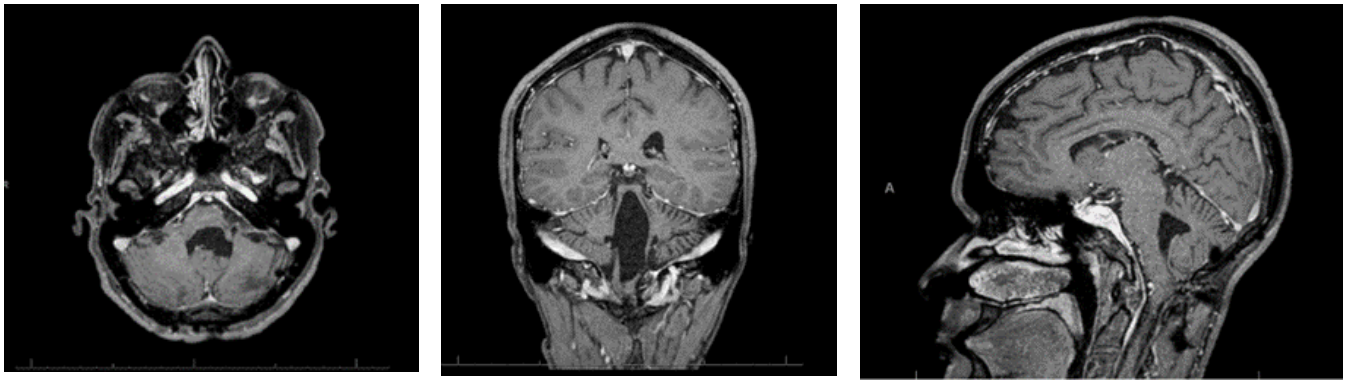

Figure 3- Postoperative MRI contrast enhanced T1-weighted axial image (a), coronal (b) and sagittal image (c) showing absence of residual tumor

Few Days after placement of the shunt, he underwent microsurgery through a median suboccipital approach. (figure 3)

At the surgery, the lesion origin was identified in the choroid plexus of the fourth ventricle, with vascular nutrition by posterior inferior cerebellar artery (PICA) branches. Coagulation of the supplying artery allowed complete resection of the lesion without significant bleeding. No blood transfusion was necessary.

Tumor resection was complete including the portion extending into the CPA through Luschka foramen.

Pathology and immunohistochemistry confirmed Choroid Plexus Papilloma (CPP).

The patient evolved well and a few days after surgery he was asymptomatic. We maintained the Ventricular Peritoneal Shunt (VPS) postoperatively.

On 2-years follow-up, there was no evidence of lesion recurrence.

DISCUSSION

Concept

Choroid plexus tumors (CPT) are rare pathologies and represent 1-4% of all brain tumors in childhood, with a higher incidence in the first year of life in all subtypes. (1)

According to the 2016 WHO classification of tumors, choroid plexus tumors have 3 pathological subtypes: Choroid plexus papillomas (CPP) - WHO Grade I, Atypical Choroid Plexus Papillomas (aCPP) - WHO GRADE II and Choroid Plexus Carcinomas CPC- GRADE III WHO. (2) The classification of choroid plexus tumors remained unchanged in the 2021 update, although they are now listed separately from glial and glioneuronal neoplasms. (3)

Regarding location, choroid plexus tumors can be: INTRAVENTRICULAR (most cases) or EXTRAVENTRICULAR (through ectopic choroid plexus tissue due to metaplasia or cerebrospinal fluid dissemination). (4,5,6,7) In children, the most common location is the lateral ventricles. Infratentorial lesions are rare in childhood, with predilection for older patients in relation to supratentorial ones. (8) Trybula et al published an average age of 9 years in cases of infratentorial lesions. (9) In our case the patient was 16 years old when diagnosed.

Among Infratentorial lesions, the most frequent locations are IV ventricle, cerebello-pontine angle (CPA) and Cerebellomedullary cistern (in descending order). (10)

In the case presented, the lesion was primarily centered in the fourth ventricle, but extended to CPA region.

CPT arising from CPA are rare during childhood. The most recent systematic review by Gaddi et al showed only 11 cases published in the literature. (9, 8)

Anatomical Aspects

The choroid plexus of the fourth ventricle has as its main vascular supply the PICA (roof and Magendie foramen) and the Anterior Inferior Cerebellar Artery - AICA (CPA and lateral recess).

According to previous studies, most of the CPTs occupying fourth ventricle have the PICA as their main feeder artery, while lesions primarily in the CPA have the AICA as their main feeder. (6,11) In the case of tumors with intraventricular and extraventricular components, it is possible to have supply from both arteries (AICA and PICA). (9,6)

Surgery

Adib et al proposed an anatomical subclassification of extraventricular CPT of the posterior fossa that serves as a tool to facilitate the choice of surgical access: Type 1- tumor in the CPA without any portion into the cerebellomedullary angle (CMA)

(give preference to the retrosigmoid approach in semi-sitting position); Type 2- Tumor located in the CMA without any portion in the CPA (suggests subtonsillar median suboccipital with patient in prone position) and Type 3- Tumor located in the CPA and CMA. The authors recommend this classification in the preoperatory for surgical planning, since the relationship and proximity of the tumor to the cranial nerves varies according to the type of lesion (cranial nerves VII and VIII in CPA lesions versus lower cranial nerves in lesions centered on the CMA). (10)

The choice of the best surgical approach for infratentorial CPT should evaluate not only the location of the tumor but also vascular aspects, since the initial objective is to access the main arterial feeders to facilitate the resection and avoid hemorrhagic complications. Unlike supratentorial lesions (which are known to be hypervascularized tumors), infratentorial CPTs are less vascularized, especially those with an extraventricular location.(9,8) This fact justifies a lower rate of hemorrhagic complications in posterior fossa CPT. (9,8) In the case operated by our team, the lesion was not hypervascularized and there was no technical difficulty in bleeding control.

For CPA lesions,in previous studies, the suboccipital retrosigmoid is the standard approach. It is important to emphasize that lesions originated in the 4th ventricle, even with CPA extension, may benefit from the median suboccipital access through the cerebellar-medullary fissure with the primary objective of interrupting the vascular pedicle of the tumor before debulking (12,13,10). We used this approach, and despite the large size of the tumor, it was completely resected without additional complications. After interruption of the main vascular supply, piecemeal resection can be performed safely. (12)

Despite the importance of knowing the vascular anatomy in preoperative planning, contrary to what was done in the past, digital angiography is currently not routinely indicated, especially in posterior fossa lesions in pediatric population. The main reasons are the risk inherent to the procedure and thin vessels supplying this region. (10,14)

Preoperative high-quality resonance imaging can provide the necessary tumor information, and suggesting its origin from the vascular supply. (6)

Standard surgery is total tumor resection (which was achieved in most cases reported in the literature). If the lesion has an infiltrative appearance on the floor of the fourth ventricle, it is preferable to perform safe partial resection. Small residual lesions, usually, do not recur in the follow-up, even in the absence of adjuvant therapy, given the benign aspect of most lesions. (9)

According to Panniza et al, when total resection of the CPP is not achievable, intraoperative coagulation of the remaining lesion may reduce the chance of local recurrence. (14)

Preoperative Embolization

Due to the hemorrhagic characteristics of CPT some authors recommend preoperative transarterial embolization of vessels supplying the tumor in order to minimize surgical bleeding. (REF 10). Therefore with considerable risk of additional complications, this practice has fallen into disuse, especially in infratentorial tumors that have less vascularization than supratentorial. (15)

Hydrocephalus

Most patients with infratentorial CPT present with hydrocephalus at diagnosis, which is due both to CSF hyperproduction and to obstructive factors, and less frequently to disabsorptive factors (impaired absorption - due to hemorrhage or hyperproteinorrhachia produced by the tumor). (16,17)

In cases of lesions originating from the CPA, a lower rate of hydrocephalus (67%) is observed in the literature when compared to infratentorial intraventricular CPT. Previous

reports discuss that this difference is due to the lower obstructive component of CPA lesions and lower CSF production, as they are less vascularized tumors. (6,8,18,19)

As complete tumor resection is achieved in most cases (with resolution of obstructive factors and overproduction of CSF), only 27% of cases remains shunt dependent and most of them hydrocephalus persists due to disabsorptive problems. (8,16)

Regarding the best treatment for hydrocephalus and the choice between external ventricular drainage (EVD), endoscopic third ventriculostomy (ETV) or VPS is still controversial.

As most cases will be shunt independent (about 80%) (20), the EVD (before or during the tumor microsurgery) seems to be a good option: intracranial pressure is controlled and allows blood and protein residues drainage, further reducing shunt dependence rate in the postoperative period.

Although some authors advocate ETV as the first treatment option (8), this is a matter of discussion (10), and VPS is most widely performed for persistent postoperative hydrocephalus.

In the case reported, patient underwent VPS placement at another hospital on an emergency basis and was referred to our hospital days after the shunt. Although total resection of the lesion was performed, patient required maintenance of VPS postoperatively.

Adjuvant Therapy

For CPP with total resection there is no indication of complementary therapy. For cases with partial resection and small residual lesions, observation and follow up with imaging, is a good alternative because of small recurrence rates.

The use of chemotherapy and radiotherapy should be evaluated in cases of malignant lesions or recurrence, as CPCs are 20 times more likely to have local recurrence or metastases (20) . Di Rocco et al (15) reported a case treated with preoperative chemotherapy for a giant lesion, to reduce vascularization and tumor volume and facilitate subsequent surgical approach. Despite the reported success in this case, most authors did not adhere to this practice (18).

Survival

From a general point of view, choroid plexus papillomas have a good prognosis. Patients with Choroid Plexus Carcinomas have a 5 times higher mortality rate than those with papillomas. (1) Surgery plays a fundamental role in the prognosis with increased survival (close to 100% in 10 years) in cases of total resection of the PPC. (9,21)

CONCLUSION

Infratentorial tumors of the choroid plexus are rare pathologies in childhood. Surgery with total resection is the standard treatment. An important step in surgery is early vascular control of the supplying arteries. Adjuvant therapy is reserved only for cases of aggressive lesions and with recurrence. Given the benign characteristics of these lesions and the high chances of total surgical resection, most cases have a good long-term prognosis.

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DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the local Ethics Committee,

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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CONTRIBUTIONS

-**Amanda de Oliveira López**: Methodology, Project administration, Resources, Writing – original draft, Writing – review & editing
-**Patricia Dastoli**: Conceptualization, Data curation, Visualization
-**João Gabriel Ribeiro Gomes**: Resources, Writing – original draft, Writing – review & editing
-**Jardel Mendonça Nicácio:** Data curation, Methodology
-**Marcos Devanir Silva da Costa**: Data curation, Software
-**Sergio Cavalheiro**: Data curation, SupervisionData curation,

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