



## Case Report

# Spinal drop of atypical choroid plexus papilloma in an adult patient: A case report and literature review

Diego Ochoa-Cacique<sup>1</sup>, María Córdoba-Mosqueda<sup>1</sup>, José Ramón Aguilar-Calderón<sup>1</sup>, Martha Cristina Sánchez-Silva<sup>2</sup>, Rosa María Vicuña-González<sup>3</sup>, Abraham Ibarra-de la Torre<sup>1</sup>, Victor Andrés Reyes-Rodríguez<sup>4</sup>, José de Jesús Lomeli-Ramírez<sup>5</sup>, Óscar Medina-Carrillo<sup>1</sup>, Mauricio Daniel Sánchez-Calderón<sup>1</sup>, Erick Alberto Castañeda-Ramírez<sup>1</sup>, Ulises García-González<sup>1</sup>

Departments of <sup>1</sup>Neurosurgery, <sup>2</sup>Imaging, <sup>3</sup>Anatomical Pathology, Hospital Central Sur de Alta Especialidad PEMEX, Tlalpan, Ciudad de Mexico, Mexico, <sup>4</sup>Department of Neurosurgery, Hospital Central Norte PEMEX, Azcapotzalco, Estado de Mexico, Mexico, <sup>5</sup>Department of Neurosurgery, Hospital Vossan, Campeche, Mexico.

E-mail: \*Diego Ochoa-Cacique - diego2\_doc@hotmail.com; María Córdoba-Mosqueda - dramaelenacmosqueda@gmail.com; José Ramón Aguilar-Calderón - jcalderon02@yahoo.com; Martha Cristina Sánchez-Silva - marthacss@gmail.com; Rosa María Vicuña-González - vicus67@yahoo.com.mx; Abraham Ibarra-de la Torre - abraham\_ibarra2017@outlook.com; Victor Andrés Reyes-Rodríguez - neurovican@hotmail.com; José de Jesús Lomeli-Ramírez - joselomeli@prodigy.net.mx; Óscar Medina-Carrillo - dromecar@gmail.com; Mauricio Daniel Sánchez-Calderón - maurisancal@gmail.com; Erick Alberto Castañeda-Ramírez - erickcastanedar@gmail.com; Ulises García-González - ulises.med@gmail.com



### \*Corresponding author:

Diego Ochoa-Cacique,  
Department of Neurosurgery,  
Hospital Central Sur de Alta  
Especialidad PEMEX, Tlalpan,  
Ciudad de Mexico, Mexico.

diego2\_doc@hotmail.com

Received : 16 September 2020

Accepted : 22 October 2020

Published : 25 November 2020

### DOI

10.25259/SNI\_649\_2020

### Quick Response Code:



## ABSTRACT

**Background:** Choroid plexus papillomas (CPPs) are benign extra-axial tumors that originate from the choroid plexus; these tumors rarely have metastases, being at the spinal level the location with few reported cases.

**Case Description:** We report the case of a 48-year-old man with a history of atypical fourth ventricular CPP and gross total resection (GTR) in 2008. In 2015, he presented with radicular pain, decreased strength, and paresthesia in the left leg. Magnetic resonance imaging revealed a well-defined intradural ovoid lesion in the vertebral canal at the level of the L3-L4 intersomatic space. Subtotal resection of the tumor was performed. The patient recovered well, with relief of pain and no neurological deficit. A literature research few cases of CPP metastasis in adults. We describe here a fifth-decade male patient with a lumbar neoplasm, which according to the histopathologic characteristics and location is the first case of an atypical papilloma implant of the choroid plexus at this spinal level.

**Conclusion:** GTR of primary lesions and associated implants remains the gold standard for surgical treatment of CPP. Radiotherapy, stereotactic radiosurgery, and chemotherapy are adjuvant therapies for CPP but there is no definitive protocol for the management of implants. Proper follow-up of these patients is essential since spinal drop can appear many years after the initial presentation of CPP.

**Keywords:** Choroid plexus papilloma, Metastasis, Spinal drop

## INTRODUCTION

Choroid plexus papillomas (CPPs) are extra-axial benign tumors that originate from the neuroepithelial lining of the choroid plexus. These tumors represent about 0.4–0.6% of all intracranial neoplasms and 2–4% of brain tumors in children and 1% of those in adults.<sup>[1,4,8,11]</sup> CPPs can be found in any choroid plexus, but the fourth ventricle is the most common location in adults. Diffuse craniospinal seeding involving the supra- and infratentorial compartments and spine has been observed.<sup>[8]</sup> Tumor metastasis or implants have been reported along cerebrospinal

This is an open-access article distributed under the terms of the Creative Commons Attribution-Non Commercial-Share Alike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

©2020 Published by Scientific Scholar on behalf of Surgical Neurology International

fluid (CSF) pathways; 35 such cases have been reported in the literature and of these, three exhibited atypical transformation of CPP spinal drop metastases.<sup>[8]</sup> Here, we describe the diagnosis and management of a patient with atypical CPP in the fourth ventricle with atypical spinal metastasis.

## CASE EXAMINATION

A 48-year-old male with a history of fourth ventricle atypical CPP diagnosed in 2008 underwent gross total resection (GTR) without any postoperative neurological deficits [Figure 1]. In 2015, he presented radicular pain, a slight decrease in strength, and paresthesia in the left leg.

The neurological examination showed decreased left thigh strength quantified as 4+ according to the Royal Medical Research Council of Great Britain scale, left patellar reflex quantified as 1+ according to the muscle stretch reflex (deep tendon reflex) grading scale, and hypoesthesia in the third and fourth lumbar dermatomes. Spinal magnetic resonance imaging revealed a well-defined intradural ovoid lesion in the vertebral canal at the level of the L3-L4 intersomatic space with a length of  $1.7 \times 1.2$  cm that was isointense on T1 and T2 sequences with homogeneous enhancement [Figure 2].

### Differential diagnosis, investigations, and treatment

The patient underwent microscopy-assisted tubular microsurgery with neural decompression through left



**Figure 1:** Brain magnetic resonance imaging. Axial brain contrast material-enhanced T1-weighted image. Within the fourth ventricle, there is a well-defined lesion, homogenous enhancement, and causing partial obliteration of the fourth ventricle.

L3 hemilaminectomy with subtotal resection (STR). Intraoperatively, we found a soft yellow-colored lesion that was firmly adhered to the fourth lumbar nerve root [Figure 3].

The final pathology report described a neoplasm with papillary architecture, with four mitoses in 10 high-power fields [Figure 4]. Immunohistochemistry showed positivity for cytokeratin 7, S100, and synaptophysin, with a proliferation index (Ki67+) < 1% [Figure 5]. In contrast to the pathology examination performed in 2008, here we observed a neoplasm with papillary architecture and several layers of epithelial cells that were ovoid and uniform, with no evidence of necrosis, pleomorphism, or anaplasia and a small mitotic fraction [Figure 6].

### Outcome and follow-up

Postoperatively, there was improvement of the radicular pain and an increase in left leg strength, with no changes in left leg dermal sensation. 2 months later, the patient was treated with radiotherapy (RT).

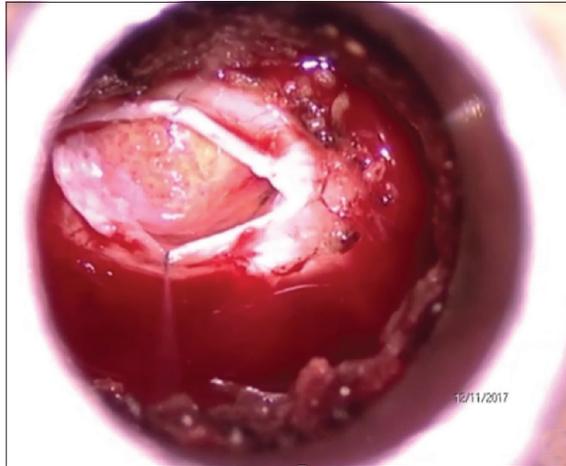
## DISCUSSION

### Background

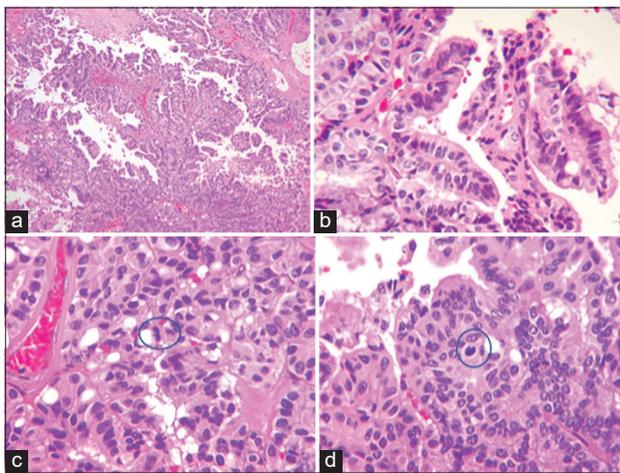
CPPs are rare central nervous system tumors that account for <1% of all intracranial neoplasms.<sup>[2]</sup> A review of the literature



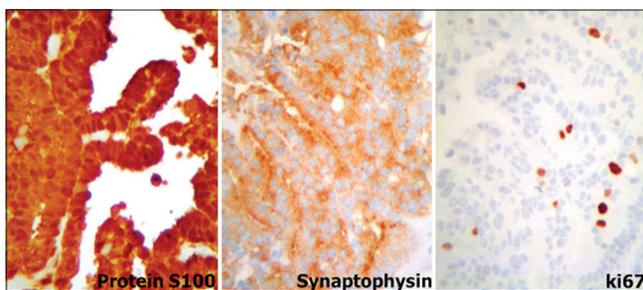
**Figure 2:** Spinal magnetic resonance imaging. (a) Sagittal T2-weighted image (b) sagittal lumbar spine contrast material-enhanced T1-weighted image (c) coronal T2-weighted image (d) axial lumbar spine contrast material-enhanced T1-weighted image. In the vertebral canal at the level of the intersomatic space, L3L4 presents a well-defined ovoid lesion, isointense on T2 sequence, with a length of  $1.7 \times 1.2$  cm, and intradural-extramedullary behavior that involves the spinal canal with homogeneous enhancement.



**Figure 3:** Surgical view. Soft yellow-colored lesion, firmly adhered to the nerve root of L4, with a length of 1.6 × 0.6 cm.

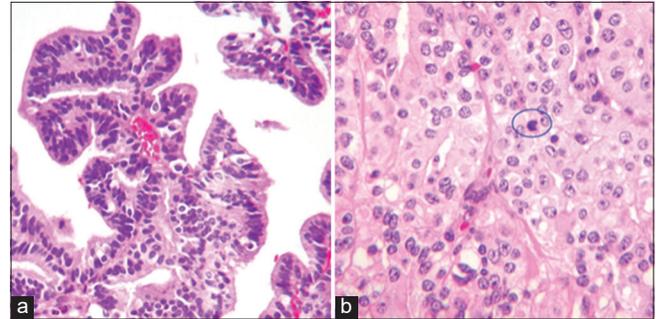


**Figure 4:** Photomicrographs of the spinal implant resected in 2017. A neoplasm with papillary architecture is observed, similar to that of 2008 (a and b) with solid areas (c) that present mitosis figures (c and d) of which four mitoses were counted in ten high-rise fields.



**Figure 5:** Immunohistochemical reactions of the spinal implant showing positivity for cytokeratin seven, positivity for S100 protein, synaptophysin, and with a proliferation index (Ki67) of <1%.

revealed 35 cases of CPP metastasis in adults, with a mean age at presentation of 40.3 years and a larger proportion of



**Figure 6:** Photomicrographs of atypical fourth ventricle tumor resected in 2008. A neoplasm with papillary architecture is observed with several layers of epithelial cells in some portions, they are ovoid and uniform (a) with a small amount of mitosis (b).

females affected (54%) than males.<sup>[8]</sup> The sites of metastasis or implants were the ventricular system, cranial fossa, and spinal axis.<sup>[1,2,4,7,8,11]</sup> Only three cases have been reported in which the CPP was at a spinal location (i.e., cervical, thoracic, and sacral).<sup>[8]</sup> We describe here a fifth-decade male patient with a lumbar neoplasm, which according to the histopathologic characteristics and location is the first case of an atypical papilloma implant of the choroid plexus at this spinal level.

### Clinical features

The clinical presentation of CPP depends on the site of metastasis. The principal clinical manifestation was radicular pain accompanied by paresis of the affected myotome.<sup>[8]</sup> The time between diagnosis of primary CPPs and distant spread varied from 1 month to 5 or more years.<sup>[1,8]</sup> Thus, for patients with a history of CPP and new-onset radicular pain spinal metastasis involvement must be considered, as in the present case.

### Treatment

The decision to perform surgery on this patient was based on the presentation of neurological deficit and pain. According to the previous reports, the indications for surgical intervention of spinal metastasis are intractable pain; the presence of a growing tumor resistant to RT, chemotherapy, or hormonal therapy; evidence of spinal instability, progressive deformity, or neurological deficit; and clinically significant neural compression.<sup>[5]</sup> The aim of spinal metastasis treatment is to relieve pain and preserve neurological function.<sup>[3]</sup>

Diverse scales have been proposed for evaluating the spinal metastasis treatment; the most popular are the Tokuhashi<sup>[12]</sup> and Tomita<sup>[13]</sup> scales which analyze the expected survival of patients. Based on the scores, clinicians can choose surgery or conservative treatment as the best approach for the patient.<sup>[5,12,13]</sup> According to both scales, the treatment of

choice for our patient was GTR as the survival prognosis was good.

In recent years, minimally invasive surgery (MIS) has become increasingly popular for the treatment of spinal pathology. This approach is associated with lower operative blood loss, diminished narcotic use, shorter postoperative stay, and lower costs of hospitalization compared to open surgery. Although the mortality associated with MIS is 0%, there are other complications including CSF leakage (3%), asymptomatic pseudomeningocele (3%), superficial surgical site infections (2%), sinus vein thrombosis (1%), and neurological deterioration (4%).<sup>[6,14]</sup> Intradural extramedullary tumors are candidates for MIS when the patient has a Karnofsky score >70% and normal (E) preoperative American Spinal Injury Association (ASIA) score, and craniocaudal tumor length less than 38 mm.<sup>[6,14]</sup> Our patient had a Karnofsky score of 90%, ASIA E score, and craniocaudal tumor length of 17 mm and was therefore an excellent candidate for MIS.

Neurological deterioration is expected in cases of GTR.<sup>[6,14]</sup> In our patient, total resection was not performed because the tumor was firmly attached to the nerve roots and the risk of neurological damage is greater with GTR.

### Histology

The world health organization (WHO) classifies ventricular papillary neoplasms into different grades: Grade I CPPs resemble normal papillary tissue, Grade II or atypical tumors are benign CPPs transforming to a malignant type, and Grade III or choroid plexus carcinoma (CPC) is characterized by anaplasia, increased mitotic activity, nuclear pleomorphism, and necrosis.<sup>[11]</sup> Approximately 80% of ventricular papillary neoplasms are CPPs; 15% are atypical, and < 5% are CPC.<sup>[11]</sup> Only three cases with metastatic lesions that progressed to atypical features have been reported.<sup>[8]</sup> The histopathologic findings from our patient suggested atypical CPP (WHO II) as an implant of a previous atypical CPP (WHO II) in the fourth ventricle.

### Prognostic

For CPPs, GTR is considered to be curative while STR produces good long-term results. Although GTR resulted in excellent tumor control at the 1- and 5-year follow-ups compared to STR, this advantage was lost after 10 years.<sup>[10]</sup> The utility of adjuvant therapy remains unclear. In cases of STR, RT is reserved for tumor progression documented by imaging; the radiation dose is between 25 and 54 Gy depending on the location of metastasis. GTR is the gold standard of treatment for metastatic CPP.<sup>[9,10]</sup> In this case, the first surgery was GTR of the atypical CPP in the fourth ventricle. In one study, progression-free survival after GTR of these tumors was found to be almost 95% and the patient did not receive adjuvant therapy.<sup>[9,10]</sup> For the spinal implant,

we performed STR and according to the previous studies the patient received adjuvant RT (25 sessions of 1.8 Gy, and for a total of 45 Gy).

Given the unpredictable behavior of CPPs, even with total resection, patients should be closely monitored during the post-surgical period as spinal implants have been reported 10 years after the primary tumor; in the present case, it was detected 7 years later. It is also important to consider the atypical location of CPP implants, which are very rare at the spinal level. In fact, ours is the first case reported at the lumbar level. CPP cases should be monitored for at least 10 years by routine cranial and spinal imaging, after which imaging should be performed in the event that there are root symptoms or the presence of spinal syndrome.

### CONCLUSION

CPPs are extra-axial benign tumors that account for < 1% of intracranial tumors in adults. The presence of spinal metastasis or implants is very rare; only three cases of spinal metastasis have been reported to date. An adequate follow-up of these patients is essential since implants can occur up to 10 years after the initial presentation of CPP.

The clinical manifestation of compression of nerve root or spinal syndrome in patients with a history of CPP precludes spinal metastasis, necessitating a complete study protocol as in the present case.

GTR of primary lesions and associated implants remains the gold standard for surgical treatment of CPP. There are no prophylactic treatments for preventing CPP metastasis. RT, stereotactic radiosurgery, and chemotherapy are adjuvant therapies for CPP but there is no definitive protocol for the management of implants.

### Acknowledgments

Primary authorship was the responsibility of D. Ochoa was responsible for all statistical aspects, analysis, editing, and writing. ME. Cordoba was responsible for data analysis, translation, preparation of figures, and verification. The team coordinating consisted of JR. Aguilar, U. García, A. Ibarra, VA. Reyes, JJ. Lomelí, MC. Sánchez, and RM. Vicuña. The group writing was O. Medina, MD. Sánchez, and EA. Castañeda.

### Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

### Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

## REFERENCES

1. Abdulkader MM, Mansour NH, van Gompel JJ, Bosh GA, Dropcho EJ, Bonnin JM, *et al.* Disseminated choroid plexus papillomas in adults: A case series and review of the literature. *J Clin Neurosci* 2016;32:148-54.
2. Akil H, Coupe NJ, Singh J. Spinal deposits of a benign choroid plexus papilloma. *J Clin Neurosci* 2008;15:708-12.
3. Babu ML, Gupta R, Kumar A, Gupta M. Spinal intradural metastasis. *JK Sci* 2004;6:48-50.
4. Bahar M, Hashem H, Tekautz T, Worley S, Tang A, de Blank P, *et al.* Choroid plexus tumors in adult and pediatric populations: The Cleveland Clinic and University Hospitals experience. *J Neurooncol* 2017;132:427-32.
5. Deng Z, Xu B, Jin J, Zhao J, Xu H. Strategies for management of spinal metastases: A comprehensive review. *Cancer Transl Med* 2015;1:94-100.
6. Halvorsen CM. Minimally invasive microsurgical resection of primary, intradural spinal tumors is feasible and safe: A consecutive series of 83 patients. *Neurosurgery* 2017;1:1-7.
7. Jinhu Y, Jianping D, Jun M, Hui S, Yepeng F. Metastasis of a histologically benign choroid plexus papilloma: Case report and review of the literature. *J Neurooncol* 2007;83:47-52.
8. Morshed RA, Lau D, Sun PP, Ostling LR. Spinal drop metastasis from a benign fourth ventricular choroid plexus papilloma in a pediatric patient: Case report. *J Neurosurg Pediatr* 2017;20:471-9.
9. Safaee M, Clark AJ, Bloch O, Oh MC, Singh A, Auguste KI. Surgical outcomes in choroid plexus papillomas: An institutional experience. *J Neurooncol* 2013;113:117-25.
10. Safaee M, Oh MC, Sughrue ME, Delance AR, Bloch O, Sun M, *et al.* The relative patient benefit of gross total resection in adult choroid plexus papillomas. *J Clin Neurosci* 2013;20:808-12.
11. Stuivenvolt M, Mandl E, Verheul J, Fleischeuer R, Tijssen CC. Atypical transformation in sacral drop metastasis from posterior fossa choroid plexus papilloma. *BMJ Case Rep* 2012;2-5.
12. Tokuhashi Y, Matsuzaki H, Oda H, Oshima M. A revised scoring system for preoperative evaluation of metastatic spine tumor prognosis. *Spine (Phila Pa 1976)* 2005;30:2186-91.
13. Tomita K, Kawahara N, Kobayashi T. Surgical strategy for spinal metastases. *Spine (Phila Pa 1976)* 2001;26:298-306.
14. Wong AP, Lall RR, Dahdaleh NS, Lawton CD, Smith ZA, Wong RH, *et al.* Comparison of open and minimally invasive surgery for intradural-extramedullary spine tumors. *Neurosurg Focus* 2015;39:E11.

**How to cite this article:** Ochoa-Cacique D, Cordoba-Mosqueda ME, Aguilar-Calderón JR, Sánchez-Silva MC, Vicuña-González RM, de la Torre AI, *et al.* Spinal drop of atypical choroid plexus papilloma in an adult patient: A case report and literature review. *Surg Neurol Int* 2020;11:409.