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Extraventricular choroidal plexus papilloma. Report of one case.

Authors**:** Angel J. Lacerda-Gallardo MD PhD[1]

Daisy Abreu- Perez MD ScM[2]

José Antonio Gálvez MD[1]

1- Department of Neurosurgery.

2- Pediatric Intensive Care Unit.

Correspondence: alacerda@infomed.sld.cu

Background:

Choroid plexus tumors are primary neoplasms of the central nervous system, of neuroectodermal origin. In children they have a special predilection for the lateral ventricles and the third ventricle, while in adults they are more frequently located in the fourth ventricle. The location of this tumor outside the ventricular cavity is rare butit can be located in brain parenchyma, in the brainstem, in the suprasellar region or even in the spinal canal. We are showing an ectopic location of a choroid plexus papilloma in a young female patient who underwent surgery in the Neurosurgery service of the General Teaching Hospital "Roberto Rodríguez" in Morón, Ciego de Ávila, Cuba.

Key Words: Choroid plexus papilloma, Brain tumors, Ectopic location.

Introduction:

The first description of a primary choroid plexus tumor was in a biopsy sample from a 3-year-old girl by Gerard, in 1832 (1). Choroid plexus tumors are primary neoplasms of the central nervous system, of neuroectodermal origin, representing between 0.3% and 3% of all intracranial tumors in children and less than 1% in adults (2,3). In children they have a special predilection for the lateral ventricles and the third ventricle, while in adults they are more frequently located in the fourth ventricle, and are rare in the lateral ventricles (2,4). Choroid plexus papillomas (CPP) can be located outside the ventricular cavity, in the brain parenchyma (5), in the brainstem, in the suprasellar region or even in the spinal canal (6), exceptionally. The objective of this study is to present an ectopic location of a choroid plexus papilloma in a young female patient who underwent surgery in the Neurosurgery service of the General Teaching Hospital "Roberto Rodríguez" in Morón, Ciego de Ávila, Cuba.

Case Presentation: A 31-year-old white female patient with a history of type I diabetes mellitus and hypertension, for which she was on insulin and enalapril treatment respectively.

The patient begins with a severe, oppressive holocranial headache that does not change with posture, that relieves little with analgesics, and is accompanied by a barely perceptible motor defect of the right half of the body. She is taken to the emergency department of the hospital, where she is evaluated by internal medicine and complementary tests are indicated.

Imaging: Simple cranium Computerized Tomography scan (CT scan): hyperdense, homogeneous image with well-defined edges is seen in the left

parieto-occipital region, with its postero-external border located 1.4 cm from the cerebral cortex and with diameters of 2.75 cm X 2.32 cm and its inner posterior border located 1.9 cm from the midline, surrounded by a hypodense area, with no mass effect on the midline structures. Contrast CT: The image described above captures contrast very slightly and homogeneously (Figure 1).

Abdominal ultrasound: Normal.

PA chest X-ray: Normal.

Treatment:

Medical: In the queuing period for allocation of surgical shift, treatment was applied with brain dehydrating agents of mannitol type 20% at a dose of 1g / kg / day and steroids such as dexamethasone 4mg every 8h, as well as symptomatic treatment.

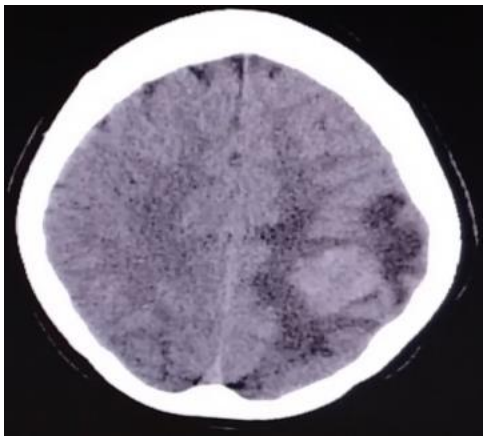

Figure 1. Preoperative contrasted CT scan.

Surgical: cytoreductive neurosurgical treatment was planned to be performed through a left parieto-occipital approach. The lesion was localized and dissection was performed through a brain parenchyma-tumor cleavage plane that was well-defined towards the surface and slightly less-defined towards the deep planes of the lesion, total resection of the lesion was achieved (Figures 2 and 3).

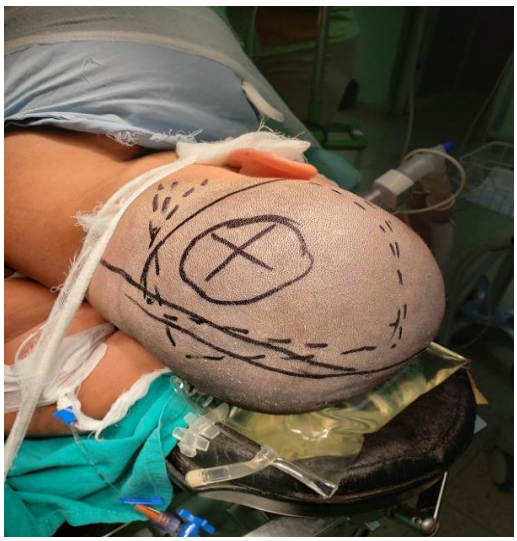

Figure 2. Intraoperative planning of the surgical approach.

Post-operative evolution: The evolution was satisfactory and patient was discharged 7 days after surgery.

Post-operative CT images: Post-operative CT images showed total resection of the lesion (Figure 4).

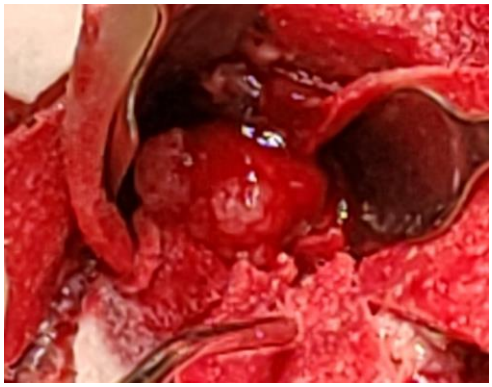

Figure 3. Transoperative view of the tumor dissection.

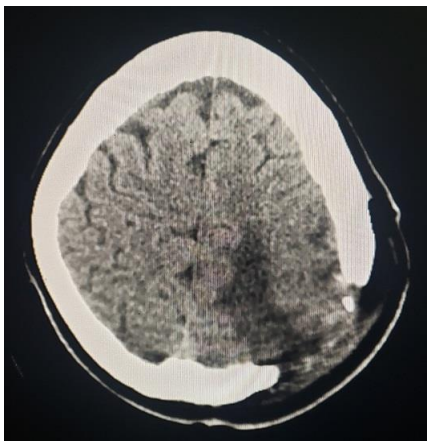

Figure 4. Post-operative CT image.

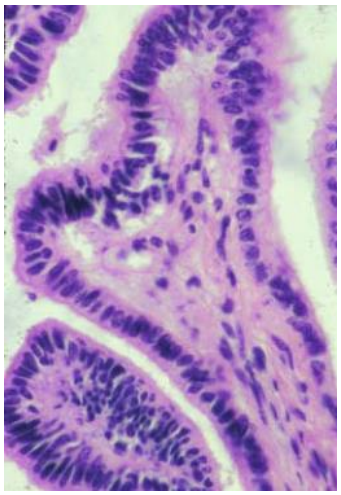

Figure 5. Histological features of the choroidal plexus papilloma. Hematoxylin and eosin tincture.

Biopsy: Choroid plexus papilloma (Figure 5).

Discussion and conclusion:

Primary choroid plexus papillomas are tumors that most frequently occur in pediatric ages and inside the ventricles. The extraventricular location is presented by two fundamental causes: they can develop from normal choroid plexus in the IV ventricle, which extend through the foramina of Luschka or Magendie and they appear in the cerebellopontine angle. Second by ectopic remains of choroid plexus in the parenchyma, which would give rise to the parenchymal presence of the tumor.

The first description of this type of tumor was made by Guerard in 1832 (1). According to its topography, there are, in addition to purely intraventricular locations, intraventricular-extraventricular forms (6) and purely extraventricular tumors, including parenchymal (5-8), cistern and intraparenchymal presentations (9), intra-spinal presence (6), in the cerebellum (10), and sellar and suprasellar region (11) has also been described.

The World Health Organization classifies choroid plexus tumors into three histological grades: Grade I is choroid plexus papilloma (CPP), grade II is atypical papilloma (aCPP) and grade III is choroid plexus carcinoma (CPC). (7). Total surgical resection is the main objective in grades I and II, and adjuvant treatment is reserved primarily for grade III. The prognosis not only depends on its histological grade but also on its size, location and association with alterations related to the free circulation of the cerebrospinal fluid (CSF) (12).

As they are intraventricular tumors, their clinical presentation is closely related to the manifestations of intracranial hypertension that appear with hydrocephalus that is very frequently associated with these histological types, secondary to obstruction of free circulation or hyperproduction of CSF (1,5), however they can also present with seizures, hemorrhages and focalization signs (1).

Unlike typical lesions, parenchymal extraventricular CPP are little associated with hydrocephalus (5), as happened in the case reported, it is difficult to distinguish from other types of tumors, that`s why the biopsy represents the most accurate form of diagnosis.

These lesions tend to behave isodense or slightly hyperdense on CT scan, with homogeneous contrast uptake. On magnetic resonance images (MRI), they are usually isointense or slightly hypointense on T1 and hyperintense on T2 (1), as occurred in this case.

The main objective of surgical treatment is total resection, and survival rate has been linked closely with the degree of resection achieved during surgery, with the histological grade of the lesion, with tumor progression and the presence or absence of leptomeningeal metastasis (12). In our patient we achieved total resection.

In addition to surgical treatment, adjuvant treatment in these cases represents an important aspect, the use of chemotherapy and radiotherapy have been recommended especially with choroid plexus carcinoma, but the existing evidence on its benefit is poor due to the small number of cases in the different series reported in international literature (12).

Conclusion:

We can conclude that the ectopic location in the brain parenchyma is a rare location for choroid plexus papillomas and that its diagnosis will most often be through the histological study of the sample obtaining from the specimen.

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Abbreviations

CPP: Choroid plexus papilloma

aCPP: Atypic Choroid plexus papilloma

CPC: Choroid plexus carcinoma

CT: Computarized Tomography.

CSF: Cerebrospinal fluid.

MRI: Magnetic Resonance Images.

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Ethics approval and consent to participate: This case report was approved by the Institutional Review Board of "Roberto Rodriguez" General Teaching Hospital.

Consent for publication: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.