

Choroid plexus papilloma, a cause of hydrocephalus in children. A case report

Papiloma de plexos coroideos, tumor infrecuente en la edad pediátrica. A propósito de un caso

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Summary
Choroid plexus papilloma, an uncommon benign tumor of the central nervous system, represents less than 5% of all brain tumors in the pediatric age group. Its peak incidence occurs in the first year of life, with sporadic cases in adults. This neoplasm causes hyperproduction of cerebrospinal fluid caused by a disruption in its reabsorption mechanism, leading to hydrocephalus, which is characteristic in these patients. Imaging plays a crucial role in the diagnosis, determined by its typical appearance, intraventricular, predominantly supratentorial in the lateral ventricles, and its cauliflower-like morphology, which shows marked enhancement with contrast medium.

Resumen
El papiloma de plexos coroideos, tumor benigno infrecuente del sistema nervioso central, representa menos del 5 % de todos los tumores cerebrales en la edad pediátrica, su pico de incidencia es en el primer año de vida, con casos esporádicos en los adultos. Esta neoplasia ocasiona hiperproducción de líquido cefalorraquídeo provocada por una alteración en su mecanismo de reabsorción, que causa hidrocefalia, característica en este tipo de pacientes. La imagenología cumple un rol fundamental en su diagnóstico, determinado por su apariencia típica en forma de coliflor, de localización intraventricular, de predominio supratentorial en los ventrículos laterales con realce marcado al medio de contraste.

Introduction
Choroid plexus papilloma is a rare brain tumor located within the ventricles. Most cases have been documented in patients around one year of age, and it has become the cause of hydrocephalus in this age group. Imaging plays a fundamental role in its diagnosis due to its characteristic findings; however, it is indistinguishable from its malignant variant.

The following is a case detected at the institution where the authors work.

Case
The case of a 7-month-old infant was presented, with a personal medical history of Chiari II malformation (cerebellar tonsillar descent associated with lumbosacral myelomeningocele) and hydrocephalus; the myelomeningocele was surgically corrected in the neonatal period and the hydrocephalus required the placement of a ventriculoperitoneal shunt. He was admitted to the health center due to decompensation of his underlying condition. Physical examination revealed a globular abdomen, positive ascites wave, marked increase in head circumference, and bulging fontanelles.

The following tests were performed: abdominal ultrasound, which revealed 2,500 mL of free fluid in the abdominal cavity; a simple CT scan of the skull, which showed significant dilation of the supratentorial ventricular system, resulting in overall thinning of the cerebral parenchyma; an Evans index of 0.52 (Figure 1), also associated with a heterogeneous, lobulated lesion with internal calcifications, located in the left lateral ventricle, with a cauliflower-like morphology (Figure 2).

A complementary study with contrast medium revealed marked homogeneous enhancement of the entire lesion (Figure 3).

The lesion was excised and the ventriculoperitoneal shunt (VPS) was replaced with an external ventricular drain (EVD). Histopathological examination confirmed the diagnosis of choroid plexus papilloma.

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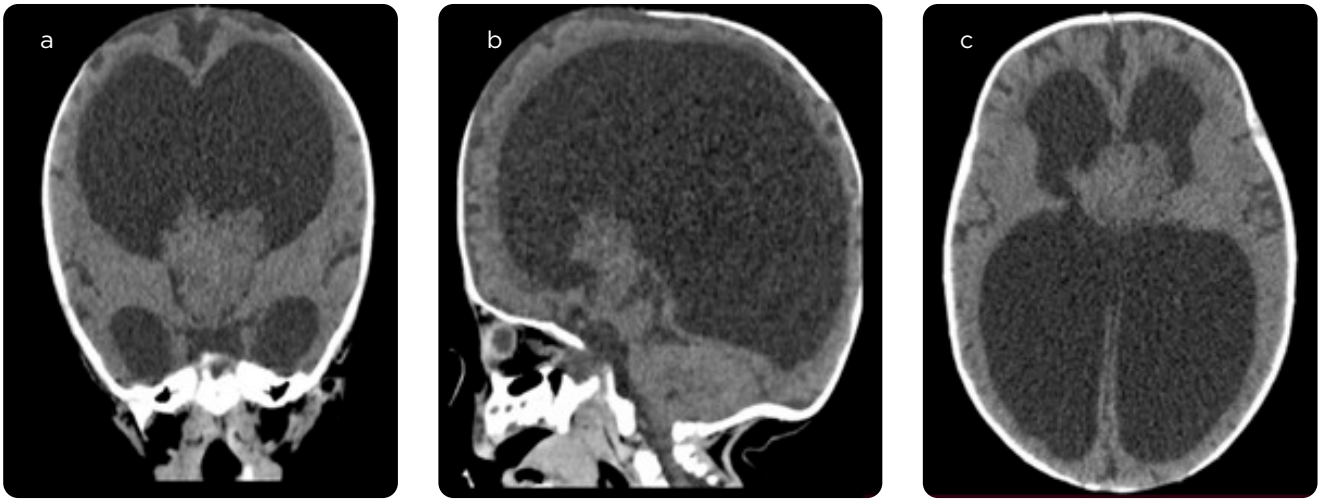


Figure 1. Plain CT scan of the skull. a) Coronal; b) sagittal; c) axial. Severe hydrocephalus with lobulated intraventricular lesion associated with cerebellar tonsillar herniation, Chiari malformation.

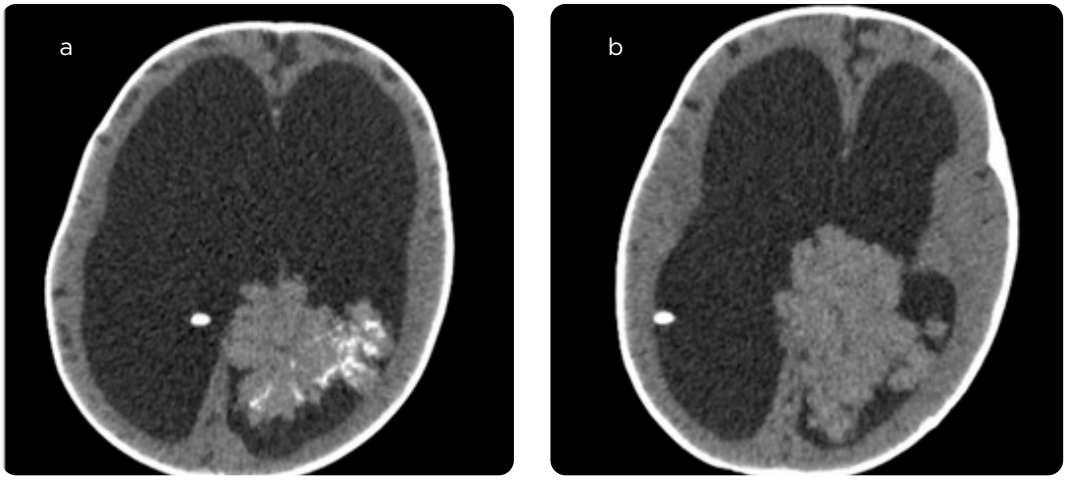


Figure 2. Plain CT scan of the skull. a) Axial. Calcifications in the lesion. b) The lesion is lobulated with a "cauliflower" morphology. Ventricular catheter entering the lateral wall of the posterior region of the right lateral ventricle.

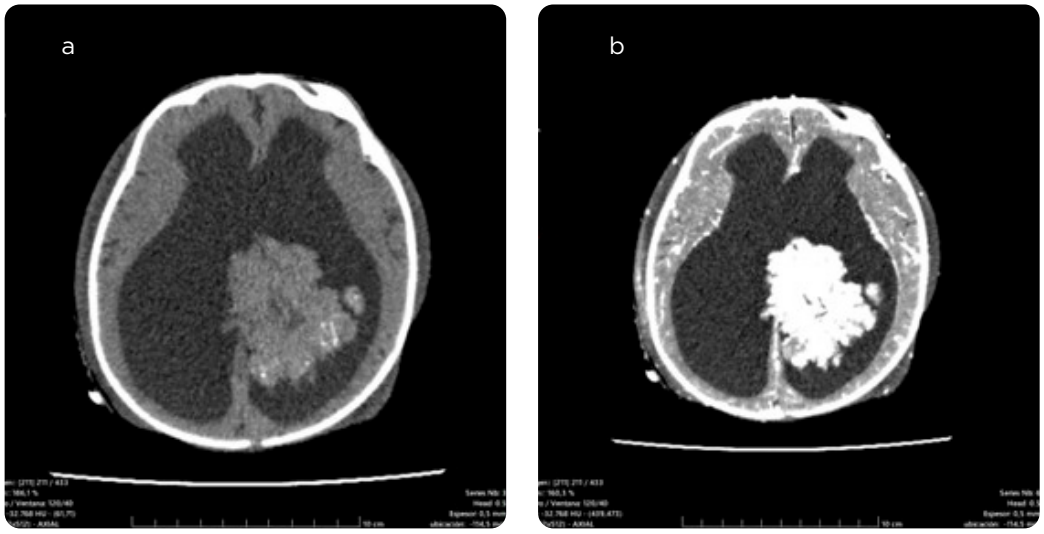


Figure 3. a) Plain CT scan of the skull, axial: lobulated lesion in the ventricular atrium. b) With contrast medium: marked and homogeneous enhancement of the lesion.

Discussion

The choroid plexuses are formed from the neuroectoderm and are located throughout the ventricular system. Their main function is the production of cerebrospinal fluid (CSF) (1). Pathology can occur in any area of the cerebral ventricular system (1, 2).

Choroid plexus papilloma or papillary neoplasia is a rare benign intraventricular tumor that occurs most frequently in children, although it accounts for less than 5% of brain tumors in this age group and has a higher incidence in the first year of life (2); however, isolated cases have been reported in adults (3).

In the ventricular system, it is most commonly located in the atrium of the lateral ventricles, followed by the fourth and third ventricles. To a lesser extent, it can be located extraventricularly, such as in the pontocerebellar angle, suprasellar region, frontal lobe, posterior commissure, pineal gland, and posterior fossa (4,5). In children, it is more commonly found in the ventricular atrium (4, 5), while in adults it is most frequently located in the fourth ventricle (2). In less than 5% of cases, it may have multiple locations (5).

Histologically, choroid plexus papilloma has been classified into three grades.

- Choroid plexus papilloma (WHO grade I).
- Atypical choroid plexus papilloma (WHO grade II).
- Choroid plexus carcinoma (WHO grade III), a malignant variant (6).

No predilection for either sex has been described; however, in adults, more cases have been reported in males (5).

Its association with Chiari II malformation, which occurs in the patient presented, has not been documented in the literature; however, this is an aggravating factor in the management of the patient, as it is a difficult-to-control hydrocephalus, since all the pathways of hydrocephalus pathogenesis are affected: excess CSF secretion, CSF absorption disorder caused by the papilloma, and obstruction of the circulation pathways by cerebellar descent in Chiari II malformation (6).

The clinical presentation corresponds to the symptoms and signs of hydrocephalus (2-4), which is determined by the three mechanisms mentioned above: CSF overproduction, obstruction of drainage secondary to the tumor, and impaired CSF absorption (5).

On CT scan, it presents as a solid, low-density, lobulated intraventricular lesion with a "cauliflower" morphology, associated with cystic areas, hemorrhage, internal calcifications, and homogeneous and marked enhancement with contrast medium administration; the latter being its main difference from its malignant variant, carcinoma, which usually presents with irregular margins, is infiltrative, and has heterogeneous enhancement (2, 5).

On magnetic resonance imaging (MRI), it appears as a solid, well-defined, cauliflower-like lesion with medium to low signal intensity on T1-weighted sequences and intermediate signal intensity on T2-weighted sequences, with prominent enhancement on contrast-enhanced sequences, with or without hemorrhage and calcifications. It does not restrict diffusion sequences, and spectroscopy shows a decrease in N-acetyl aspartate (NAA) and a peak in choline (Cho) (4, 5).

On ultrasound, it appears as a lesion with circumscribed, regular, echogenic, homogeneous, and lobulated margins, with increased vascularity on color Doppler (5).

The treatment of choice for choroid plexus papilloma is total excision of the tumor, in the case of the patient presented here, associated with correction of his concomitant Chiari malformation to correct hydrocephalus.

Conclusion

Choroid plexus papilloma has its own characteristics, being intraventricular, lobulated, with a cauliflower-like morphology, with or without concomitant hemorrhages or calcifications; its avid and homogeneous enhancement with contrast medium makes it easily identifiable. It is associated with hydrocephalus.

Its relationship with Chiari malformation has not been documented, and there are no data in the literature on this relationship; however, this denotes the difficult management of hydrocephalus in this patient, as all the mechanisms of its pathogenesis are involved, including hypersecretion, reabsorption disorders, and obstruction, which in the patient described required immediate valve replacement.

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