Title : Choroid plexus papilloma : Case report .

Abstract :

Choroid plexus papilloma (CPP) is a benign intraventricular neuroepithelial tumor classified as WHO Grade 1. Despite being rare, it happens in both adult and pediatric populations, albeit it is more common in children. . Clinical symptoms, molecular and genetic markers, imaging characteristics, and most importantly histological investigation are the four primary methods used to diagnose CPP. We report the case of a 53-year-old patient admitted for signs of intracranial hypertension (isolated headache) with standing and walking disturbances and heaviness of the left hemibody diagnosed with choroid plexus papilloma through histological exam.

Keys words: choroid plexus papilloma , intraventrucular masse, benign tumor, papillary architecture.

Introduction ;

Choroid plexus tumors are papillary intraventricular masses that arise from the choroid plexus[1]. They are more frequently seen in children than in adults and make up between 0.3% and 0.6% of brain tumors[2].

According to the World Health Organization (WHO), these tumors are classified into three categories based on histological characteristics:

1. **Grade I: Choroid Plexus Papilloma (CPP)** – A slow-growing, benign tumor.
2. **Grade II: Atypical Choroid Plexus Papilloma (aCPP)** – An intermediate-grade tumor with a higher risk of recurrence.
3. **Grade III: Choroid Plexus Carcinoma (CPC)** – A malignant and aggressive neoplasm of the choroid plexus.

This classification system helps guide diagnosis and treatment strategies based on the tumor's behavior and associated risks [3]. Herein, we present the case of a 53-year-old patient diagnosed choroid plexus papilloma .

Case report:

A 53-year-old patient operated on in 2016 for VP shunt + choroid plexus papilloma of the fourth ventricle (V4), admitted for signs of intracranial hypertension (isolated headache) with standing and walking disturbances and heaviness of the left hemibody evolving for 1 month. Clinical examination revealed:Statokinetic syndrome ,neurological focalization sign: left hemiparesis rated 4/5. Routine hematological tests were normal.Brain MRI

revealed a multilobulated fourth ventricular mass (arrow), and display hyperintense signal on T2WI/FLAIR and low ADC value. This mass caused obstruction leading to hydrocephalus (figure 1).

Microscopic findings revealed a benign tumor proliferation with a papillary architecture, lined by cells with regular nuclei and moderately abundant eosinophilic cytoplasm. The core of the papillae is thin and fibrovascular (figure 2).

The immunohistochemical study shows diffuse positivity for anti-PS100 antibodies, focal positivity for anti-synaptophysin antibodies, weak and focal expression of anti-GFAP antibodies, negativity for anti-EMA antibodies, and a Ki67 proliferation index estimated at 2%, suggesting a benign tumor with neuroectodermal differentiation(figure 3).

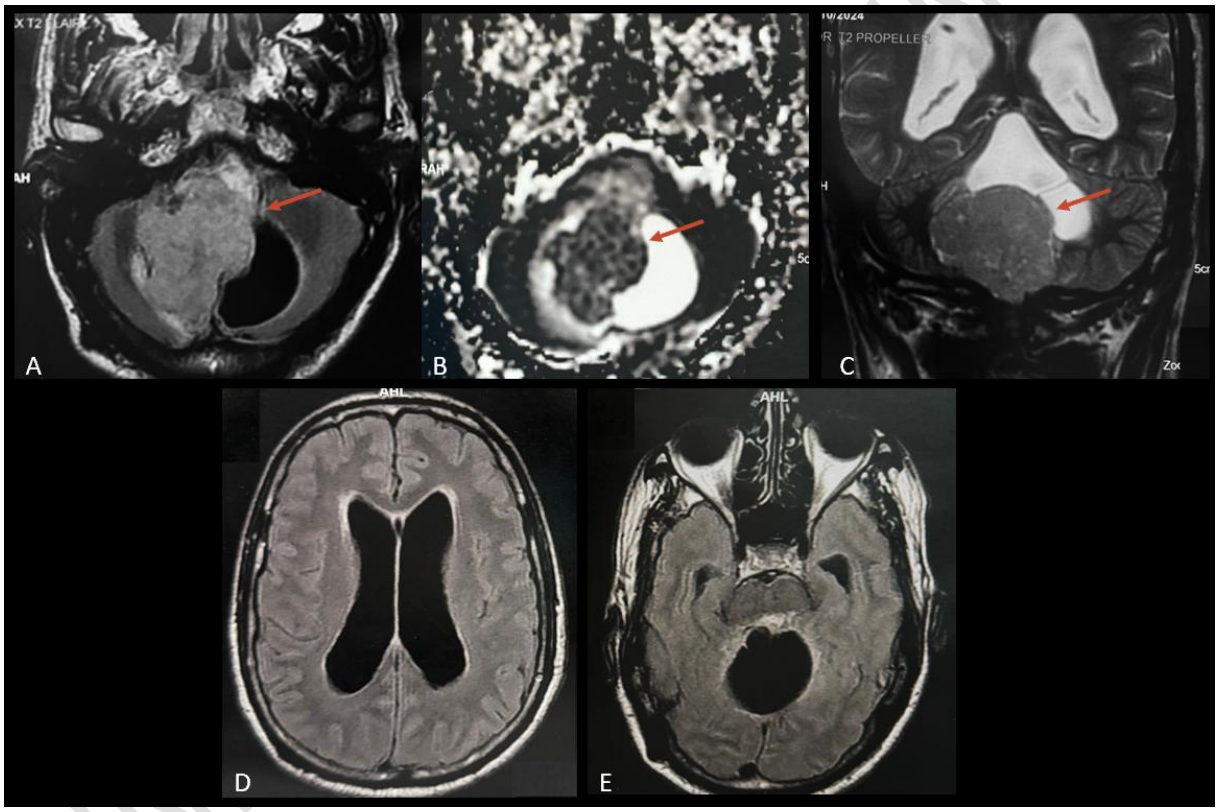

Figure 1 : Brain MRI in axial T2 FLAIR (A), axial ADC map (B) and coronal T2WI (C) sequences demonstrated a multilobulated fourth ventricular mass (arrow), and display hyperintense signal on T2WI/FLAIR and low ADC value. This mass caused obstruction leading to hydrocephalus (D, E).

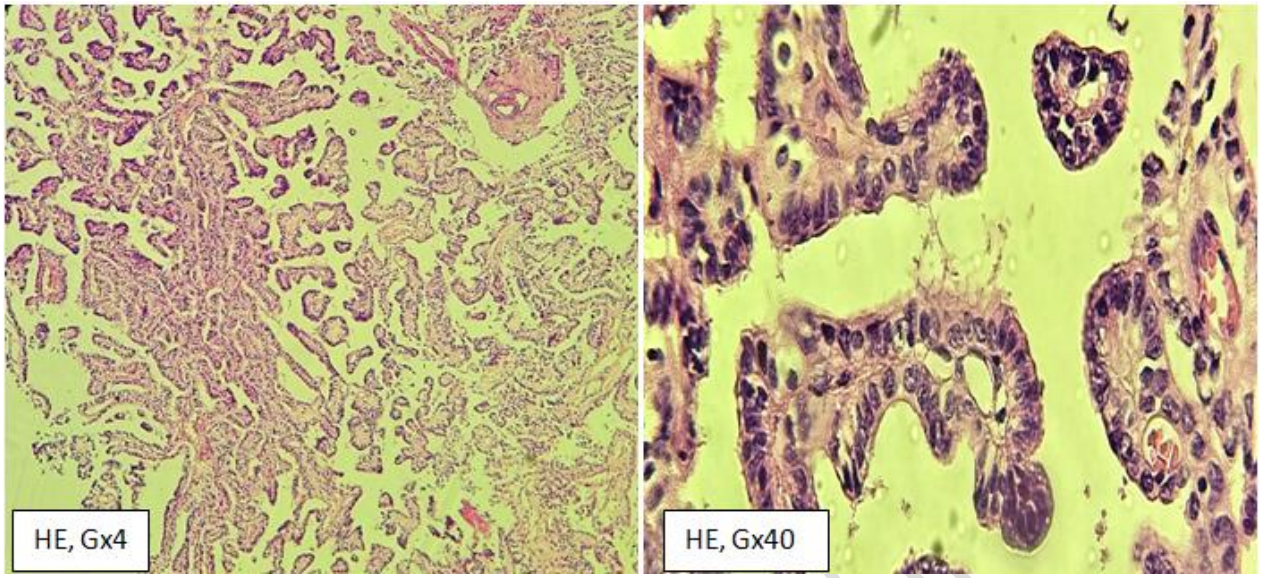

Figure 2:Anatomopathological examination reveals a papillomatous proliferation consisting of papillae of variable size(Image on the left). Papillae are lined with monomorphic cells (image on the right).

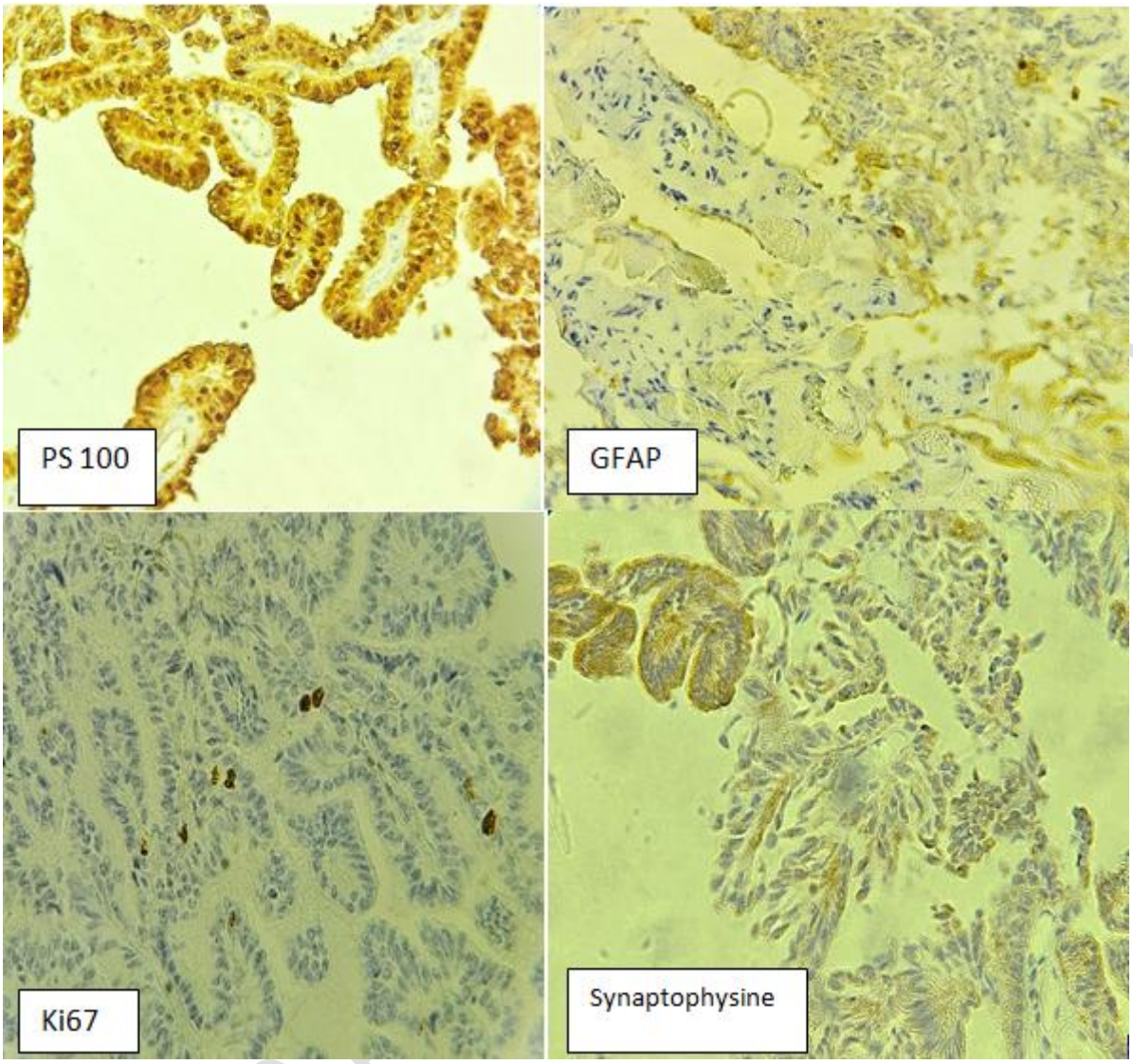

Figure 3 : Immunohistochemical findings : malignant cells were positive for anti-Ps100,anti-synaptophysine,anti-GFAP with Ki67 proliferation index estimated at 2%.

Discussion:

The choroid plexus is a highly vascularized structure in the central nervous system (CNS), composed of fenestrated and discontinuous capillaries. Its cells are tightly connected by junctions and are lined with ependymal cells within the brain's ventricles. This unique structure can be affected by various pathological conditions [4].

Choroid plexus papillomas (CPPs) are rare neuroectodermal tumors that originate from the choroid plexus epithelium. Representing 0.4% to 0.6% of all brain tumors, they are associated with a favorable long-term prognosis. CPPs can occur at any age, commonly

appearing in the lateral ventricles (supratentorial) in children and the fourth ventricle (infratentorial) in adults[5].

The symptoms of CPPs depend on their size and location. Most symptoms are caused by increased intracranial pressure due to blockage of cerebrospinal fluid (CSF) flow, and less frequently, by the mass effect of the tumor. Common clinical presentations include cranial nerve III and VI palsies, visual disturbances, headaches, nausea, balance problems, and drowsiness [6-7].

On MRI scans, choroid plexus papillomas (CPPs) typically appear as lobulated, homogeneous masses with uniform enhancement, while choroid plexus carcinomas (CPCs) tend to be more heterogeneous due to the presence of necrosis, calcification, or hemorrhage [8].

Angiography highlights the highly vascular nature of CPPs, showing an intense vascular blush. The tumor may also display enlarged choroidal arteries supplying blood to it, along with potential shunting[9]. Common angiographic features include small spiral arteries, a meningioma-like blush with early tumor circulation, displacement of vessels such as the internal cerebral veins, and signs of ventricular dilatation[10].

A range of imaging techniques, including computed tomography (CT), magnetic resonance imaging (MRI), and angiography, play a crucial role in diagnosing choroid plexus papillomas (CPP). However, these imaging methods alone are not enough to conclusively confirm the diagnosis of CPP. To achieve a definitive diagnosis and ensure effective treatment, a complete surgical excision of the tumor followed by histological examination is necessary[11].

The WHO classification of choroid plexus tumors is based on their histological features. Choroid plexus papillomas (CPP) are distinguished from other choroid plexus tumors by their low mitotic activity (<2 mitoses per 10 high-power fields) and unique histological characteristics[12].

Compared to normal choroid plexus tissue, the cuboidal to columnar epithelial cells that border the fibrovascular papillary projections in well-differentiated CPPs are more dense, elongated, and stratified. The cytologically bland epithelial cells found in CPPs often have fine chromatin and moderately eosinophilic or transparent cytoplasm. In contrast to more aggressive choroid plexus tumors, CPPs may not exhibit malignant traits such nuclear pleomorphism, necrosis, or brain invasion, while they may occasionally exhibit oncocytic alterations and degenerative features like calcification, hyalinization, or vacuolization[13].

Immunohistochemical analysis shows that CPP shows express cytokeratin, vimentin, and S-100. The lack of Epithelial Membrane Antigen (EMA) and Glial Fibrillary Acidic Protein (GFAP) supports the diagnosis of CPP, although GFAP can occasionally appear focally. In

atypical CPP (aCPP), immunohistochemistry highlights positivity for vimentin, synuclein, and S-100[14].

Genetic and molecular studies on CPPs are complicated due to the rarity of the tumor and the complexity of its biological processes. Many studies have small sample sizes, which limits the ability to make definitive conclusions. This presents challenges when developing therapeutic strategies or identifying genetic markers. Nevertheless, despite these difficulties, recent research has drawn attention to the potential role of molecular biology and genetic analysis in improving the diagnosis of CPP, offering hope for better differentiation from other choroid plexus tumors[15].

A total surgical excision is usually curative, with few cases of recurrence[16]. However, complete removal can sometimes be difficult [17].

Conclusion:

In conclusion, while choroid plexus papilloma is uncommon, it demands timely and comprehensive multidisciplinary management. Raising awareness among healthcare professionals about its symptoms and ensuring early recognition are crucial for optimizing patient outcomes and enhancing quality of life.

Authors contribution

All authors participated actively in elaboration of this scientific Document.

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

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