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Case Report

Comprehensive Review of Fourth Ventricle Epidermoid Cysts

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ABSTRACT:
Epidermoid cysts located within the fourth ventricle are exceptionally uncommon intracranial anomalies arising from epithelial remnants during neural tube closure. Constituting a minority of intracranial neoplasms, these benign extracerebral intradural lesions account for less than 2% of all intracranial tumors, with a mere 5 to 18% manifesting within the confines of the fourth ventricle. Given their rarity, this report endeavors to provide a comprehensive delineation of a fourth ventricle epidermoid cyst, elucidating its clinical, radiological, therapeutic, and prognostic attributes within the context of a 32-year-old male patient.

keywords: Epidermoid cyst, Intracranial lesion, Papilledema, Cysticlesion, Telovelar approach ,Surgical excision, Ventriculoperitoneal shunt, Keratin Proteins

INTRODUCTION:

Epidermoid cysts are rare and benign slow-growing tumors of ectodermal origin, representing 0.2 to 1.8% of all intracranial tumors.[11] Due to their pearly appearance, in 1829, Cruveilhier coined these tumors as "Tumeur Perlée." Virchow and Bailey introduced the term "pearly tumor"; and in 1836, Müller introduced the designation "Cholesteatoma."[2] They typically occur throughout the neuraxis, most commonly in the cerebellopontine angle (60%), but can also appear in other organs, such as the middle ear or orbit [11]. Their occurrence in the fourth ventricle is unusual, accounting for only 5 to 18.5% of all epidermoid cysts appearing in the brain [1], [10], [12]. Only 100 cases have been described in the literature.[3,4] They result from ectodermal inclusion during the closure of the neural tube during the 3rd to 5th week of embryogenesis and have an extremely slow linear growth due to their avascular nature and composition with cholesterol in a solid crystalline state and keratin in the tumor. Epidermoid tumors typically have long T1 and T2 relaxation times and show no enhancement on post-contrast MRI images. Microscopically total excision is the only guarantee of optimal patient recovery, with minimal risk of recurrence.

Case Report:

This study concerns a 32-year-old man complaining of occipital and pulsatile headaches approximately eight months before his admission. Not responding to analgesic use, associated with nausea, dizziness, and vomiting. After four months, he began to develop gait disturbances. Radiological exploration conducted by his family physician found an intracranial lesion, and he was referred to our service for surgical intervention. Clinical examination revealed a conscious, cooperative patient complaining of headaches. Fundoscopy revealed bilateral stage I papilledema and a state-kinetic cerebellar syndrome with ataxia.

Magnetic resonance imaging (MRI) revealed a large extra-axial cystic lesion filling the fourth ventricle, expanding into the cisterna magna with invasion of the right Luschka foramen, displacing the cerebellar parenchyma upwards, and compressing the medulla oblongata. On T1-weighted images, the lesion appeared hypointense and hyperintense on T2-weighted images with irregular contours.(figure1)

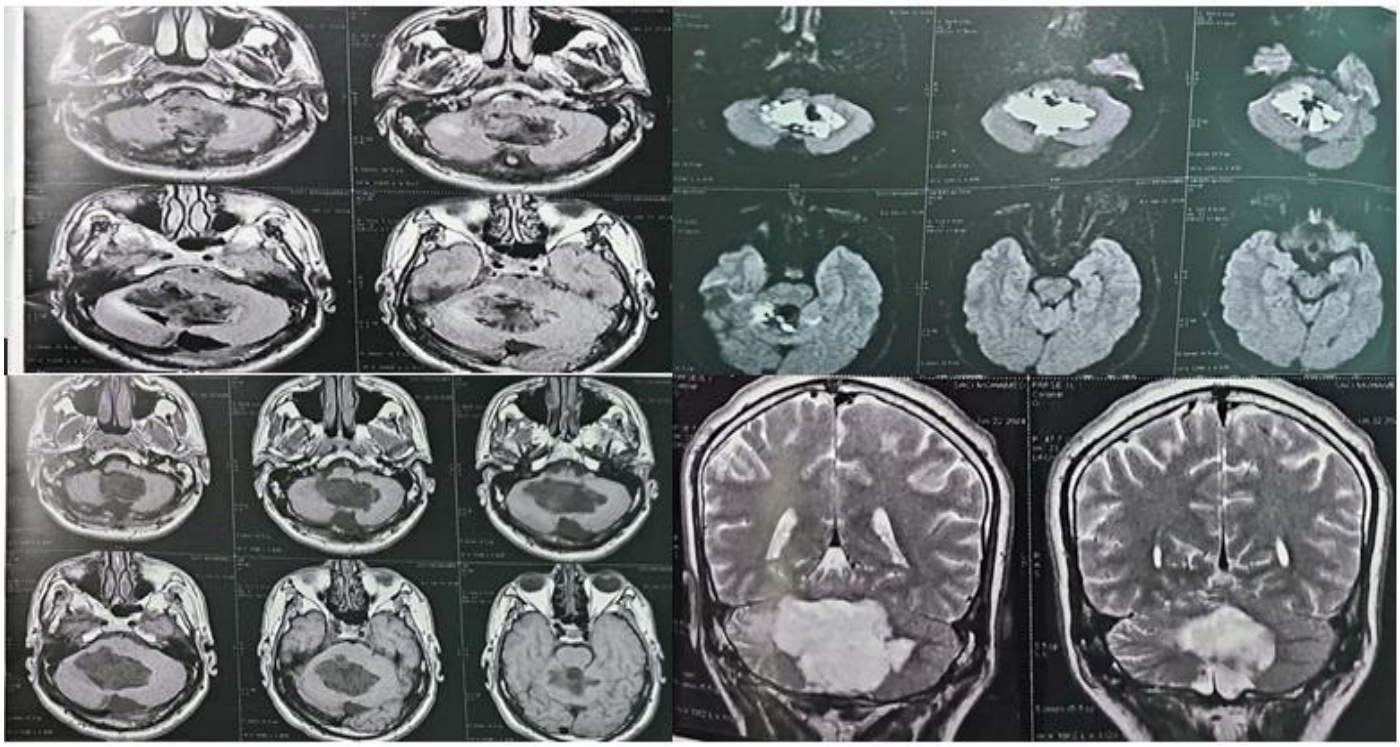

Figure 1: Preoperative MRI Showing a Cystic Lesion of the Fourth Ventricle: Axial T1-weighted MRI Sequence, Demonstrating a Lesion Isointense to Cerebrospinal Fluid; Coronal T2-weighted MRI Sequence, Depicting a Hyperintense Lesion; Axial Diffusion-Weighted Imaging (DWI), Displaying a Hyperintense Lesion.

There was restriction on diffusion-weighted imaging, and the lesion did not enhance with gadolinium administration. There was heterogeneity on FLAIR sequence and hyperintensity on diffusion-weighted imaging. There was no ventricular dilation or tonsillar herniation. The MRI was suggestive of a typical intraventricular epidermoid cyst, and the decision to operate on the lesion was made. For the surgical procedure, the patient was positioned prone. We performed a midline occipital craniotomy to approach the lesion through a unilateral telovelar approach.(figure2)

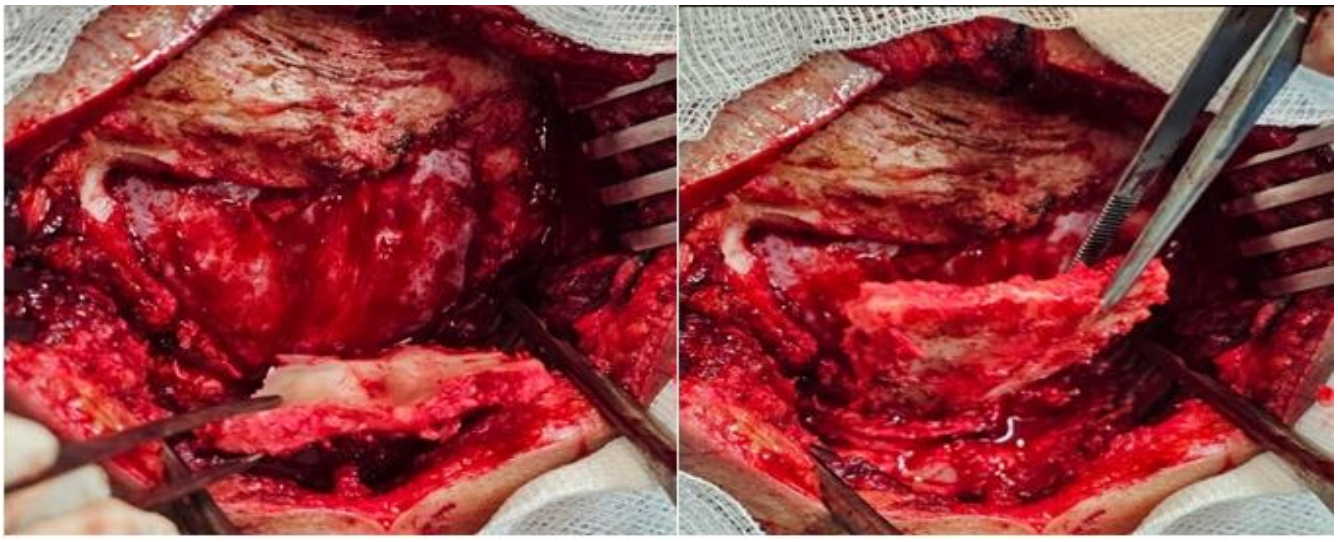

Figure 2: Median Suboccipital Approach, Bone Flap.

We visualized a pearly appearance of a whitish tumor, resembling the characteristic appearance of an epidermoid cyst with cisternal extension.(figure03)

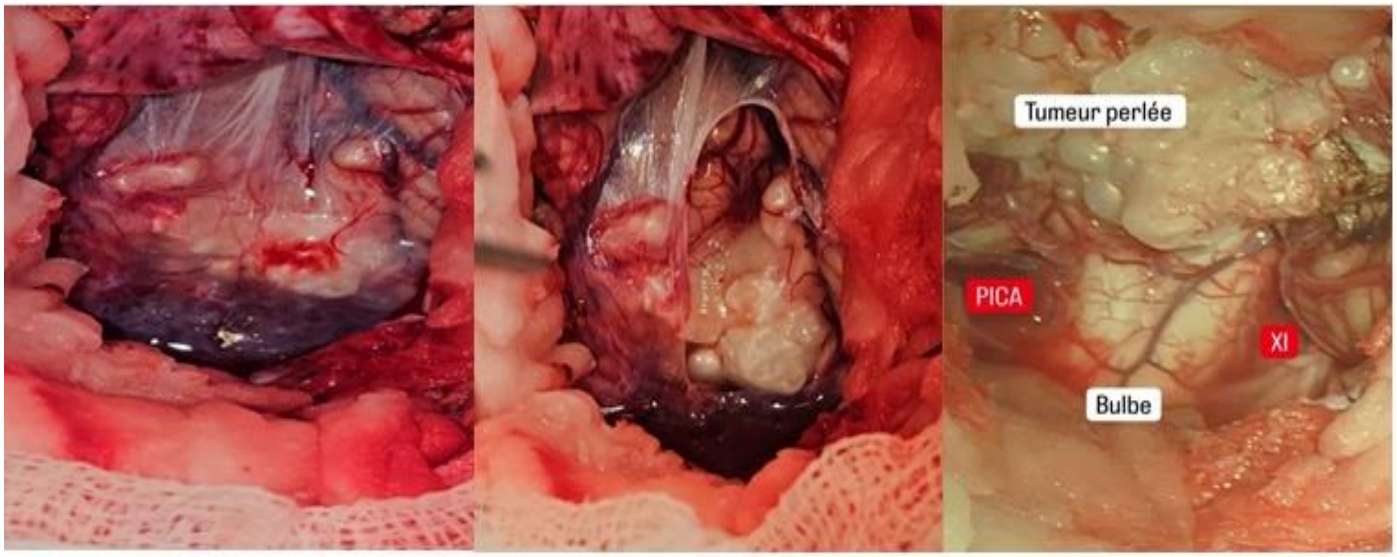

Figure 3: Intraoperative Appearance of the Cyst with Identification of a Pearly Mass on the Fourth Ventricle Topography Before and After Opening the Cisterna Magna, Identifying its Relationship with the XI Cranial Nerve, the Posterior Inferior Cerebellar Artery (PICA), and the Bulb.

Despite the difficulty encountered due to the intimate adhesion of the tumor capsule to the lower part of the floor of the fourth ventricle, the telovelar approach allowed us to achieve total surgical excision, visualizing the various recesses of the fourth ventricle, namely the two lateral recesses, the Sylvian aqueduct, and the Obex.(figure4/5)

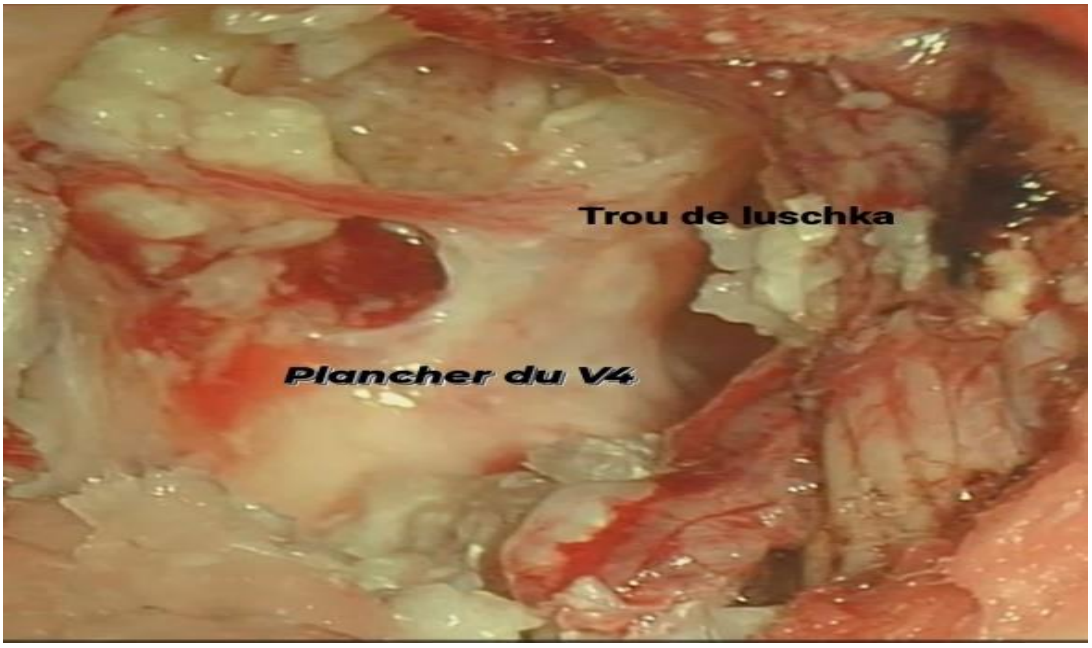

Figure 4: Unilateral Telovelar Approach

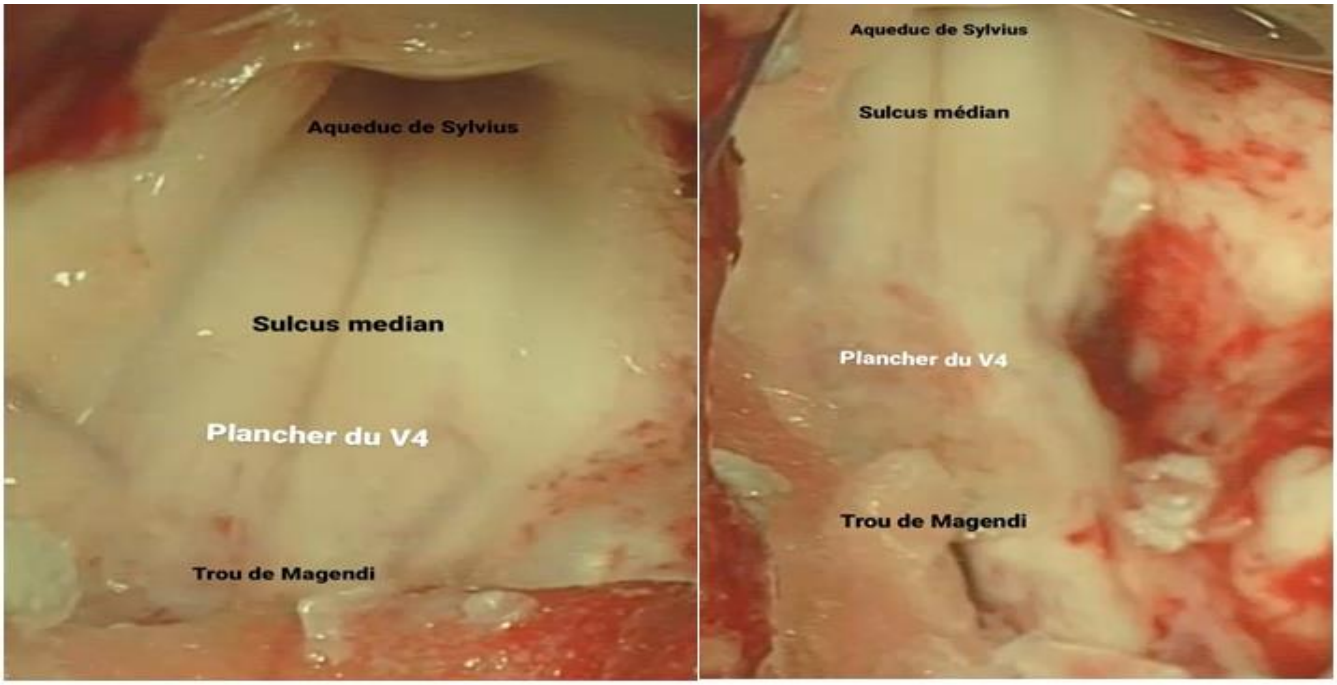

Figure 5: Completion of Resection and Complete Visualization of the Floor of the Fourth Ventricle

A ventriculoperitoneal shunt was not necessary due to the absence of ventricular dilation. In the immediate postoperative period, there were no complications, and the patient clinically improved during a six-month follow-up. A brain CT scan was performed, demonstrating the emptiness of the fourth ventricle.(figure6).Histopathological study showed a capsule tissue rich in keratin and proteins and predominantly cystic content, consistent with an epidermoid cyst.

Figure 6: Follow-up CT Scan Showing the Emptying of the Fourth Ventricle

DISCUSSION:

Epidermoid cysts are benign, congenital, and rare, accounting for approximately 0.2 to 1.8% of all intracranial tumors.[1] Formerly called cholesteatoma or Cruveilhier's pearly tumor. The usual location is found in the cerebellopontine angle (40 to 50%); or in the parasellar and temporal regions (30%).[1] Its localization in the fourth ventricle is very rare (5%), with only 100 cases described in the literature.[3,4] These tumors are generally asymptomatic until the fourth or fifth decade of life, with a mean age of 40 years. Clinically, cerebellar syndrome is the most common manifestation, while intracranial hypertension syndrome is less frequent, as hydrocephalus occurs late and is observed in less than 50% of cases in accordance with our case.[1] Similarly, the very slow growth of the tumor and the likely persistence of cerebrospinal fluid flow space between the capsule and the ventricle walls explain the lack of correlation between the size of the tumor volume and the presence of hydrocephalus at the time of tumor discovery.[15] Extension into the cerebellopontine angle through the foramina of Luschka leads to cranial nerve lesions (mixed nerves, acoustic-facial bundle, trigeminal nerve).[3,7] In our case, a 32-year-old patient whose initial symptoms were headaches, cerebellar syndrome, and gait ataxia, consistent with those found in the literature, contrary to his young age, with the median described being 40 to 50 years. The diagnosis of these tumors is made by correlating symptoms with radiographic findings. On T1-weighted Magnetic Resonance Imaging (MRI) sequences, they typically appear as a hypointense mass and hyperintense on T2 sequences and diffusion-weighted imaging with sharp but irregular boundaries, without perilesional edema or contrast enhancement. Indeed, the signal is often heterogeneous; its intensity may vary depending on the protein content of the tumor. Atypical forms have been reported, with a mass spontaneously hyperintense on T1 and hypointense on T2, probably due to the presence of calcifications and a high protein content.[17] Differential diagnoses include arachnoid cysts, dermoid cysts, hydatid cysts, and neurocysticercosis.[1,8] The differential diagnostic problems of arachnoid cysts and tumoral cysts are bypassed by the heterogeneous appearance on FLAIR sequence, increased signal on diffusion sequence, and especially the hyperintense and heterogeneous appearance on CISS-3D sequence.[18,19]

The standard treatment for epidermoid tumors is total gross surgical resection.[1] Histological analysis of epidermoid cysts is the same regardless of intracerebral location. From a therapeutic point of view, total excision of the cyst and its capsule remains the only guarantee of definitive cure, relying on intracapsular reduction and extracapsular dissection.[11] However, the intimate adhesion of the tumor capsule to delicate neurovascular structures, as observed in our intraoperative case, where it was very adherent to the bulbar triangle of the fourth ventricle floor, makes this total excision very laborious given the neurological and vital risks involved, notably the risk of involvement of cardiorespiratory centers as well as various cranial nerve nuclei.

Thus, in a literature review by Tancredi A. et al.,[13] on 66 patients operated for a V4 epidermoid cyst between 1974 and 2003, total excision was achieved in only 30% of cases. In such situations, total resection must be replaced by a subtotal approach.[3]

The postoperative course is generally straightforward; however, two complications may alter the course of the disease: aseptic meningitis and malignant transformation.[9] Chemical meningitis can occur from a single contact of the cystic tumor content with cerebrospinal fluid during surgery.[1] and lead to communicating hydrocephalus, the prevention of which requires as complete removal as possible, avoiding dispersion of cyst content intraoperatively, and irrigation of the operative field with hydrocortisone.[1,18] For the treatment of aseptic meningitis, it relies on repeated lumbar punctures and corticosteroids.[9] In contrast, malignant transformation is an extremely rare complication and, when present, is associated with the development of squamous cell carcinoma.[14,15]

In the case of incomplete tumor resection, although the growth rate of the residual lesion is as slow as that of the original tumor, annual magnetic resonance imaging follow-up is necessary.[16] In such cases, studies indicate tumor recurrence at a time interval equal to the patient's age at diagnosis plus nine.[4]

Surgical approach should be careful and based on a combination of symptoms and radiological data, considering the slow growth of residual tumor.[11] The literature indicates that adjuvant treatment of epidermoid cysts is rare, according to some authors, nonexistent.[17] However, Parikh et al. reported a successful case of inhibiting the recurrence of a benign intracranial epidermoid cyst using radiation during two years of follow-up.[18]

In postoperative surveillance, diffusion imaging is used to determine whether excision is complete or not. In case of residual tumor, annual MRI follow-up assesses the evolutionary potential of the residue [12].

CONCLUSION:

Epidermoid cysts of the fourth ventricle are rare and benign intracranial lesions. They tend to attain large sizes before manifesting symptoms or cerebellar dysfunction. Initial symptoms result from increased intracranial pressure, which can be mitigated by placing a ventriculoperitoneal shunt. MRI is of great diagnostic value, and total surgical excision should be considered the procedure of choice. However, due to slow growth and adherence to delicate nervous system

structures, subtotal approach should be contemplated and is associated with satisfactory outcomes.

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