Epidermoid cyst of the pineal region:report of a case

Comment [ak1]: A case report

Abstract :

Intracranial epidermoid cysts are one of the rare intracranial tumors. They represent 0.2 to 1% of intracranial tumors and 7% of cerebellopontine angle tumors. The pineal location is exceptional for this type of tumor. Cushing was the first to report the pineal location of the epidermoid cyst in 1928. We report a clinical case concerning a 30-year-old young man who presented with intracranial hypertension for 6 months. Clinical examination found stage III papilledema at the fundus and the rest of the clinical examination was unremarkable. MRI showed a pineal region process. The patient underwent surgery and underwent total excision of the lesion.

Comment [ak2]: This line may be omitted from abstract

Introduction :

Epidermoid cysts (EC), also called “primary cholesteatomas” or “pearled Cruveilhier tumors”, are rare, benign, congenital tumors developed from ectodermal inclusions [1]. The classic locations are: the cerebellar pontine angle (half of the cases), the temporal fossa, the suprasellar region and the quadrigeminal region. The location in the pineal region is rare.

Comment [ak3]: are the

Materials and methods :

Comment [ak4]: Case Report

We report the case of a 31-year-old patient, who had intracranial hypertension for 6 months, the clinical examination found a conscious patient GCS 15 without sensory-motor deficit with papilledema stage III in the fundus, Brain CT showed a lesion developing in the pineal region, hypodense, not enhancingcontrast, withhydrocephalus (figure 1).MRI showed a process in the pineal region with regular contours, well limited, hypo-intense in T1, hyper-intense in T2 (Figure 2), heterogeneousFLAIR, filling the supra-vermiancisternwith mass effect on the vermis and Sylvius aqueduct with tri-ventricular hydrocephalus. The patient benefited from a first ventriculo-cisternostomy (VCS), then through a supra-cerebellar infratentorial approach he benefited from a macroscopically total excision of a tumor with a pearlyappearance. The evolution was marked by a good improvementwithcompletedisappearance of clinicalsigns, radio-clinical follow-up wasrecommended (figure 3).

Comment [ak5]: Start the case report with brief history, presenting complaints, symptoms related to intracranial hypertension. Then general physical examination findings and relevant investigative findings should be mentioned.
Also mention the sex of the patient.
The abbreviated forms such as CT, MRI must be elaborated at the first mention.

Comment [ak6]: Mention full form

Comment [ak7]: Histopathological findings have not been mentioned.

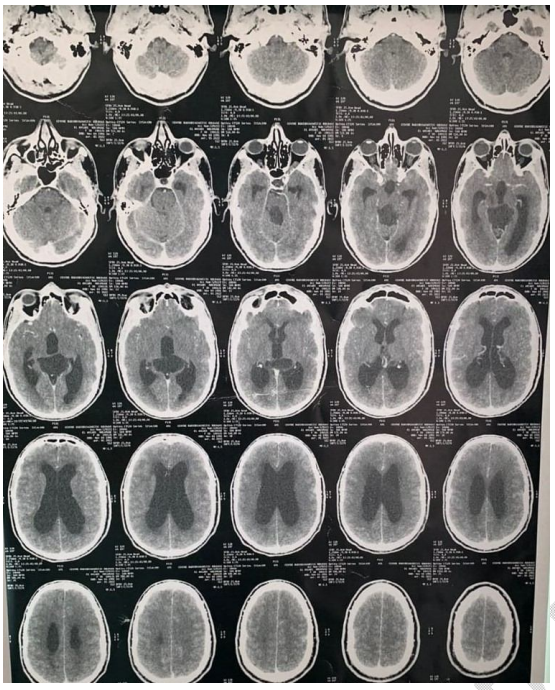
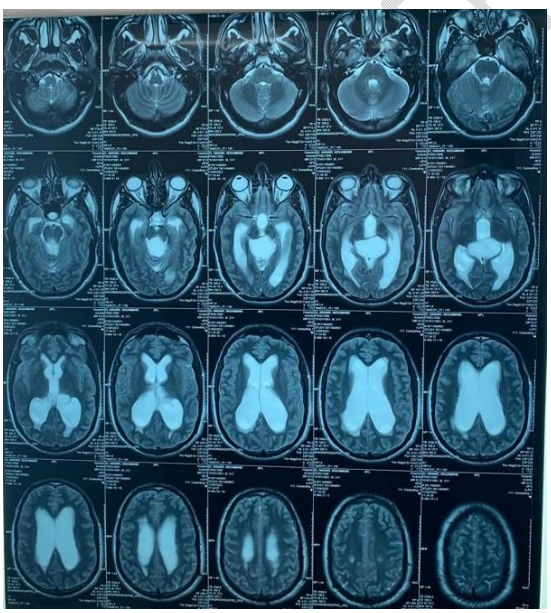


Figure 1: Brain CT C+ axial sections showing a lesion in the hypodense pineal region that does not take up the contrast.



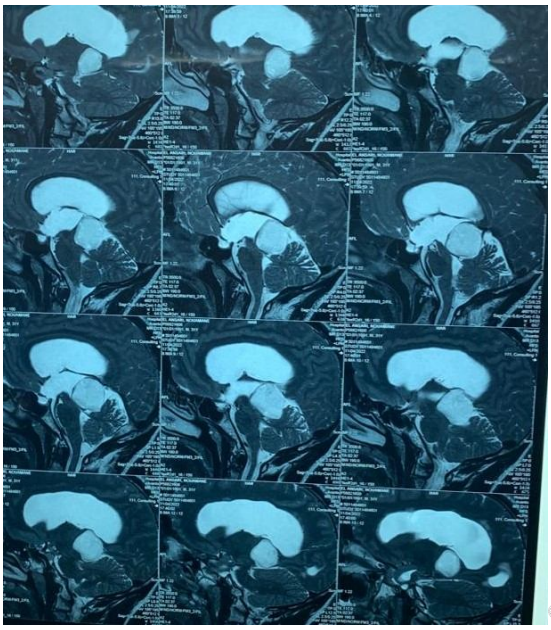


Figure 2: Brain MRI T2 sequence axial and sagittal section showing a hyperintenselesion in the pinealregionwithregular contours.

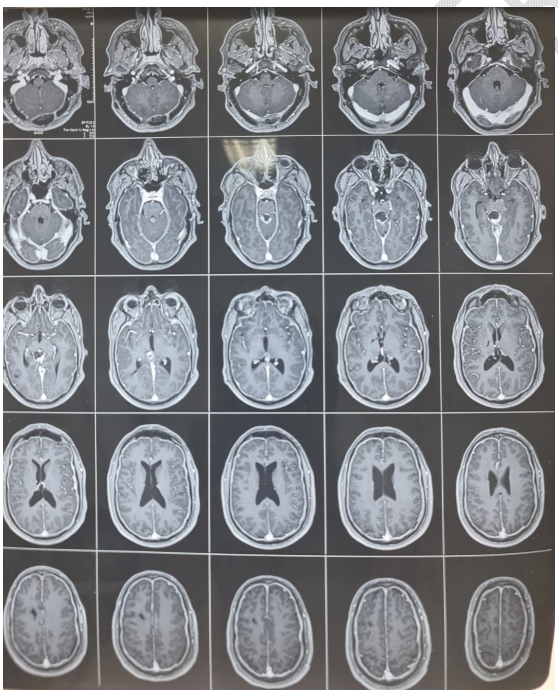


Figure 3: Axial T1-enhanced brain MRI, performed postoperatively, showing a small residue of the epidermoid cyst in the pineal region.

Discussion :

Epidermoid cysts, also called primary cholesteatomas or “Cruveilhier pearl tumors”, are rare, benign, dysembryoplastic tumors; they constitute approximately 1% of all primary intracranial tumors [1]. Developed from ectodermal inclusions [1]. Theymostoftenresult from an incomplete cleavage of the neural ectoderm and the cutaneous ectoderm, at the time of closure of the neural tube between the third and fifth week of gestation, with retention of ectodermal cells in the nervous system [1.2]. They can, more rarely, be secondary to post-traumatic or iatrogenic penetration [3] of the epidermis at the level of the subarachnoidspaces.

This lesion is often located at the cerebellopontine angle while the dermoid cyst prefers the medial location [4]. Pineal localization is a very rare form of this intracranial lesion. It represents 0.2-1% of all intracranial tumors [5]. Cushing was the first to report the pineal location of the epidermoid cyst in 1928 [5]. Then, many other authors have reported some cases of pineal epidermoid cyst [12]. Until 1974, 9 cases were reported in the literature [13]. In 1999, 11 cases were analyzed by Mackay et al. [14]. Since then, many other articles have been published focusing primarily on surgical treatment.

The clinical presentation is often characterized by Parinaud's syndrome and hydrocephalus. Hemiparesis and cerebellarsignsmayalsobenoted [14].

Previous studies have described depressive symptoms in pineal injury[6]; as well as a case of schizophrenia cured after excision of the lesion[6].

An unusual presentation for this rare tumor in an adolescent, diplopia and lateral rectus muscle paralysis[7].

These lesions are made of a pearly white, soft mass surrounded by a capsule often adherent to the adjacent walls. The cystic content is avascular and presents, when cut, a yellowish content, of more or less viscous consistency, reminiscent of candle wax and arranged in concentric strips [8]. The lesion grows slowly, and has a flexible and deformable character, adapting to the spaces in which it evolves [8]. Cysts grow by progressive desquamation of epithelial cells whichtransformintokeratin and cholesterolcrystals.

CT scan shows a cystic lesion. The density is similar to that of cerebrospinal fluid. We can find a lesion of the quadrigeminal cistern sometimes causing hydrocephalus without enhancement after injection of contrast product. Variable imaging is due to the difference in cholesterol and protein content and the presence of hemorrhage. On MRI, the epidermoid cyst is hypointense on T1-weighted images and hyperintense on T2-weighted and FLAIR images without contrast enhancement [4]. Diffusion sequences (DWI) can differentiate between an epidermoid cyst and an arachnoid cyst [14]. In diffusion, epidermoid cysts are bright compared to other cystic lesions [5].

The main point of surgical treatment is radical excision of the epidermoid cyst with its capsule. However, it is a real challenge due to this location. Some authors prefer to intentionally leave fragments of the adherent capsule in situ in the deep veins of this region to avoid any risk. Konovalov et al. [4] specified that radical excision was only possible in 50% of the cases presented in this series. Two approaches were described by Yasargil [4] in the

Comment [ak8]: Merge these sentences

Comment [ak9]: [1,2]

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Comment [ak11]: After reference 5, suddenly reference 12, 13, 14 have been cited. Then reference 6, 7 8 appear.
References must be numbered chronologically.

surgical management of pineal epidermoid cysts; the supra-cerebellar infra-tensorial approach and the occipital trans-tentorial approach. The latter is preferred for a lesion with a significant supratentorial component [4,5].

The supracerebellar infratentorial approach is the mostused for surgicaltreatment[4] Otherapproaches are used, including the trans-callosal interhemispheric approach [9], the trans-ventricular approach [5], the combined supra-infratentorial approach [14]. The ventriculoperitoneal shunt could be used in certain cases of hydrocephalus with intracranial hypertension [5]. Therapeutic stereotaxic aspiration is also proposed for the treatment of epidermoid cyst. Kitchen et al. [5] reported a case of VP shunt and stereotaxic aspiration. This technique still has many disadvantages. First of all, aspiration does not detach the capsule which represents a high risk of recurrence, spontaneous rupture of the cyst, aseptic meningitis and malignant transformation of the epidermoidcyst.

Comment [ak12]: Treatment [4].

The direct surgical approach appears to be more useful for these patients. Mackay [14] analyzed 12 reported cases of pineal epidermoid cyst since 1968. The outcome was good in 10 of the 12 cases. Two cases of aseptic meningitis. One deathwasrecorded.

Comment [ak13]: Elaboarate

As part of postoperative monitoring, diffusion sequence MRI allows precise verification of the completeness or incompleteness of the excision [8]. Two complications can modify the evolutionof KE:rupture and malignantdegeneration [8]. Cystic rupture is the most frequent complication, it is most often secondary to surgical manipulations, more rarely spontaneous and manifests as aseptic chemical meningitis. As for malignant forms, they are rare and exist in the form of squamous cell carcinomas [11].
In the event of incomplete excision, the growth of the residue is as slow as that of the native tumor; it nevertheless requires annual monitoring, allowing its evolving potential to be assessed [1].

Comment [ak14]: ?

Conclusion :

The pineal epidermoid cyst is a rare tumor. The treatment of choice is a total resection of the tumor. But sometimes, this is not always possible due to the characteristics of the tumor and the pineal region.

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