Asian Journal of Research in Surgery

Volume 7, Issue 2, Page 411-415, 2024; Article no.AJRS.120050

Epidermoid Cyst of the Pineal Region: Report of a Case

A Regragui [a,b*], MA Daraabou [a,b], M Hakkou [a,b], MY Ouadghiri [a,b], A Melhaoui [a,b], M Boutarbouch [a,b], Y Arkha [a,b] and A Ouahabi [a,b]

[a] Department of Neurosurgery, Specialty Hospital, Ibn Sina University Hospital, Raba, Morocco.
[b] Faculty of Medicine and Pharmacy of Rabat, Mohammed V University, Morocco.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/120050

Case Report

Received: 20/05/2024
Accepted: 23/07/2024
Published: 23/10/2024

ABSTRACT

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

Keywords: Epidermoid cyst; intracranial tumors; MRI; hypertension; pineal region.

**Corresponding author: E-mail: regragui.asmae7891@gmail.com, asmae.regragui123@gmail.com;*

Cite as: *Regragui, A, MA Daraabou, M Hakkou, MY Ouadghiri, A Melhaoui, M Boutarbouch, Y Arkha, and A Ouahabi. 2024. "Epidermoid Cyst of the Pineal Region: Report of a Case". Asian Journal of Research in Surgery 7 (2):411-15. https://journalajrs.com/index.php/AJRS/article/view/238.*

1. 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.

2. CASE PRESENTATION

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. A brain CT showed a lesion developing in the pineal region, hypodense, not enhancing contrast, with hydrocephalus . MRI showed a process in the pineal region with regular contours, well-limited, hypo-intense in T1, hyper-intense in T2 (Fig. 1), heterogeneous FLAIR, filling the supra-vermian cistern with mass effect on the vermis, and Sylvius aqueduct with tri-ventricular hydrocephalus. The patient benefited from a first ventriculo-cisternostomy (VCS), and then, through a supra-cerebellar infratentorial approach, he benefited from a macroscopically total excision of a tumor with a pearly appearance. The evolution was marked by a good improvement with the complete disappearance of clinical signs; radio-clinical follow-up was recommended (Fig. 2).

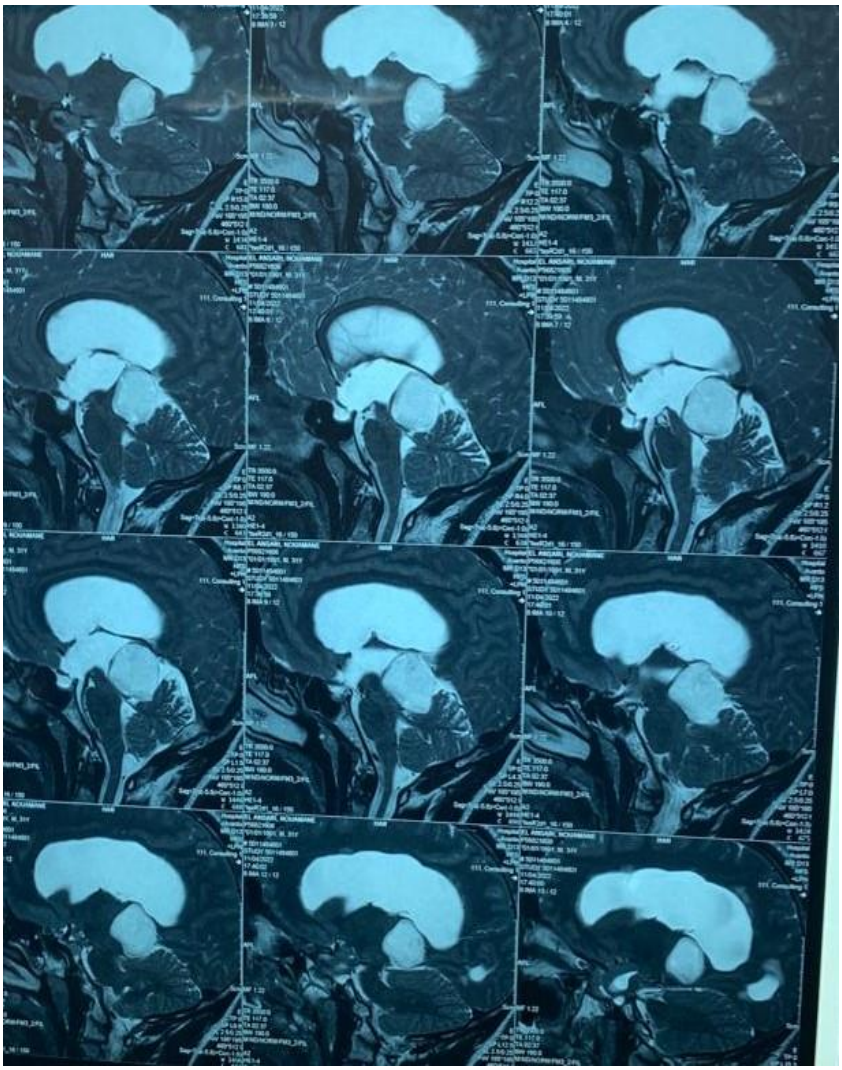

Fig. 1. Brain MRI T2 sequence axial and sagittal section showing a hyperintense lesion in the pineal region with regular contours

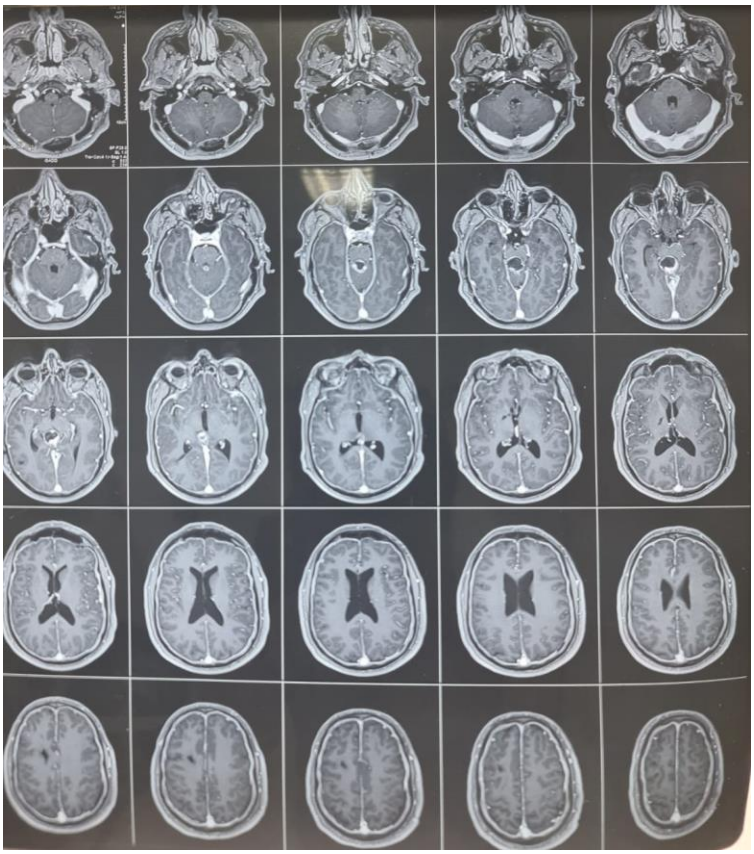

Fig. 2. Axial T1-enhanced brain MRI, performed postoperatively, showing a small residue of the epidermoid cyst in the pineal region

3. 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]. “They most often result 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 weeks 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 subarachnoid spaces.

“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 cysts” [6]. “Until 1974, nine cases were reported in the literature” [7]. In 1999, 11 cases were analyzed by Mackay et al. [8]. 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 cerebellar signs may also be noted” [8].

Previous studies have described depressive symptoms in pineal injuries [9], as well as a case of schizophrenia cured after the excision of the lesion [9].

An unusual presentation for this rare tumor in an adolescent is diplopia and lateral rectus muscle paralysis [10].

“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" [11]. "The lesion grows slowly and has a flexible and deformable character, adapting to the spaces in which it evolves" [11]. Cysts grow through the progressive desquamation of epithelial cells, which transform into keratin and cholesterol crystals.

"A 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" [8]. In diffusion, epidermoid cysts are bright compared to other cystic lesions [5].

The main point of surgical treatment is the 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 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 most commonly used for surgical treatment" [4]. Other approaches are used, including the trans-callosal interhemispheric approach [12], the trans-ventricular approach [5], and the combined supra-infratentorial approach [8]. "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 cysts. 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 epidermoid cyst.

The direct surgical approach appears to be more useful for these patients. Mackay [8] analyzed 12 reported cases of pineal epidermoid cysts since 1968. The outcome was good in 10 of the 12 cases. There are two cases of aseptic meningitis. One death was recorded.

As part of postoperative monitoring, diffusion sequence MRI allows precise verification of the completeness or incompleteness of the excision [11]. Two complications can modify the evolution of KE: rupture and malignant degeneration [11]. 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 [13,14].

"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].

4. 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.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of this manuscript.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Lopes M, Capelle L, Duffau H, Kujas M, Sichez JP, Van Effenterre, R. Supported surgical treatment of intracranial epidermoid cysts About 44 patients and review of the literature. Neurosurgery. 2002;48(1): 5-13.
2. Doll A, Abu Eid M, Kehrli P, Esposito P, Gillis C, Bogorin A. Aspects of Flair sequences, 3D-CISS and diffusion-weight MR imaging of intracranial epidermoid cysts. J Neuroradiol. 2000;27(2):101-6.
3. Talacchi A, Sala F, Alessandrini F, Turazzi S, Bricolo A. Assessment and surgical management of posterior fossa epidermoid tumors: report of 28 cases. Neurosurgery. 1998;42(2):242-51; Discussion 251-2
4. Konovalov AN, Spallone A, Pitzkhelauri DI. Pineal epidermoid cysts: diagnosis and management. J Neurosurg. 1999;91(3): 370-4.
5. Fahd Derkaoui Hassani, Abdelali Bouchaouch, Nizare El Fatemi, Rachid Gana, Najia El Abbadi, Moulay Rachid Maaqil. Pineal epidermoid cyst: Case report and review of the literature. The Pan African Medical Journal ; 2014.
6. Smaltino F, Cucciniello B. A case of epidermoid of the epiphysary region. Rass Int Clin Ter. 1967;47(16):881-8.
7. Sambasivan M, Nayar A. Epidermoid cyst of the pineal region. J Neurol Neurosurg Psychiatr. 1974;37(12):1333-5.
8. MacKay CI, Baeesa SS, Ventureyra EC. Epidermoid cysts of the pineal region. Childs Nerv Syst. 1999;15(4):170-8.
9. Jiang X, Chen Y, Zhou Z, Luo L, Hu W, Zheng H, Zhu Z, Wang J, Chen Z, Surgical resection of pineal epidermoid cyst contributed to relieving schizophrenia symptoms, World Neurosurgery ; 2018.
10. Koushik Handattu. Unilateral lateral rectus palsy: an unusual presentation of pineal epidermoidcyst. Pan African Medical Journal. 2020;36(252).
11. Noureddine Oulali, Faycal Moufid, Mohamed Rachid Ghailan, Brahim Hosni. Epidermoid cyst of the greater cistern and the fourth ventricle. The Pan African Medical Journal ; 2012.
12. Jia W, Ma Z, Liu IY, et al. Transcallosal interforniceal approach to pineal region tumors in 150 children. J Neurosurg Pediatr. 2011;7(1):98-103.
13. Goldman SA, Gandy SE. Squamous cell carcinoma as a late complication of intracerebroventricular epidermoid cyst: Case report. J Neurosurg ; 1987. Apr;66(4):618-20. This article on PubMed
14. Hila H, Bouhaoula MH, Darmoul M, Jelassi H, Yedeas M. Vermian epidermoid cyst Revealed By Head Injury. Neurosurgery. 2006;52(1):63-6.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/120050