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Case Report

Dermoid cyst, unusual location of the pterion: About a case and review of literature [☆]

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ABSTRACT

Intracranial dermoid cysts are benign tumors of congenital origin. The intradiploic forms are rare, exceptional at the pterional level. We report the case of a 10-year-old girl who presented with a cutaneous fistula in the left frontotemporal region. The blind end of the fistula was an intradiploic dermoid cyst in the rare location of the pterion confirmed by imaging and histopathology. Once this lesion is suspected, it is important to identify its location and morphology using imaging techniques and to complete excision of the cyst to avoid complications associated with infection and to mitigate the risk of subtotal resection.

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Introduction

The dermoid cyst is a congenital tumor usually located intradurally and rarely occurs in the intradiploic space in the skull. Imaging is very useful for the precise diagnosis of this lesion.

Case presentation

We report the case of a 10-year-old girl, with no previous history, who consulted with a skin lesion in the left frontotemporal region. The mother confirmed the presence of a small tumefaction in the same region since birth, which had in-

creased in volume over the past year, with recurrent discharge of pus and blood.

At the admission, the patient was in good general status, Glasgow Coma Scale was 15/15 with normal neurologic examination. The general examination revealed a left temporal skin lesion.

A cerebral computed tomography (CT) scan showed an intradiploic cystic lesion in the pterional region on the left side. A cerebral MRI revealed a cystic lesion without intracranial extension in favor of a dermoid cyst of the left pterion fistulized to the skin (Fig. 1).

The affected person underwent a left pterional craniotomy (Fig. 2).

Intraoperatively, the temporalis muscle was disinserted and then reclined with a harder part corresponding to the path of the fistula; the exophytic lesion was entirely surrounded by bone matrix.

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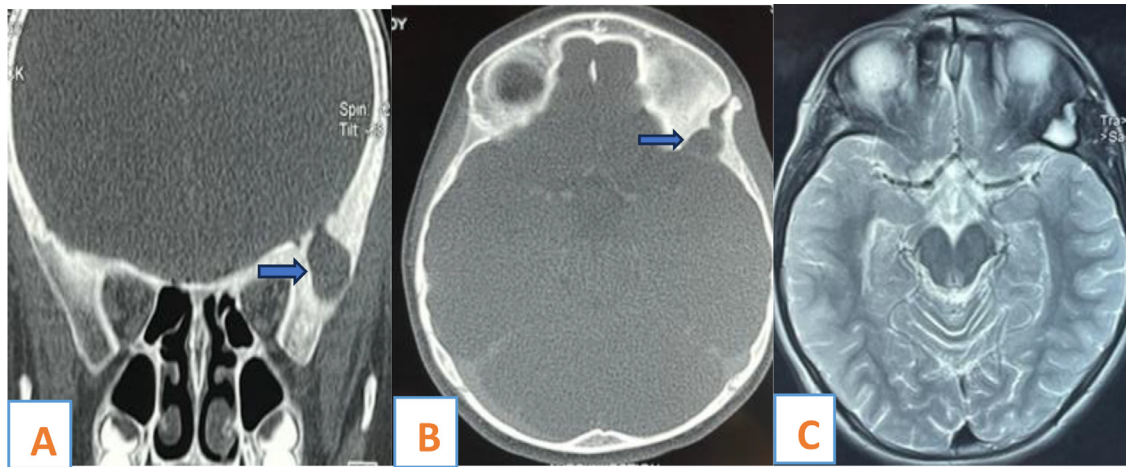


Fig. 1 – CT brain bone window coronal (A) and axial (B) sections of intradiploid dermoid cyst fistulizing at the skin (blue arrow). MRI well-circumscribed extracranial tumor component (C).

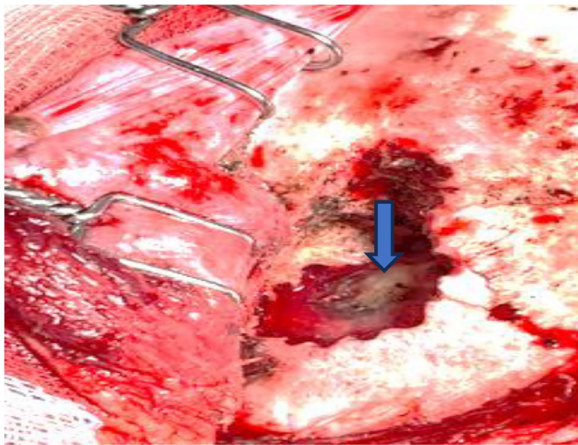


Fig. 2 – Intraoperative discovery of yellowish fatty tumor contents (blue arrow).

The intraoperative discovery of a purulent fluid discharge corresponding to the cystic portion and lumps evacuated by abundant washing with dirty serum with subsequent aspiration, afterward a complete resection of the cystic wall with co-

agulation of the hard meridian contact surface. No intradural tumor component was found. The extradural one was carefully removed.

The postoperative course was uneventful. The control CT scan (Fig. 3A), after 24 hours, confirms resection of the affected part of the pterion. The patient was doing well (Fig. 3B) and was discharged home on postoperative day 5 in stable condition, and back to school 2 weeks later.

The immunohistochemistry (Fig. 4) revealed a ruptured intradiploid dermoid cyst.

Discussion

The intradiploid dermoid cyst account for 0.04%–0.7% of cranial tumors [1]. They are composed of both ectodermal and mesodermal elements and form either as a result of incomplete closure of the neural tube during the third to fifth weeks of fetal development, or due to traumatic implantation of skin elements [2,3]. This lesion occurs most frequently in females, with a 2:1 ratio [4], and the fistula opening is present on the lateral side of the orbit (the eyebrow) [5], as in our case.

In the literature, the dermoid cyst is frequently described in the anterior fontanel. The other localizations are rarer [4]

Table 1 – Details of the present case and reported cases of frontotemporal intradiploid dermoid cyst DC and location of skin fistula.

Authors	Age	Sex	Localization of dermoid cyst	Localization of skin fistula
Hong [9]	2 years 6 months	F	Frontotemporal	Right temporal region
Vega et al. [5]	2 years	M	Intradiploid frontotemporal right coronal lateral suture	No fistula
Yamawaki et al. [10]	7 years	F	Right frontozygomatic suture	Right lateral orbital region
Hong et al. [1]	43 years	F	Frontotemporal	Left superolateral subcutaneous mass
Barkley et al. [11]	15 months	F	2 intradiploid pterional cysts	frontotemporal
Our case (2021)	10 years	F	Intradiploid pterion	frontotemporal

F, female; M, male.

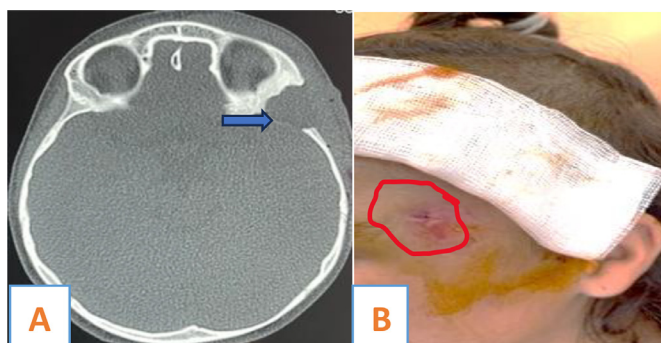


Fig. 3 – Postoperative CT scan bone window axial, resection of the intradiploic dermoid cyst (blue arrow) (A). The fistula skin scar (B) (red round).

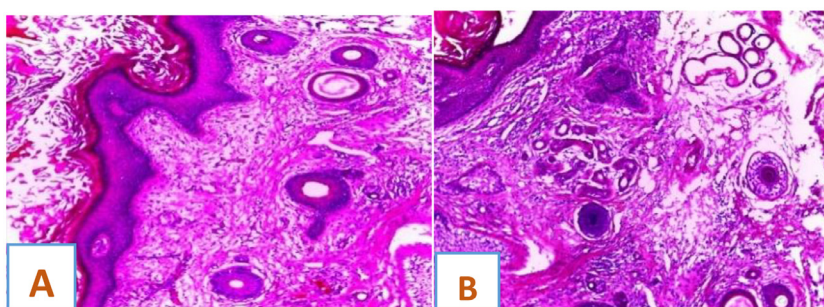


Fig. 4 – Pathologic analysis of intraoperative specimens. Hematoxylin and eosin, magnification $\times 10$. A cyst bordered by a keratinized squamous lining (A), with appendages: hair follicles (A) and sweat glands (B).

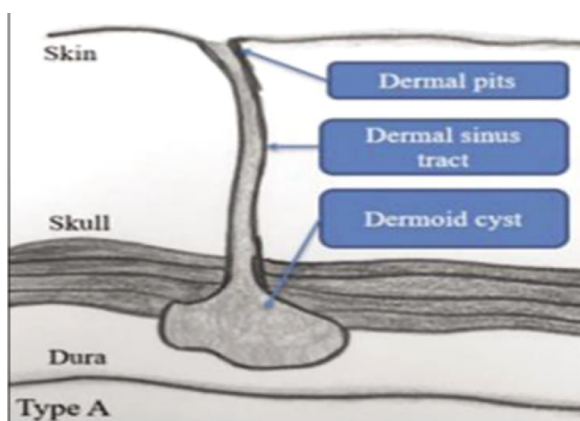


Fig. 5 – Type A of Hong et al. classification [1].

include the petrous apex, the eustachian tube, the periorbital or perinasal soft tissues, the cavernous sinuses, the anterior cranial fossa or in the skull suture near the skin, the skull base, the infratemporal region, the supratentorial region or the fourth ventricle [2]. In our case, the cyst was located at the pterional level, an unusual localization (Table 1).

According to a classification established by Hong et al. [1], our case is a type A (Fig. 5), characterized by a dermal fistula terminating in a pocket with a dermoid cyst in the extradural intradiploic areas. The dermoid cyst can be seen more or less at birth, and has a tendency to enlarge, which

is the most frequent reason for consultation [4]. This type is diagnosed exclusively in childhood, due to recurrent local infections [1].

Their clinical presentation depends on the size and location of the tumor. The patients with the dermoid cyst may be asymptomatic, the only clinical sign being a skin lesion, as in our case. This, therefore, highlights the importance of radiology in determining the presence of any intracranial extension, as well as aiding postoperative follow-up of these tumors.

The CT scans can easily depict the dermoid cyst as low-density lesions, a feature that is consistent with their fat content. In addition, peripheral calcifications can be spotted in the clear zone [6]. The cyst wall may become partially enhanced after administration of iodinated contrast medium on CT [7].

On MRI, they tend to appear as hyperintense lesions on T1-weighted imaging and as a heterogeneous lesion on T2-weighted imaging due to their varied content including bone, cartilage and calcifications.

The differential diagnosis can be made with squamous cell cysts, lipomas, teratomas, cystic craniopharyngiomas, and arachnoid cysts [2].

Despite their benign nature, the dermoid cysts have a high morbidity and mortality rate in the event of rupture. The latter, whether spontaneous or traumatic, leads to severe aseptic or chemical meningitis, responsible for transient cerebral ischemia secondary to vasospasm, and ultimately leading to infarction, coma and death. It can also lead to chronic granulated arachnoiditis, aqueduct stenosis or ventriculitis [2].

The complete surgical resection of the dermoid cyst is the only effective treatment to prevent future recurrence and/or complications [8].

Conclusion

Our case is a rare presentation of an intracranial extradural dermoid cyst which highlights the need for early imaging in the evaluation of any skin fistula in the head and neck region prior to surgical treatment.

Patient consent

Informed consent was obtained from the parents of the patient prior to the submission of this article. Also, this article respects both the Consensus-based Clinical Case Reporting Guideline and the Recommendations for the Conducting, Reporting, Editing, and Publication of Scholarly Work in Medical Journals.

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