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## Case Report

# Intradural dermoid cyst with complete dermal sinus of the posterior fossa: Contribution of 3D imaging with histopathological correlation<sup>☆,☆☆</sup>

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## ARTICLE INFO

## Article history:

Received 19 December 2021

Revised 11 March 2022

Accepted 12 March 2022

## Keywords:

Intradural dermoid cyst

complete dermal sinus

posterior fossa

3D CT

restricted diffusion

## ABSTRACT

A 5-year-old girl who presented with two episodes of meningitis, had a patchy red area and a small skin dimple in the midline of the occiput on physical examination. Imaging revealed a well-demarcated oval intradural lesion of the posterior fossa with restricted diffusion and peripheral enhancement, raising the possibility of an abscess. The 3D volume rendering of CT images of the inner surface of bone showed chronic bone remodeling and a tiny bone defect of the outer table. This detailed anatomical evaluation has an added value to MRI characteristics to orient for a preoperative diagnosis of an intradural dermoid cyst with a dermal sinus, that was confirmed by histopathological analysis after surgical excision.

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## Introduction

Intracranial dermoid cysts are rare, benign, slow-growing congenital abnormalities, accounting for 0.1%–0.7% of all intracranial tumors [1]. They arise from ectopic deposition of epithelial cells during neural tube closure, between 3 and 5 weeks of pregnancy [2]. Enclosed ectodermal cysts develop when the surface ectoderm fails to completely detach from

the underlying neural tube. They usually present in the second and third decades but can occur in infants and young children [2–4]. The initial presentation of an infected dermoid cyst associated with a dermal sinus is uncommon. This persistent dermal sinus establishes a connection between the intracranial tumor and the extracranial occipital bone and skin. The present case highlights the added value of 3D volume rendering of CT images, in addition to MRI, to establish the diagnosis.

<sup>☆</sup> Acknowledgments: None.

<sup>☆☆</sup> Competing Interests: None.

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<https://doi.org/10.1016/j.radcr.2022.03.047>

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## Case presentation

### Clinical presentation

A 5-year-old girl was admitted to our emergency department complaining of a 3-day history of high-grade fever, lethargy, headache, and vomiting. One month prior to the current admission, she had similar symptoms, diagnosed in another institution as having bacterial meningitis, and treated with intravenous Vancomycin and Ceftriaxone for 14 days. She had no history of brain trauma. Upon admission, she was febrile (38.6°C), tachycardic (heart rate:144), tachypneic (respiratory rate 22/min) with a normal oxygen saturation on room air (96%), and normal blood pressure (110/69 mmHg). On physical examination, she had severe neck stiffness and a patchy red discoloration with a small skin dimple at the midline of her occiput.

### Laboratory evaluation

Blood tests revealed a high C-reactive protein of 140 mg/L (<5 mg/L), increased white blood cells ( $21 \times 10^3$  cells/mm<sup>2</sup>, normal range: 4.5–13.5 cells/mm<sup>2</sup>) with neutrophil predominance (90%). She underwent a lumbar puncture. The cerebrospinal fluid analysis revealed increased white blood cells (1920 cells/mm<sup>2</sup>, normal value <7 cells/mm<sup>2</sup>) with a predominance of neutrophils (88%), red blood cells (60 cells/mm<sup>2</sup>), a decreased glucose level (11 mg/dl, normal range:40–80 mg/dl) and a normal protein level (9.9 mg/dl, normal range:10–60 mg/dl).

The patient was started on Cefepime and Vancomycin. The cerebrospinal fluid microbiological analysis, including polymerase chain reaction, was negative for the following viruses and bacteria: herpes simplex virus 1&2, Epstein-Barr virus, cytomegalovirus, varicella-zoster, human herpesvirus 6, *Neisseria meningitidis*, *Hemophilus influenza*, *streptococcus pneumonia*, group B *streptococcus*.

### Imaging

A CT scan with intravenous contrast injection was performed in the emergency department. It revealed a  $21 \times 13$  mm oval hypodense collection with peripheral enhancement located between the cerebellar hemispheres (Fig. 1). Three-dimensional volume rendering of CT images showed smooth, well-corticated funneling of the inner table of the occipital bone, with an overlying 1 mm midline defect of the outer table (Fig. 1).

The patient underwent a brain MRI with intravenous contrast administration. The inter-cerebellar collection showed restricted diffusion, heterogeneous T2 signal, and peripheral enhancement, communicating with a subcutaneous nodular lesion with a low T2 signal intensity and homogeneous enhancement (Figs. 2–4). There were features of ventriculitis and cerebellitis (Fig. 5). Based on MR images, the differential diagnosis included an epidural abscess or an infected dermoid cyst (Table 1) [5,6]. The presence of bone remodeling of the inner table on 3D CT reformats was a sign of a slow growing, pre-existing process, favoring the diagnosis of a dermoid cyst.

### Surgery

The patient underwent surgical excision due to a persisting high-grade fever despite antibiotic therapy. The lesion was adherent to the inner layers of the dura mater and the surrounding cerebellar structures. It was carefully dissected and resected en bloc with its sinus tract, with preservation of cerebellar tissues.

### Histopathology

Macroscopic inspection showed a complex cystic lesion measuring  $22 \times 15$  mm containing pilar structures and a white friable material. Microscopic examination revealed a ruptured cyst lined by a stratified squamous epithelium and containing keratinaceous material. A granulation tissue was present, along with numerous histiocytes. A granulomatous inflammation, with a foreign body giant cell reaction consisting of multinucleated giant cells, was seen in contact with hair shafts (Fig. 6). The diagnosis of a ruptured dermoid cyst was confirmed. There was no sign of malignancy. Multi-resistant *staphylococcus aureus* (MRSA) was isolated from surgical swab culture, and a three-week course of vancomycin was continued.

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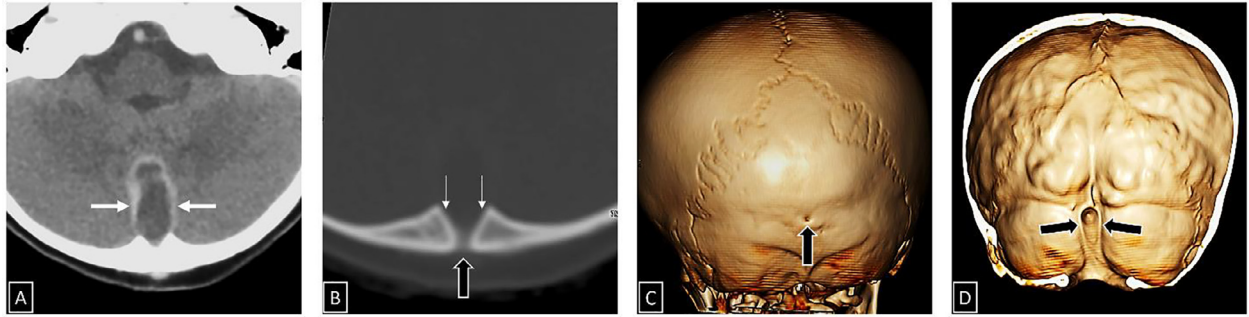
## Discussion

We report imaging findings of a complete dermal sinus associated with an infected intradural dermoid cyst with histopathological correlation, and to the best of our knowledge, only a few case reports have correlated imaging findings with histopathology [7,8].

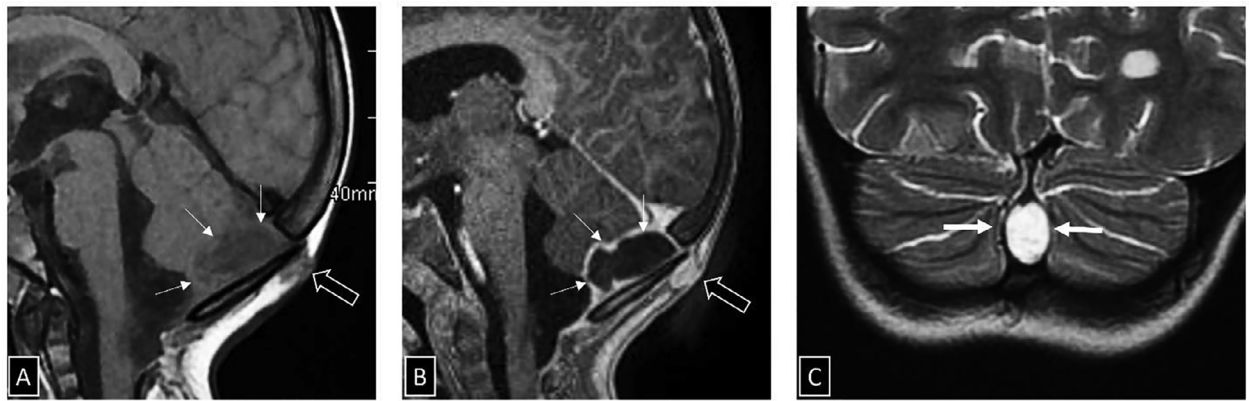
Dermoid cysts may remain asymptomatic, with a subtle skin dimple on physical examination [9,10]. They can rupture and manifest as recurrent meningitis due to the release of their content into the CSF (aseptic meningitis), or they can become infected, most commonly with *staphylococcus aureus* [11]. They can be complicated by abscess formation, hydrocephalus, epilepsy, cerebrovascular accident, or mass effect [3,9,10].

Dermoid cysts of the posterior fossa are rare and have a predilection for occurring in the midline. They are classified into four categories depending on the status of the occipital dermal sinus: (1) an extradural dermoid cyst with a complete dermal sinus; (2) an intradural dermoid cyst with no dermal sinus; (3) an intradural dermoid cyst with an incomplete dermal sinus; and (4) an intradural dermoid cyst with a complete dermal sinus [9]. This is related to the embryology of the falx and tentorium, presumably resulting from ectodermal fragments being pulled in during the invagination of the dural folds [7,9]. Infratentorial dermoid cysts may retain their attachment to the skin through a persistent narrow dermal sinus through an occipital bone defect, unlike those located in the supratentorial area. Therefore, they are more prone to getting infected [9].

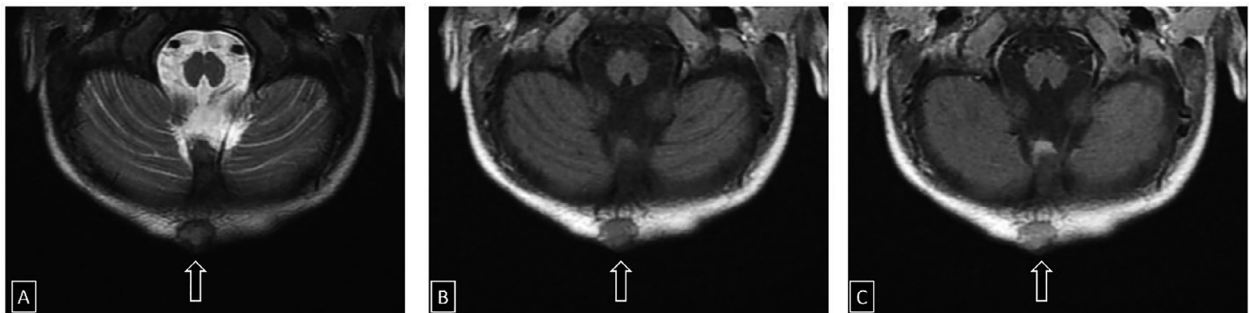
CT scan and MRI play an important role in the diagnosis of dermoid cysts and the assessment of their complications [12]. Findings on imaging are determined by the content of



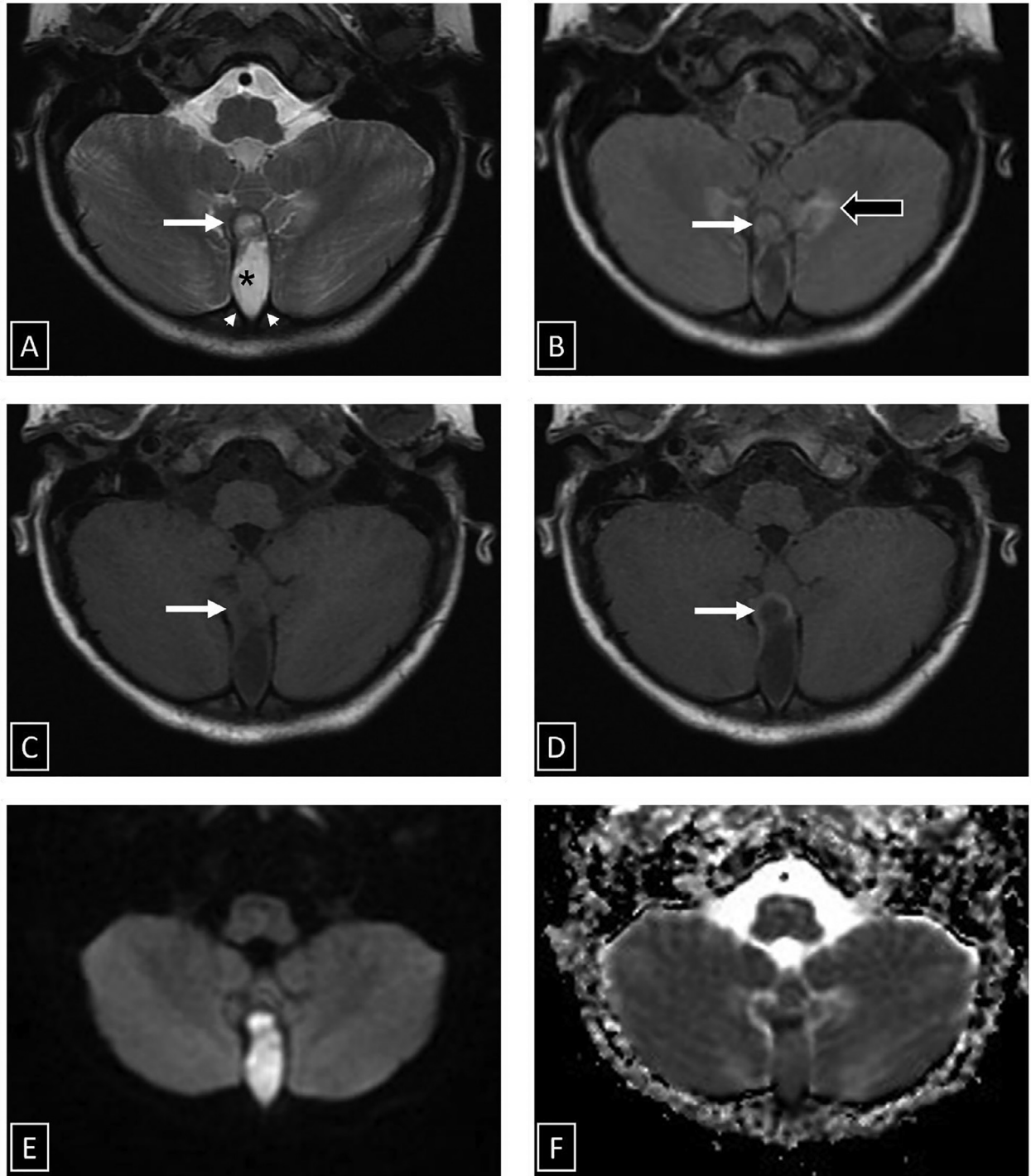
**Fig. 1** – Contrast-enhanced CT images in a 5-year-old girl with fever and meningismus. (A) axial image at the level of the posterior fossa shows an oval hypodense collection with peripheral rim enhancement (arrows) between the cerebellar hemispheres. (B) axial image reconstructed with bone kernel shows smooth funneling of the inner tables (white arrows) and a 1 mm defect in the outer table (black arrow). (C) 3D volume rendering of the outer surface of the skull shows a 1mm bone defect (black arrow). (D) 3D volume rendering of the inner surface of the skull shows an oval smooth funneling of the inner table with sharp well-defined edges (black arrows), a sign of a chronic process that probably occurred before complete ossification.



**Fig. 2** – (A) Sagittal nonenhanced T1 weighted MR image at the level of the midline structures of the posterior fossa, and (B) Sagittal T1-WI after intravenous administration of gadolinium chelate show an oval well-circumscribed peripherally enhancing lesion (white arrows) located between the cerebral hemispheres, communicating with an enhancing nodule in the scalp (black arrows). (C) On the coronal T2-WI MR image, the lesion has a bright T2 signal intensity, that seems to split the dura (white arrows).

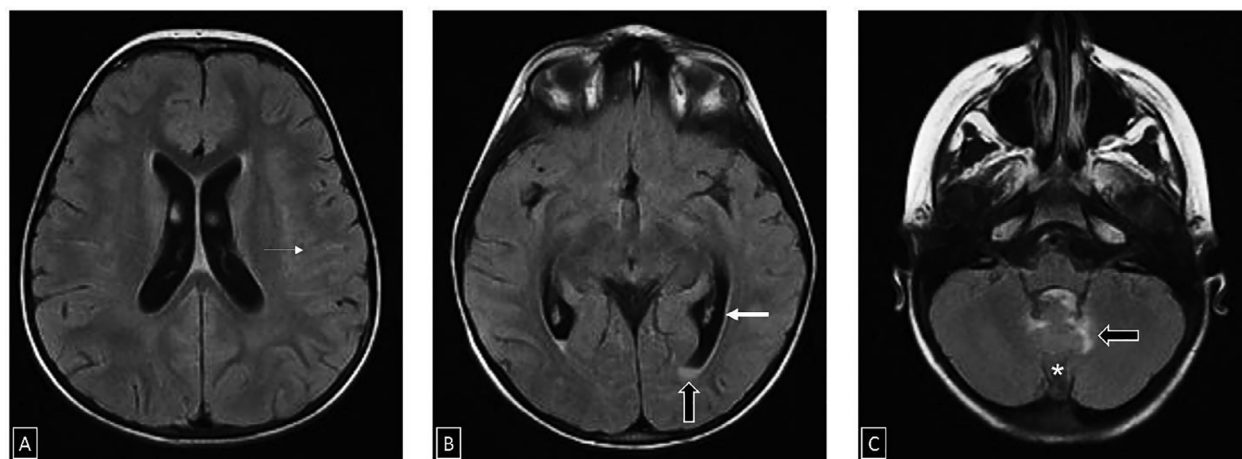


**Fig. 3** – (A) Axial T2-W MR image, (B) axial non-enhanced T1-W, and (C) contrast-enhanced T1-W MR images at the level of the posterior fossa, show a 2 cm enhancing nodule in the scalp with intermediate to low signal intensity on T2-WI (arrow in A), intermediate on T1-WI (arrow in B) with homogeneous enhancement after contrast administration (arrow in C).



**Fig. 4 – (A) Axial T2-W MR image, (B) fluid-attenuated inversion recovery (FLAIR), (C) nonenhanced T1-W, (D) contrast-enhanced T1-W, (E) diffusion-weighted (DWI), and (F) corresponding apparent diffusion coefficient map (ADC) MR images at the level of the posterior fossa, show a lesion between cerebral hemispheres. The lesion has a predominant T2 bright component (asterisk in A), and a minor nodular component with intermediate-to-low T2 signal intensity (white arrow in A), high on FLAIR (white arrow in B), intermediate on T1 (white arrow in C) with peripheral enhancement (white arrow in D). The lesion shows restricted diffusion (E and F). The posterior margin of the lesion fills the defect in the inner table of the occipital bone with sharp raised edges and seems to extend via the small defect in the outer table. Note increased signal intensity in the surrounding cerebellar parenchyma on FLAIR images (black arrow in B) suggestive of surrounding inflammation.**





**Fig. 5 – (A) Axial fluid-attenuated inversion recovery (FLAIR) image at the level of the body of the lateral ventricles shows a subtle increased signal in the sulci (arrow). (B) Axial FLAIR image at the level of the occipital horns shows thickening of the ependymal lining (white arrow) and fluid-fluid level in the occipital horns (black arrow). (C) Axial FLAIR image at the level of the cerebellar vermis shows increased signal intensity in the cerebellar parenchyma surrounding the lesion (asterisk). Findings are related to meningitis, ventriculitis, and early cerebellitis.**

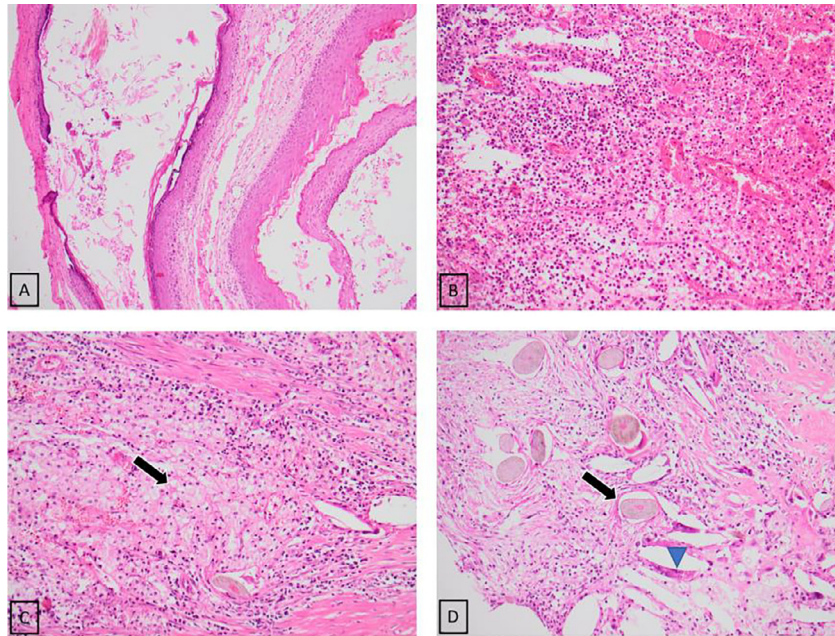
**Table 1 – Differential diagnosis of extra-axial lesions of the posterior fossa that show restricted diffusion on MRI.**

|                            | Abscess  | Epidermoid cyst   | Dermoid cyst  |
|----------------------------|--|---|---|
| <b>Composition</b>         | Collection of pus (broken-down tissues, dead bacteria and leukocytes, and extracellular fluid) [6] | Stratified squamous epithelial cells [2]  | Stratified squamous epithelial cells with sebaceous, hair, sweat glands [2]   |
| <b>Affected age</b>        | Any age  | Mostly in the 3rd-4th decades [2]   | Mostly in the 2nd-3rd decades [2]<br>May affect infants and young children  |
| <b>CT</b>                  | Thick-walled with central hypoattenuation, may contain air pockets                                 | Fluid attenuation like CSF  | Depending on composition<br>Heterogeneous fat attenuation   |
| <b>MRI</b>                 | Depending on the stage of abscess  | T1: Isointense to CSF<br>T2: Isointense to CSF  | Depending on composition:<br>T1:<br>- Heterogeneously hyperintense (fat)<br>- low signal if a low-fat component<br>T2: Heterogeneously hyperintense<br>Nonuniform suppression on fat-suppressed sequences |
| <b>Enhancement</b>         | Rim enhancement  | Minimal rim enhancement   | Rim enhancement (dermal adnexa in the wall)   |
| <b>Additional features</b> |  | Less common in the posterior midline fossa<br>More common in cerebellopontine angle [5] | If associated dermal sinus → dermoid cyst may become superinfected  |

the cyst: appendages of skin, such as hair follicles, sweat, and sebaceous glands, calcifications of dental enamel, and thick proteinaceous material [5,11]. They are frequently hypodense relative to water on CT, hyperintense on T1-weighted, and heterogeneous on T2-weighted MR images [5]. Post-contrast images may show peripheral enhancement due to dermal adnexa in the dermoid capsule [5]. Due to their thick content, they may cause restricted diffusion and resemble an abscess. The differential diagnosis of extra-axial lesions of the midline posterior fossa that show restricted diffusion lesions includes dermoid cyst, epidermoid cyst, and abscess (Table 1). In fact,

the current case report adds to the limited data available in the literature on the use of diffusion-weighted imaging to diagnose dermoid cysts [13,14]. In addition, we used 3D volume rendering of CT images that demonstrated smooth bone remodeling of the inner surface of the skull underlying a tiny aperture of the outer table, a useful ancillary sign indicating a pre-existing chronic process. To the best of our knowledge, this has not been previously described.

In summary, dermoid cysts of the posterior fossa can show restricted diffusion and peripheral enhancement on MRI, mimicking an abscess. The presence of bone remodel-



**Fig. 6 – (A) H&E staining (hematoxylin and eosin) (100X) shows a cyst lined by a stratified squamous epithelium and containing keratinaceous material. (B) H&E staining (200X) shows a florid granulation tissue consisting of numerous blood vessels and associated neutrophils. (C) H&E staining (200X) shows numerous foamy histiocytes (arrow). (D) H&E staining (200X) demonstrates multinucleated giant cells (arrowhead) in contact with hair shafts (arrow).**

ing of the inner surface of the skull on CT images, as well as a tiny aperture communicating with the overlying skin, could be useful hints to the presence of a complete dermal sinus.

### Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

### Patient consent

Written informed consent for publication of the case report was obtained from the parents.

### Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2022.03.047](https://doi.org/10.1016/j.radcr.2022.03.047).

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