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Occipital intraosseous dermoid cyst with restricted diffusion on magnetic resonance imaging in a child

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ABSTRACT

A 4-year-old girl presented repeatedly with a complicated occipital mass, which was erroneously treated as a pyogenic granuloma. Imaging performed before a planned surgical resection detected an underlying intraoccipital dermoid with a sinus tract to the skin surface and extension into the posterior fossa. This case highlights the value of high-resolution computed tomography imaging for depiction of anatomic details and the value of magnetic resonance imaging for differential diagnosis and surgical management. A comprehensive literature review of intraosseous dermoid cyst and detailed discussion of the differential diagnoses are provided.

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Case report

A 4-year-old girl presented repeatedly to a dermatology clinic with a 4-cm indurated plaque on her posterior scalp. This was initially thought to be a pyogenic granuloma and was treated with multiple extended courses of antibiotics and underwent incision and drainage twice without resolution of symptoms. The patient was then referred to a general surgery clinic for excision of the presumed subcutaneous soft tissue lesion. Intraoperatively, the lesion was found to have a sinus track extending down to the galea of the occipital bone. The

scalp lesion was removed and a cuff of pericranium was incised around the sinus track, revealing extension of the lesion into the occipital bone. An intraoperative neurosurgical consultant recommended brain magnetic resonance imaging (MRI) to better assess the full extent of the lesion. The surgery was aborted and the lesion was not excised pending neuroimaging.

The MRI of the brain with intravenous contrast showed a 2.5 × 1.6 × 1.7-cm round, well-marginated lesion centered within the midline occipital bone. The lesion demonstrated hyperintense signal on T2WI, low signal on T1WI with a thick peripheral rim of low signal on T2WI, and mild peripheral enhancement (Fig. 1). The lesion showed restricted diffusion, which can be

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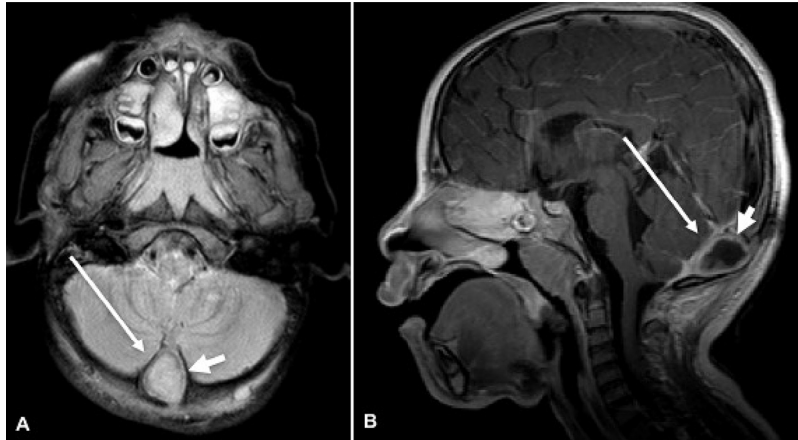


Fig. 1 – A 4-year-old girl with intraoccipital dermoid with associated dermal sinus tract. Preoperative MRI. Findings: The calvarial lesion demonstrates content that is of high T2 and low T1 signal (A and B, long arrows), and the lesion has a thick low T2 signal enhancing rim (A and B, short arrows). These imaging findings can be seen in abscess and dermoid cysts. Technique: (A) MRI, 1.5 T, T2, axial plane, TR 2500, TE 85, slice thickness 3 mm. (B) MRI, 1.5 T, T1 + contrast, sagittal plane, TR 615, TE 12, slice thickness 4.5 mm. Contrast: 5 mL MultiHance (Bracco Diagnostics Inc., Cranbury, NJ, U.S.). MRI, magnetic resonance imaging; TE, echo time; TR, repetition time.

seen with abscess or dermoid or epidermoid cyst (Fig. 2). The MRI located the lesion within the diploic space of the occipital bone, an extension into the epidural space of the posterior fossa, and a sinus tract extending to the overlying scalp (Fig. 3). On the bone windows of the computed tomography (CT) angiogram, the lesion remained posterior to the torcular and the sinus tract was identified as a calvarial lucency with sclerotic borders (Fig. 4).

Following MRI and CT angiogram, the patient underwent neurosurgical resection of the lesion via a transoccipital approach. Gross examination of the specimen at the time of

surgical resection demonstrated fat and hair follicles within a cystic lesion consistent with an occipital intraosseous dermoid cyst.

For several days following the procedure, the patient was noted to have slurred speech and ataxia. Postoperative MRI of the brain demonstrated T2/fluid-attenuated inversion recovery hyperintense, ring-enhancing lesions with restricted diffusion consistent with right cerebellar abscesses (Figs. 5). These were surgically drained and treated medically with antibiotics. The patient's symptoms gradually improved, and she was eventually discharged without any neurologic sequelae.

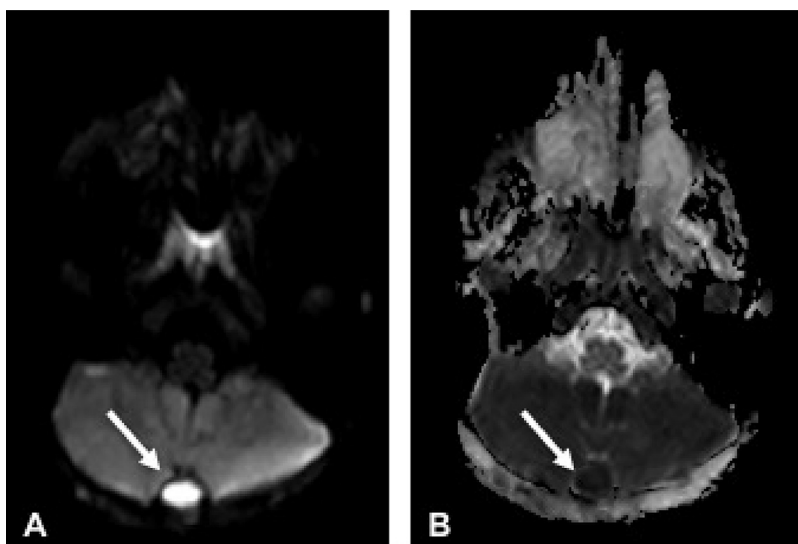


Fig. 2 – A 4-year-old girl with intraoccipital dermoid with associated dermal sinus tract. Preoperative MRI. Findings: The calvarial lesion demonstrates restricted diffusion with high signal on DWI (A) and low signal on ADC map (B). This finding can be seen both in an abscess and a dermoid cyst. Technique: (A) MRI, 1.5 T, DWI, axial plane, eB1000, TR 6011, TE 82, slice thickness 4 mm. (B) MRI, 1.5 T, DWI, ADC map, axial plane, TR 6011, TE 82, slice thickness 4 mm. ADC, apparent diffusion coefficient; DWI, diffusion weighted imaging; MRI, magnetic resonance imaging; TE, echo time; TR, repetition time.

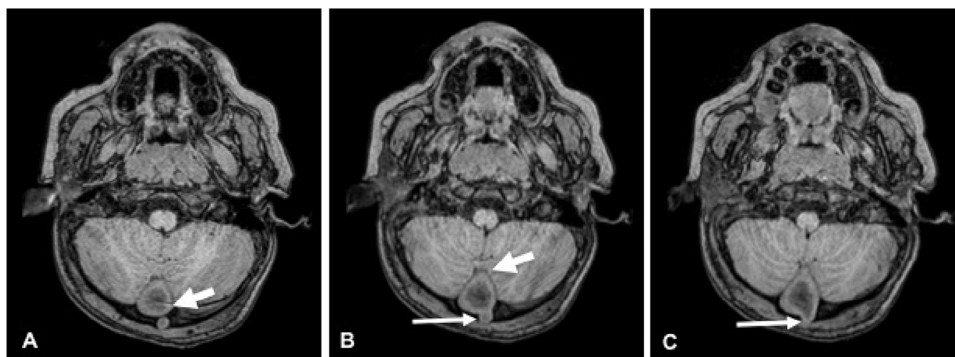


Fig. 3 – A 4-year-old girl with intraoccipital dermoid with associated dermal sinus tract. Preoperative MRI. Findings: Reformatted noncontrast axial 3D T1 images. The lesion is localized within the occipital diploic space (A), with an intracranial extradural component (B, short arrow), and a sinus tract traversing through the occipital bone to the overlying skin (B and C, long arrows). **Technique:** (A-C) Sequential images. MRI, 1.5 T, noncontrast MPRAGE 3D T1, axial plane, TR 14, TE 7, slice thickness 2 mm. MPRAGE, Magnetization-prepared rapid gradient-echo; MRI, magnetic resonance imaging; TE, echo time; TR, repetition time.

Discussion

We present a case of a rare intraosseous location of a dermoid cyst within the occiput with a subcutaneous sinus tract that was initially wrongly diagnosed as an occipital pyogenic granuloma and underwent unnecessary medical and surgical treatment that delayed the definite diagnosis and treatment.

Intracranial dermoid cyst accounts for 0.1%-0.7% of all intracranial tumors [1]. Dermoid cysts tend to manifest in the second and third decades of life [2]. Intracalvarial dermoid cysts are typically located in the anterior fontanelle [3]. The midline occipital bone represents a very rare location for dermoid cysts [4]. Intracalvarial dermoid cysts can become symptomatic when they become infected, which can result in abscess formation

or recurrent meningitis; cyst rupture can result in chemical aseptic meningitis; they can also be locally compressive [1]. They may become symptomatic even in infants and young children [2] like in the presented case, where it became symptomatic at 4 years of age.

Congenital dermal sinuses may present as a small cutaneous pit or a subcutaneous mass [5], they can arise anywhere in the midline of the body at any level of the craniospinal axis, most commonly involving the lumbosacral and occipital regions [5]. Occipital dermal sinus may terminate in the scalp, skull, extradural space, or within the cranial cavity [5]. Those that extend to the meninges may end in the subarachnoid space and be connected to epidermoids or dermoids in the posterior fossa [6]. The most frequent association of a dermal sinus of the occipital region is with a congenital dermoid or epidermoid that is generally situated intradurally [5].

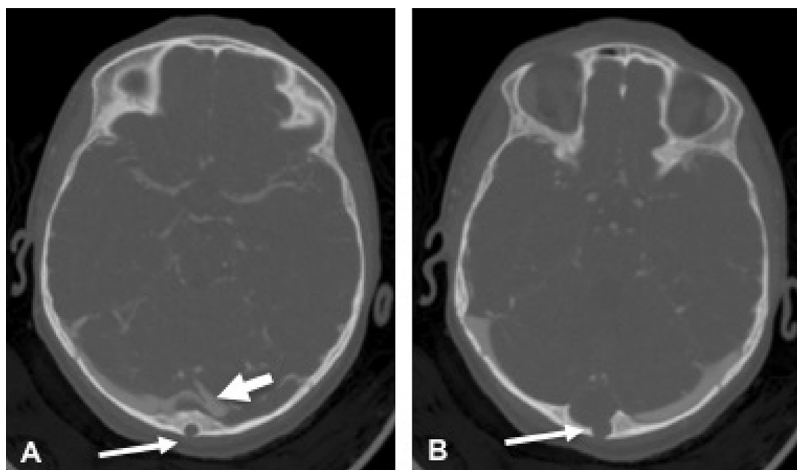


Fig. 4 – A 4-year-old girl with intraoccipital dermoid with associated dermal sinus tract. Preoperative CT angiogram. Findings: The lesion is located posterior to the torcular (A, short arrow). The sinus tract is noted as a round track with sclerotic borders (A and B, long arrows). **Technique:** (A) Contrast-enhanced CT angiogram, axial plane, 120 kVP, 240 mA, slice thickness 0.625 mm, contrast: 40-mL Isovue 370, 20-s delay. (B) Contrast-enhanced CT angiogram, sequential image, axial plane, 120 kVP, 240 mA, slice thickness 0.625 mm, contrast: 40 mL Isovue (Bracco Diagnostics Inc. 259 Prospect Plains Road, Building H Monroe Township, New Jersey 08831 USA) 370, 20-s delay. CT, computed tomography.



Fig. 5 – A 4-year-old girl, s/p excision of the intraoccipital dermoid with associated dermal sinus tract. Postoperative course was complicated by abscesses presenting with acute symptoms of slurred speech and ataxia required postoperative MRI. Findings: The patient’s postoperative course was complicated by right cerebellar abscesses. Postcontrast T1W demonstrates several adjacent rim-enhancing lesions (A, arrows) with corresponding hyperintense signal on DWI (B) and hypointense signal on ADC restricted diffusion (C). Surgical cavity is marked with an asterisk (A-C). Technique: (A) MRI, 1.5 T, postcontrast T1W, TR 9383, TE 5, slice thickness 2 mm. Contrast: 3.5 mL MultiHance. (B) MRI, 1.5 T, DWI, axial plane, eB1000, TR 6412, TE 82, slice thickness 4 mm. (C) MRI, 1.5 T, ADC, axial plane, TR 6412, TE 82, slice thickness 4 mm. ADC, apparent diffusion coefficient; DWI, diffusion weighted imaging; MRI, magnetic resonance imaging; TE, echo time; TR, repetition time.

Dermoid tumors are thought to arise in the early stages of embryonic development, between the third and fifth week of intrauterine life, probably due to faulty separation of the neuroectoderm and cutaneous ectoderm [6]. Dermoid cysts of the posterior fossa tend to lie in the midline of the skull, likely related to the development of the falx and tentorium which occurs as an invagination of 2 folds of dura and may draw in fragments of the cutaneous ectoderm [7]. Dermoid tumors frequently connect to the skin surface via a dermal sinus, which permits extrusion of cyst contents exterior to the scalp and poses a risk for bacterial infection [6].

Dermoid cysts may be completely asymptomatic or present as an incidental dermal sinus. At the same time, they can present with acute symptoms due to development of complications including: dermoid rupture with meningitis, epileptic seizures, increased intracranial pressure from dermoid cyst mass effect, bacterial infection and abscess formation [8]. Table 1 summarizes facts regarding skull dermoid cysts.

The MRI findings of dermoid cysts depend on the composition of the dermoid. In general, dermoids contain thick,

viscous, greenish-brown fluid composed of lipid metabolites and liquid cholesterol from decomposed epithelial cells, which are usually hyperintense on non-contrast T1 and heterogeneous on T2 [9]. On post-contrast imaging, dermoids may demonstrate peripheral enhancement related to the dermal components within their walls [2]. Use of diffusion weighted imaging for the diagnosis and follow-up of dermoids has not been well-described, only a few reports commented on presence of restricted diffusion in dermoids. In our case, MR and CT imaging played an important role in the diagnosis, preoperative localization of the dermoid cyst and to determine the relationship with adjacent veins and arteries [2]. Important facts about dermoids are summarized in Table 2.

Several differential considerations apply to dermoids (Table 2). Dermoids and epidermoids are often considered together as a diagnosis due to their similar etiology [2], but epidermoids usually demonstrate imaging appearance similar to CSF on CT and MRI [2]. For dermoid cysts with intrinsic high T1 signal differential diagnosis include intracranial lipoma. Intracranial lipomas generally demonstrate more uniform fat

Table 1 – Summary for skull dermoid cysts.

Etiology	Benign tumors resulting from defective closure of the neural tube with thick capsules lined by squamous epithelium which contains skin appendages
Incidence	Rare; 0.7%-1% of all intracranial tumors
Gender	Slight male predilection
Age	Usually manifest in the second to third decades although may manifest earlier
Treatment	En bloc resection
Prognosis	Generally good prognosis; no reported cases of recurrence following en bloc resection, although complications such as meningitis, cerebellar abscesses, hydrocephalus affect prognosis
Findings on CT	Unilocular, cystic, midline, central fat attenuation; intraosseous dermoids may cause expansion of the cortex with sclerotic remodeling of adjacent bone; +/- enhancement with contrast administration
Findings on MRI	Heterogeneously hyperintense T2W, occasionally hyperintense T1W, restricted diffusion on DWI and ADC map; capsule may demonstrate enhancement on postcontrast images due to dermal lining; if present, more likely to identify dermal sinus on MRI than on CT
ADC, apparent diffusion coefficient; CT, computed tomography; DWI, diffusion weighted imaging; MRI, magnetic resonance imaging.	

Table 2 – Differential considerations and imaging patterns.

	CT	MRI—T1, T2, DWI	Enhancement pattern	Other
Dermoid	Heterogeneous fat attenuation	T1: Heterogeneously hyperintense T2: Heterogeneously hyperintense DWI: Positive	May demonstrate rim enhancement	Nonuniform suppression on fat suppressed sequences; midline; may have associated dermal sinus
Epidermoid	Fluid attenuation similar to CSF	T1: Isointense to CSF T2: Isointense to CSF DWI: Positive	May demonstrate minimal rim enhancement	DWI (hyperintense to CSF) helps to differentiate from arachnoid cyst (isointense to CSF on DWI)
Teratoma	Heterogeneous, some regions of fat attenuation	T1: Hyperintense from fat T2: Soft tissue components iso- to hyperintense DWI: Positive in highly cellular parts	Soft tissue components enhance	Extra-axial masses in children usually found in pineal or suprasellar regions
Intracranial lipoma	Homogeneous fat attenuation	T1: Hyperintense T2: Hypointense on spin echo sequences DWI: No diffusion restriction	Does not enhance	Uniform suppression on fat suppressed sequences
Abscess	Thick-walled with central hypoattenuation +/- emphysema	T1 and T2: Depends on stage of abscess DWI: Restricted diffusion	Rim enhancement	Clinical presentation is important as dermoids may become superinfected

CSF, cerebrospinal fluid; CT, computed tomography; DWI, diffusion weighted imaging; MRI, magnetic resonance imaging.

attenuation on CT and more uniform hyperintense T1 signal on MR. On fat-suppressed MR sequences, lipomas also demonstrate more uniform suppression [2]. Intracranial teratomas may appear cystic with varying other tissue components, including fat, but they most commonly involve the pineal region or the suprasellar cistern [10]. Differential considerations for cystic lesions with restricted diffusion include intracranial abscess. Differentiation between dermoid and abscess should be made based on clinical history, as patients will generally show signs and symptoms of infection with an intracranial abscess. A classic triad would be fever, headache and focal neurologic deficits. However, it should be kept in mind that a dermoid cyst may and often does become superinfected, usually via the dermal sinus tract. In a published case series on cranial dermal sinus associated with extradural dermoids of the posterior fossa [6], histopathologic study showed preclinical signs of infection in 2 out of 3 patients that had not yet formed an abscess. Furthermore, in a recently published case series [8], posterior fossa abscess, hydrocephalus, and meningitis were observed in 38.9%, 38.9%, and 22.2% of children, respectively.

In our case, the patient developed cerebellar abscesses within 10 days of initial MRI and subsequently also developed hydrocephalus, another described complication of intracranial dermoids [11,12]. Therefore, our experience also reflects that of others and reinforces the need for early neurosurgical treatment of dermoids to prevent development of severe intracranial infection and its complications.

The prognosis is overall favorable. In a case series of 15 patients diagnosed with posterior fossa dermoid cysts, the overall outcome was excellent despite differences in timing of presentation and treatment [13]. As our case demonstrates, even in cases with postoperative complications, patients may still fully recover with little to no residual neurologic sequelae.

In summary, a child presenting with a dermal sinus and posterior fossa midline mass containing fat and demonstrating

restricted diffusion on diffusion weighted imaging should prompt consideration of an intracranial dermoid. Although dermoid cysts are benign lesions, early surgical resection is advised as these lesions may be complicated by infection. The clinical course of recurrent infections despite antibiotics should have raised suspicion for an associated dermoid with a sinus track.

REFERENCES

- [1] Layadi F, Louhab N, Lmejjati M, Aniba K, Ait Elqadi A, Ait Benali S. Cerebellar dermoid cyst with occipital dermal sinus. Report of two pediatric cases. *Pediatr Neurosurg* 2006;42(6):387–90. PMID 17047421.
- [2] Smirniotopoulos JG, Chiechi MV. Teratomas, dermoids, and epidermoids of the head and neck. *Radiographics* 1995;15(6):1437–55. PMID 8577967.
- [3] Ruge JR, Tomita T, Naidich TP, Hahn YS, McLone DG. Scalp and calvarial masses of infants and children. *Neurosurgery* 1988;22(6 Pt 1):1037–42. PMID 3419565.
- [4] Riebel T, David S, Thomale UW. Calvarial dermoids and epidermoids in infants and children: sonographic spectrum and follow-up. *Childs Nerv Syst* 2008;24(11):1327–32. doi:10.1007/s00381-008-0660-2. PMID 18560839. Epub 2008 Jun 17.
- [5] Wright RL. Congenital dermal sinuses. *Prog Neurol Surg* 1971;4:175–91.
- [6] Martínez-Lage JF, Ramos J, Puche A, Poza M. Extradural dermoid tumours of the posterior fossa. *Arch Dis Child* 1997;77(5):427–30. PMID 9487967. Review, PMCID: PMC1717376.
- [7] Logue V, Till K. Posterior fossa dermoid cysts with special reference to intracranial infection. *J Neurol Neurosurg Psychiatry* 1952;15:1–12.
- [8] Naderi S, Nejat F, Shahjoui S, El Khashab M. Cranial dermal sinus: presentation, complications and management. *Pediatr Neurosurg* 2012;48(2):86–92.

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- [9] Smith AS, Benson JE, Blaser SI, Mizushima A, Tarr RW, Bellon EM. Diagnosis of ruptured intracranial dermoid cyst: value MR over CT. *AJNR Am J Neuroradiol* 1991;12(1):175–80. PMID 1903577.
- [10] Smirniotopoulos JG, Rushing EJ, Mena H. Pineal region masses: differential diagnosis. *Radiographics* 1992;12(3):577–96. PMID 1609147. Review.
- [11] Akhaddar A, Jiddane M, Chakir N, El Hassani R, Moustarchid B, Bellakhdar F. Cerebellar abscesses secondary to occipital dermoid cyst with dermal sinus: case report. *Surg Neurol* 2002;3-4:266–70.
- [12] Cai CQ, Zhang QJ, Hu XL, Wang CX. Dermoid cyst of the posterior fossa associated with congenital dermal sinus in a child. *World J Pediatr* 2008;4(1):66–9.
- [13] Raghunath A, Indira Devi B, Bhat DI, Somanna S. Dermoid cysts of the posterior fossa—morbid associations of a benign lesion. *Br J Neurosurg* 2013;27(6):765–71. PMID 23530667.