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A Rare Presentation of Occipital Dermoid Cyst with Intracranial Extension and Secondary Infection: Case Report and Follow-Up

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Abstract

Craniofacial dermoid cysts are congenital anomalies that rarely have intracranial extension and can be associated with other anomalies. Common sites of these lesions are the lateral brow and parietal scalp. Presentation of the dermoid cyst in the occipital region with intracranial extension is extremely rare. We report a 2-year-old female with a presentation of an occipital dermoid cyst with intracranial extension and secondary cerebellar abscess. This case highlights the rarity of the presentation of an occipital dermoid cyst with intracranial extension and secondary infection and the importance of early imaging for suspected dermoid cysts in the occipital region for identification of intracranial extension.

Keywords
- occipital dermoid cyst
- pediatrics
- intracranial abscess
- case report

Introduction

Dermoid cysts (DCs) are characterized by a stratified squamous epithelial lining and differentiated from epidermal cysts by the inclusion of skin adnexa, such as hair and sebaceous glands, and less frequently, sweat glands, lymphoid tissue, and cartilage. Lateral brow DCs are the most common type of craniofacial DCs.[1] They usually present as slow-growing asymptomatic masses, located superficially with no deep extension, and can be managed by direct excisional biopsy. DCs with intracranial extension in the nasal and frontal regions have been widely reported in the literature.[2,3] But an occipital DC with intracranial extension is rare and reported in only nine children.[4–8] We report a rare presentation of an occipital DC with intracranial extension presenting as a cerebellar abscess with significant cerebellar edema and obstructive hydrocephalus. The patient had a successful outcome following surgical intervention and antimicrobial treatment.

Case Presentation

A 2-year-old female child presented to an outside hospital with 3 weeks of worsening ataxia, emesis, headache, and

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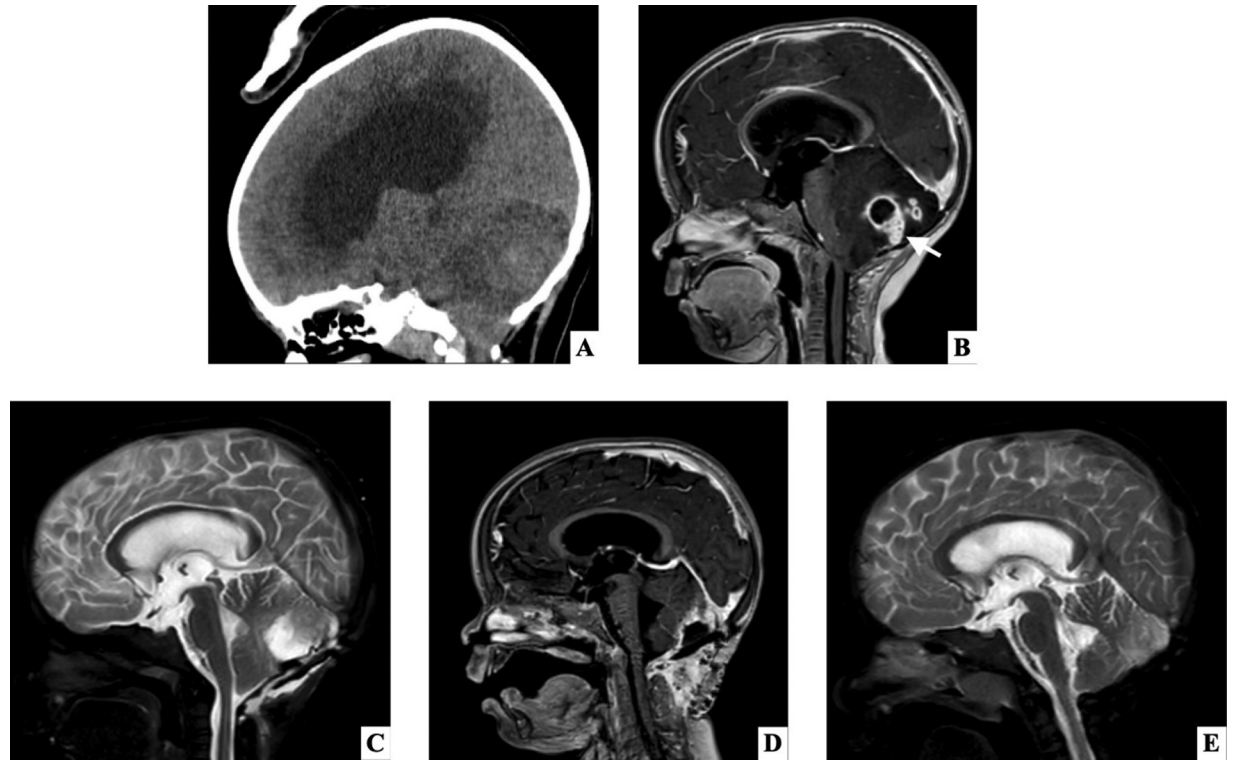

Fig. 1 Head computed tomography (CT) scan and brain magnetic resonance imaging (MRI) at the admission and follow-up. (**A**) Head CT scan demonstrated a posterior fossa mass and severe hydrocephalus. (**B**) At admission, brain MRI on sagittal T1-weighted sequence with contrast revealed a 3.7 × 2.6 cm multiloculated cystic mass (white arrow) in the right cerebellar hemisphere. (**C**) On postoperative day 7, brain MRI on sagittal T2-weighted sequence without contrast revealed significant interval improvement in cerebellar edema as well as transependymal seepage of cerebrospinal fluid (CSF). (**D**) Prior to discharge, brain MRI on sagittal T1-weighted sequence with contrast demonstrated increased leptomeningeal enhancement within the surgical bed of the posterior fossa. (**E**) At the 5-month follow-up, brain MRI on sagittal T2-weighted sequence without contrast showed there is no gross new parenchymal signal abnormality, there is no mass effect, midline shift, or basal cistern effacement, and the cerebellar tonsils terminate above the foramen magnum.

lethargy. Her head computed tomography (CT) scan (►**Fig. 1A**) demonstrated a posterior fossa mass and severe hydrocephalus. Based on these findings and neurologic deterioration, she was transferred to a tertiary care children's hospital for neurocritical care and neurosurgical management. In the emergency room, due to her altered mental status, she was intubated and subsequently underwent brain magnetic resonance imaging (MRI). The brain MRI revealed a 3.7 × 2.6 cm multiloculated cystic mass in the right cerebellar hemisphere, causing cerebellar edema and obstructive hydrocephalus (►**Fig. 1B**). On physical examination, she had a small cutaneous lesion with intermittent purulent drainage in the occipital region (►**Fig. 2**) which her mother stated that it had been present since 6 months of age. She was transferred to the pediatric intensive care unit for initial stabilization with aggressive medical management of elevated intracranial pressure. She underwent emergency placement of an externalized ventricular drain (EVD) and suboccipital craniectomy with resection of the infected dermal sinus tract and a ruptured DC with a large cerebellar abscess.

After surgery, she received vancomycin, which was then narrowed to oxacillin based on the growth of methicillin-sensitive *Staphylococcus aureus* from the cerebellar abscess. On postoperative day 7, she was extubated, and she was alert, awake, and oriented with no focal neurologic deficit with a

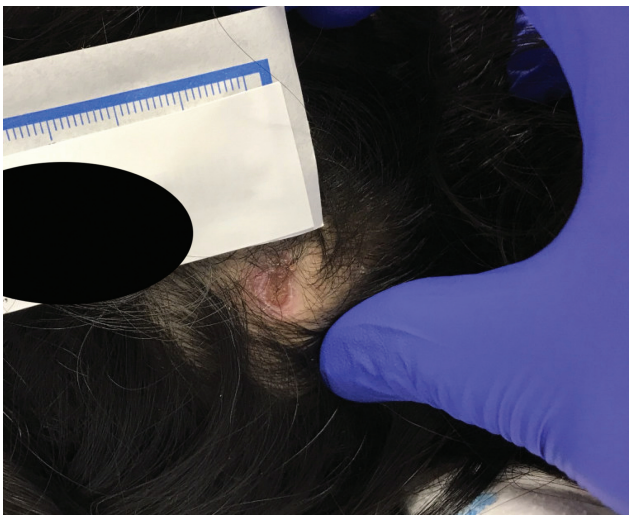

Fig. 2 A small, cutaneous lesion in the occipital region with intermittent purulent drainage since 6 months of age.

significant improvement in cerebellar edema on brain MRI (►**Fig. 1C**). On postoperative day 29, the EVD was removed. The patient completed a total of 5 weeks of antimicrobial treatment. Prior to discharge, brain MRI (►**Fig. 1D**) demonstrated decreased enhancement adjacent to the suboccipital craniotomy when compared with the previous study. Her

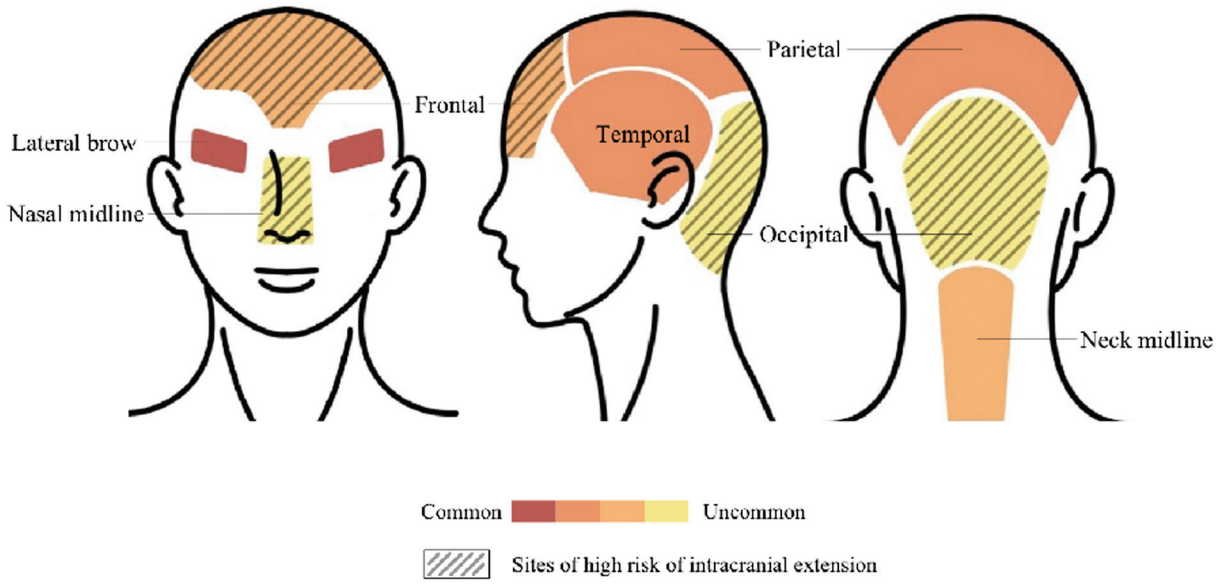

Fig. 3 Sites of craniofacial dermoid cysts.

auditory brainstem response monitoring revealed normal hearing acuity in both ears.

She was seen in the neurosurgery clinic 6 months after discharge. At that time, she was asymptomatic and there were no concerns for visual impairment. She had no neurological deficits on the physical examination. The brain MRI (►**Fig. 1E**) was reassuring and showed no new parenchymal signal abnormality, mass effect, midline shift, basal cistern effacement, or abnormal fluid collection.

Discussion

This case represents a rare presentation of an occipital DC in a pediatric patient with intracranial extension, secondary cerebellar abscess, and hydrocephalus successfully managed with surgery and antibiotic treatment.

Craniofacial DCs occur in subcutaneous tissues, especially on the face and about the eyes, and typically appear along lines of facial embryonic fusion. The most common site is the lateral brow (►**Fig. 3**). Uncommon sites include parietal scalp, temporal region, midline nasal, and occipital region.[1,3,9] Definitive diagnosis of a DC is important, particularly because DCs in certain locations have a higher risk of intracranial or intraspinal extension. Only midline nasal, frontal/anterior fontanelle, and occipital lesions have been associated with intracranial extension. In one series, 16.3% of occipital DCs had intracranial extension, and 71.4% of these lesions were identified in the midline.[3] Therefore, presurgical imaging (brain CT or brain MRI) is warranted in patients with occipital, frontal, and midline nasal DCs.

Occipital DCs are found in the center of the occipital bone in the midline, where there is a bony defect through which the dermal sinus extends intracranially. The natural history of these lesions is often asymptomatic. When symptoms are present, they include yellow discharge, pain on palpation, swelling, and pruritus.[1] A cutaneous dimple is present at the site of the lesion. In our case, the patient had a sinus tract at the site with frequent yellow discharge, and this sinus represented the route of entry for the *S. aureus* into the cerebellum. To our knowledge, this is the first reported pediatric case of an occipital DC with intracranial extension and secondary infection.

Imaging studies should be performed for cysts located along the midline, those associated with any cranial suture, or those with otherwise atypical presentations to delineate any intracranial or intraspinal extension.[10] The choice of treatment depends on the clinical status and age of the patient, as well as the size and location of the DCs. Due to the increased risk of epidural and cerebral expansion of DCs, brain MRI and surgical resection followed by systemic antibiotic therapy are indicated.[9] Positive outcomes without recurrence are observed in occipital DC after complete resection.[4–7]

In conclusion, this case demonstrates a rare presentation of an occipital DC with intracranial extension and secondary infection and the importance of early imaging for suspected DCs in the occipital region for identification of intracranial extension. Adequate preoperative studies are needed to identify any such extension before resection. Aggressive medical and surgical management of an infected, intracranial lesion is associated with successful outcomes.

Conflict of Interest

None declared.

References

1 Orozco-Covarrubias L, Lara-Carpio R, Saez-De-Ocariz M, Duran-McKinster C, Palacios-Lopez C, Ruiz-Maldonado R. Dermoid cysts: a report of 75 pediatric patients. Pediatr Dermatol 2013;30(06): 706–711

2 Hanikeri M, Waterhouse N, Kirkpatrick N, Peterson D, Macleod I. The management of midline transcranial nasal dermoid sinus cysts. Br J Plast Surg 2005;58(08):1043–1050

3 Overland J, Hall C, Holmes A, Burge J. Risk of intracranial extension of craniofacial dermoid cysts. Plast Reconstr Surg 2020;145(04): 779e–787e
4 Martens F, Ectors P, Noel P, Hanquinet S, Faverly D. Unusual cause of cerebellar abscess: occipital dermal sinus and dermoid cyst. Neuropediatrics 1987;18(02):107–109
5 Hayek G, Mercier P, Fournier HD, Menei P, Pouplard F, Guy G. Dermal sinus and dermoid cyst revealed by abscess formation in posterior fossa. Report of 2 pediatric cases and review of the literature [in French]. Neurochirurgie 2001;47(2-3 Pt 1):123–127
6 Karagöz Güzey F, Bas NS, Sencer A, et al. Posterior fossa dermoid cysts causing cerebellar abscesses. Pediatr Neurosurg 2007;43 (04):323–326
7 Ségbédji FKK, Tokpo AJ, Nubukpo-Guménu AA, et al. Infected intradural dermoid cyst with complete dermal sinus of posterior fossa. World Neurosurg 2018;116:219–224
8 Colamaria A, Blagia M, Sacco M, Iodice S, Carbone F. Infratentorial abscess secondary to dermal sinus associated with dermoid cyst in children: review of the literature and report of a rare case. Surg Neurol Int 2021;12:282
9 Khalid S, Ruge J. Considerations in the management of congenital cranial dermoid cysts. J Neurosurg Pediatr 2017;20(01): 30–34
10 Peter JC, Sinclair-Smith C, de Villiers JC. Midline dermal sinuses and cysts and their relationship to the central nervous system. Eur J Pediatr Surg 1991;1(02):73–79