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CLINICAL CASE REPORT

Giant dermoid cyst of the posterior fossa in a child

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Abstract

Introduction: Intracranial dermoid cysts are rare, congenital and, benign lesions. The etiology of these lesions is related to an embryonic defect during neurulation.
Case presentation: The present study describes a case of a 3-year-old girl with a giant cerebellar dermoid cyst, which initially manifested as hydrocephalus.
Discussion: We discuss its epidemiological characteristics as well as diagnostic and therapeutic management. The combination of high clinical suspicion, anamnesis, thorough physical examination, and adequate interpretation of neuroimaging data is crucial for the early diagnosis and timely therapeutic intervention for such cysts.
Conclusion: Surgical approach involving complete lesion resection considerably improves prognosis.

Key words: posterior fossa dermoid cyst; occipital dermal sinus; aseptic meningitis; pediatric neurosurgery; hydrocephalus; giant tumors

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Introduction

Intracranial dermoid cysts (DCs) are rare congenital lesions, accounting for less than 0.5% of primary intracranial tumors (1), and they are 4 to 9 times less common than epidermoid cysts (1). DCs are formed by the separation of the neuroectoderm during neurulation between the third and fifth weeks of embryonic development, leading to the sequestration of ectodermal remains (2,3). They usually occur at the midline of the sellar, parasellar, and frontonasal regions (1). However, they have also been found in the sylvian fissure and corpus callosum, at the cerebellopontine angle, as well as in the other regions of the posterior fossa (1,3,4). This study describes a rare case of a giant DC of the posterior fossa in a pediatric patient and provides a review of the related literature.

Case Report

A 3-year-old girl was admitted for medical care with chief complaints of headache, vomiting, neck pain, and prostration, which manifested one day prior to admission. Physical examination revealed rotatory nystagmus, right sixth nerve palsy, and occipital dermal sinus at the midline (Figure 1). The patient underwent brain magnetic resonance imaging (MRI), which revealed a large, expansive cerebellar lesion associated with supratentorial hydrocephalus (Figure 2). A cystic lesion with internal hair was identified perioperatively (Figure 3). Histopathology revealed a DC (Figure 4). The patient developed aseptic meningitis and remained hospitalized for 60 days. The patient was discharged with right sixth nerve palsy and a clinical picture compatible with akinetic mutism, which improved completely in 30 days.

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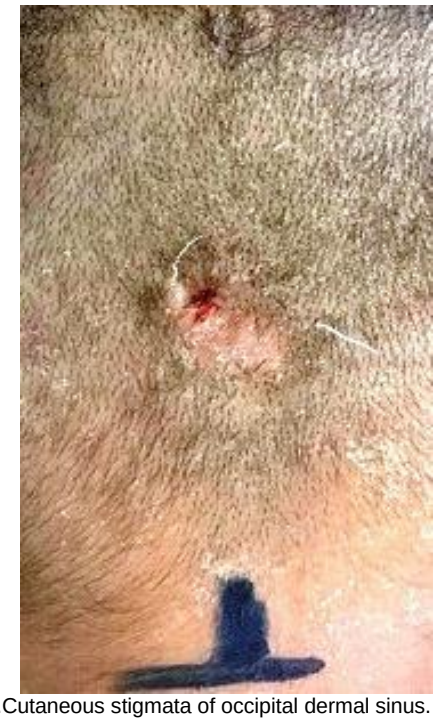

Fig.1.Cutaneous stigmata of occipital dermal sinus.

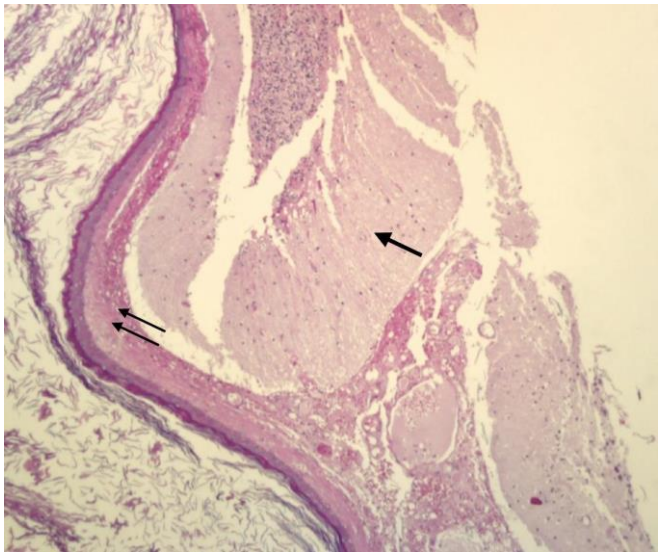

Fig 4. Microscopic view showing normal cerebellar parenchyma (Thick black arrow) limited by dermoid cyst wall (Double back arrows).

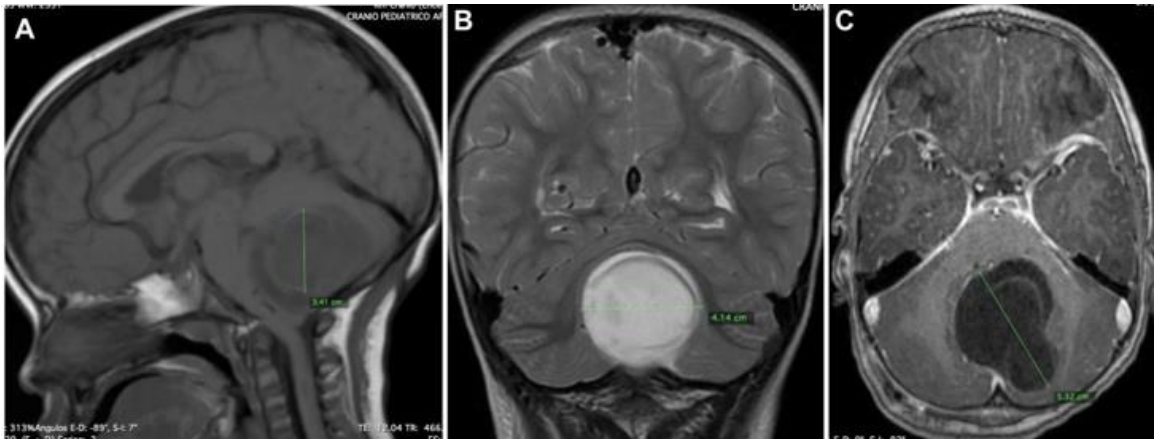

Fig. 2.Sagital T1-weighted MRI without contrast shows a heterogeneous lesion with iso/hypointense signal and 3,41cm of width(A); Coronal T2-weighted MRI with hyperintense signal and maximum diameter of 4,14cm(B) Axial T1-weighted MRI after contrast demonstrating 5,32cm lesion. None contrast enhancement was observed(C).

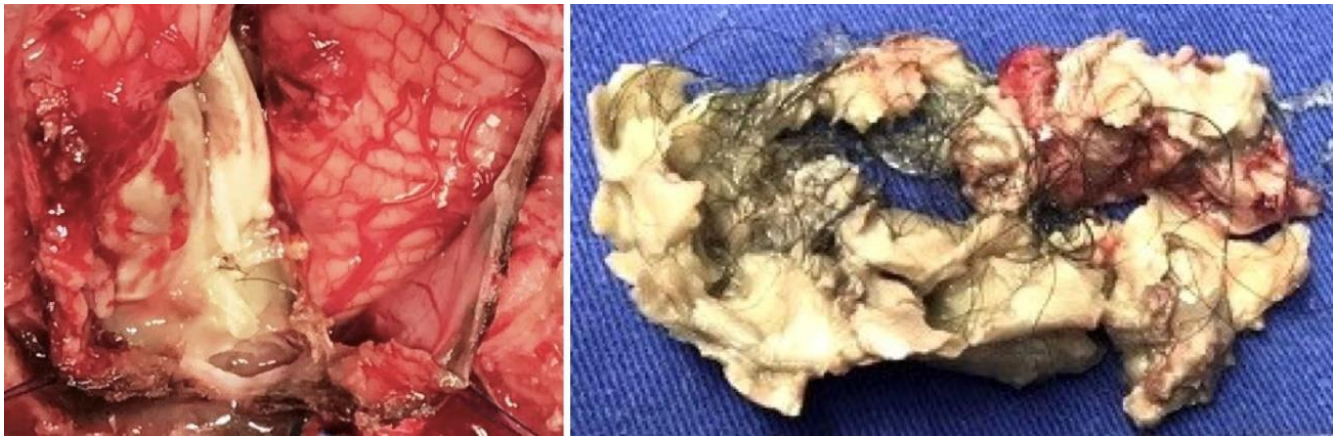

Fig. 3. Intraoperative view of the lesion in the magnum cisternal (Left) and after resection, amount of hair can be observed within the lesion (Right).

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Discussion

DCs are benign lesions primarily composed of squamous epithelium. They may contain cholesterol, keratin, sebaceous glands, hair follicles, hair, and occasionally teeth (1,5). These lesions develop slowly and are rarely diagnosed in children (6). Caldarelli et al. studied 16 patients with DCs, among whom 8 had lesions located in the posterior fossa, with the cisterna magna being the most frequently affected brain region (6). Another study involved 15 patients with DCs of the posterior fossa, among whom 10 were pediatric patients (age, 6 months to 11 years), and their average age at diagnosis was 4.8 years (7). Martínez-Lage et al. evaluated 25 patients with DCs of the posterior fossa and observed that DCs sized high than 3 cm were symptomatic (8). Although congenital, CDs are generally diagnosed in young adults. The development of this condition is extremely slow, and its clinical symptoms are related to mass effect, meningitis, hydrocephalus, or intracranial hypertension (5).

Dermal sinuses have been observed in 75% of patients diagnosed with DCs, which were usually located along a midline (dorsum of the nose, glabella, or external occipital protuberance) (6). Among these patients, 75% had signs of local infection and fever before hospital admission (6); the presence of these signs is associated with the development of meningitis in 50% of cases (9). Dermal sinuses were not found in patients with deep lesions such as in the brainstem, third ventricle, and quadrigeminal cistern. In these cases, clinical presentation is related to the mass effect of lesions (6).

Diagnostic imaging may be challenging because at early stages of lesion formation, MRI may present false-negative results (5). In addition, images may differ in cases of ruptured or unruptured cysts (1). In the latter case, T1-weighted images are hyperintense and not contrast enhanced (1,5); further, the signal intensity of their T2-weighted images is heterogeneous (1). The fat within ruptured DCs diffuses through the subarachnoid space and sulci as well as within the ventricles, and it is best observed on contrast-enhanced T1-weighted images (10). The content of ruptured DCs may cause aseptic meningitis associated with vasospasm, seizures, headache, hydrocephalus, and cerebral ischemia (11,12). These lesions may exhibit inflammatory meningeal enhancement (1).

DC treatment includes early complete surgical resection (13). This approach is necessary because growing lesions may adhere to delicate structures such as the brainstem, nerves, and blood vessels, which may increase the risk of sequelae (5). Chemotherapy and radiotherapy are not indicated for these tumors (6). Surgical resection must also be considered in asymptomatic patients (5).

Conclusion

Giant DCs of the posterior fossa are rare and infrequently reported in children. Their clinical presentation may be heterogeneous according to the location, evidence of rupture, and presence of a dermal sinus. MRI is the modality of choice for DC diagnosis and surgical planning. High clinical suspicion coupled with a relevant clinical history and physical examination as well as appropriate interpretation of neuroimaging data are crucial for the early diagnosis of DCs and timely therapeutic intervention. In this respect, the surgical approach involving complete lesion resection is considered the only effective treatment.

Statement of Ethics

The authors certify that the parents of child have given their written informed consent to publish their case.

Disclosure Statement

The authors have no conflicts of interest to declare.

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None.

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