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Occipital dermoid cyst associated with dermal sinus complicated with meningitis: A case report

Kyste dermoïde occipital avec sinus dermique compliqué d'une méningite : à propos d'un cas

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Summary

Background. Occipital dermal sinus, usually associated with dermoid cyst, is a rare entity; it results from the persistence of an abnormal embryonal communication between the skin and the intradural space. Its main complication is intracranial infection.

Case description. This 2-year-old girl was hospitalized for meningitis. Neuroradiological studies revealed a cystic mass of the posterior fossa communicating with the skin and hydrocephalus. The diagnosis of dermoid cyst associated with dermal sinus was established at surgery. The patient was treated with radical excision of both the occipital cyst and the dermal sinus associated with systemic antibiotic therapy. She had a good outcome.

Conclusion. Posterior fossa dermoid cyst should be considered in all children with chronic occipital skin lesion, especially a dermal sinus. We emphasize the importance of early neurosurgical treatment of dermoid cysts to prevent the development of severe complications.

Résumé

Introduction. Le sinus dermique occipital, fréquemment associé à un kyste dermoïde, est rare. Il résulte d'une communication anormale entre la peau et l'espace intra-dural dont la principale complication est l'infection du système nerveux central.

Observation. Cette fillette de 2 ans a été hospitalisée pour méningite purulente. L'exploration neuro-radiologique a révélé une masse kystique de la fosse postérieure communiquant avec la peau et une hydrocéphalie. Le diagnostic de kyste dermoïde associé à un sinus dermique a été établi en peropératoire. Une résection complète du kyste dermoïde avec son trajet fistuleux a pu être réalisée, associée à une antibiothérapie par voie systémique. L'évolution a été favorable.

Conclusion. Le kyste dermoïde de la fosse postérieure doit être évoqué chez tout enfant ayant une lésion cutanée chronique de la région occipitale et plus particulièrement un sinus dermique. Cette observation souligne l'importance de la précocité du traitement chirurgical des kystes dermoïdes pour prévenir la survenue de complications graves.

1. Introduction

Intracranial dermoid cyst (DC) is a rare benign congenital lesion. It occurs frequently in the posterior fossa and is usually associated with dermal sinus [1]. Embryologically, DCs occur between weeks 3 and 5 of gestation. They are thought to arise from a failure of cleavage between the ectoderm of the neuraxis and the cutaneous ectoderm, resulting in incorporation of the cutaneous ectoderm in the neural tube [2]. A dermal sinus develops when a cutaneous tract containing sebaceous materiel persists. Patients with posterior fossa DC associated with dermal sinus may develop hydrocephalus, aseptic or bacterial meningitis, or abscess formation. We report a case of occipital dermoid cyst associated with dermal sinus causing meningitis and obstructive hydrocephalus.

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2. Case report

This 2-year-old girl, the sixth child of unrelated parents, was admitted for acute fever complicated with prolonged generalized tonic-clonic seizure. She had a history of several consultations for a scalp-swelling lesion in the midline occipital region persisting since birth for which she received local treatment without any neuroradiological investigation. On physical examination, she had fever (38.5 °C), altered consciousness (Glasgow scale, 10/15), meningeal signs, tachypnea (76 cpm), tachycardia (140 bpm) with normal blood pressure (100/70 mmHg). Peripheral blood demonstrated leukocytosis (23,400/mm^3) with neutrophil count at 19,890/mm^3 and a high level of C-reactive protein (146 mg/dL). On biochemical analysis, serum sodium was 139 mEq/L, potassium 4.5 mEq/L, glucose level 4.5 mmol/L, and calcium level 2.53 mmol/L. Analysis of the cerebrospinal fluid (CSF) showed pleocytosis (1200 cells/mm^3) with predominance of polynuclear neutrophils (PNN 80%). The CSF glucose level was 0.19 g/L with $<$ 0.4 CSF glucose-to-serum glucose ratio. Culture of the CSF was negative.

A computed tomography (CT) scan of the brain showed a posterior fossa abscess complicated with hydrocephalus (*fig. 1*). Surgical exploration revealed a dermoid cyst and an occipital dermal sinus with a marked defect in the occipital skull vault. Surgical excision of both the cyst and the dermal sinus was performed. The cytopathologic analysis of the cyst revealed a dermoid cyst, containing hair and sebaceous glands. Microbiological culture of the cyst showed the presence of *Staphylococcus epidermidis*. Treatment with wide-spectrum antibiotics (cefotaxime + vancomycin) had been administered since admission and was maintained. Progression was marked by the appearance of active hydrocephalus with occurrence of nosocomial meningitis caused by *Acinetobacter baumannii*. Ventricular external drainage was performed until sterilization of the CSF, then a ventriculoperitoneal shunt was established. Two months after surgery, the patient was neurologically intact and developing well.

3. Discussion

Intracranial DCs are rare, accounting for 0.1–0.7% of all intracranial tumors [1–5]. They frequently occur in the posterior fossa, particularly in the midline position in the vermis or adjacent meninges, or in the cavity of the fourth ventricle [6]. They frequently communicate with the skin through a narrow tract lined with epithelium (dermal sinus), which can predispose to recurrent meningitis and other infections, possibly explaining why they are detected earlier during childhood [7]. Cranial dermal sinuses were first described by Ogle in 1865 [8]. They may occur in the midline anywhere from the nasion to the coccyx. The most frequent locations of these sinuses are the lumbosacral and occipital regions [3]. Dermal sinuses are often clinically occult. Their discovery is often fortuitous but can be secondary to the finding of a sluice of the occipital region, an angioma, or subcutaneous mass. Dermal sinuses can be revealed by several complications such as recurrent bacterial meningitis or cerebral abscess. *Staphylococcus aureus* is often implicated as the infecting pathogen, though infection with other pathogens has been described: *Escherichia coli* [9] and anaerobes [10]. Our patient exhibited sterile cerebrospinal fluid despite the isolation of *S. epidermidis* from the abscess and the patient's cerebrospinal fluid was typical of bacterial meningitis. This could be secondary to the previous antibiotic treatment. DCs can be responsible for recurrent aseptic or chemical meningitis, also called Mollaret meningitis, secondary to the rupture of the cyst into the meninges [11,12]. DCs can cause a mass effect by generating intracranial hypertension with or without hydrocephalus that may be due to a mass in the fourth ventricle or arachnoiditis secondary to repeated septic or aseptic meningitis [13].

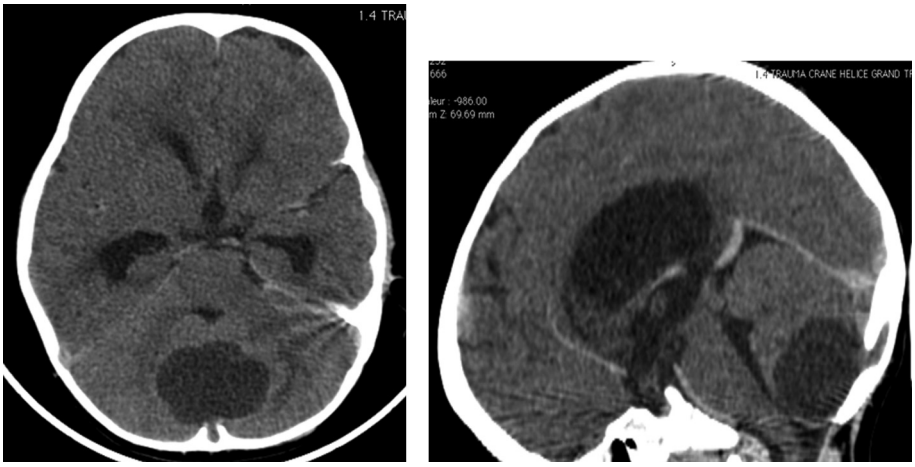

Figure 1. Computed tomography scan. Axial and sagittal contrast-enhanced images show a cystic mass in the posterior fossa midline, with discrete enhancement of the cystic walls. Note that the mass communicates with the skin.