



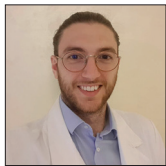
Case Report

Infratentorial abscess secondary to dermal sinus associated with dermoid cyst in children: Review of the literature and report of a rare case

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ABSTRACT

Background: Dermal sinus is usually located at either end of neural tube but most commonly lumbosacral. When occipital, it extends caudally and is mostly localized in the midline position or in the cavity of the fourth ventricle. It could communicate with the skin through a fistula with potential risk of deeper abscesses. Posterior fossa abscess secondary to dermal sinus associated with intracranial dermal cyst is an uncommon pathology.

Case Description: A 24-month-old girl was admitted to our institution with a cutaneous fistula in the midline of the occipital region. Brain imaging showed an infratentorial intradiploic cyst with peripheral enhancement to contrast medium. The mass showed hyperintensity on T1-weighted sequences, with the lower signal on T2-weighted images. A suboccipital craniotomy was performed with evacuation of the abscess and excision of the capsule. Contextually a 3 cm whitish and encapsulated cystic mass with hair component was extracted. Histology confirmed the diagnosis of abscess associated with dermal cyst and dermal sinus. The patient condition improved and 15 days after excision, was discharged. The postoperative MRI showed total removal of the lesion. A 36-month follow-up highlighted no evidence of recurrence.

Conclusion: Posterior fossa dermoid cyst should be considered in all children with a cutaneous fistula. Early neurosurgical treatment of these benign tumors should be performed to prevent the development of severe intracranial infection. Best results are associated with early diagnosis and complete removal of the abscess. The present work further reviews the few similar cases that have been reported in the literature confirming the need for future research.

Keywords: Dermoid cyst, Infratentorial abscess, Pediatric cerebellar abscess, Posterior fossa dermoid tumor

INTRODUCTION

We report an unusual case of posterior fossa abscess secondary to a dermal sinus associated with intracranial dermal cyst. Intracranial dermoid tumors are usually developmental, rare, and benign, with a linear growth rate. They represent between 0.1% and 0.7% of all intracranial tumors.^[8,19,25,35] Mostly occur in the posterior fossa, particularly in midline position, near the vermis or adjacent meninges, or in the cavity of the fourth ventricle.^[7,12,20] In the calvaria, dermoid cysts are often found in correspondence of the anterior fontanel and the occipital bone.^[21] More rarely, dermoid cysts may communicate with the skin through a narrow tract lined by epithelium

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(dermal sinus), which, therefore, contains the glandular architecture of skin, encouraging infection.^[1,16] These cysts have been reported to be associated with a higher risk of abscess formation.^[29,30] To the best of our knowledge, only 14 cases with posterior fossa dermoid cysts associated with cerebellar abscesses have been reported.^[9,10,13,17,20-22,30,33]

CASE REPORT

A 24-month-old female with a 2-month history of psychomotor retardation, weight loss, and unremarkable medical history was admitted to our institution. She presented horizontal nystagmus without sensory nor motor deficits. Physical examination revealed mild confusion and a cutaneous fistula in the mid-occipital region. The fistula appeared to communicate with intracranial space. Emergency CT scan [Figure 1; blue arrow] and brain MRI [Figure 2] showed a cystic infratentorial mass with ring enhancement of the cystic walls. In the suspect of intracranial abscess, treatment with ceftriaxone 75 mg/kg was administered every 24 h.

During preparation for surgery, a 5 mm non-purulent subcutaneous nodule with skin fistula was seen [Figure 3]. A suboccipital craniotomy was performed. The cerebellar abscess was evacuated, followed by excision of the entire capsule [Figure 4]. Total resection of a 3 cm whitish, midline,

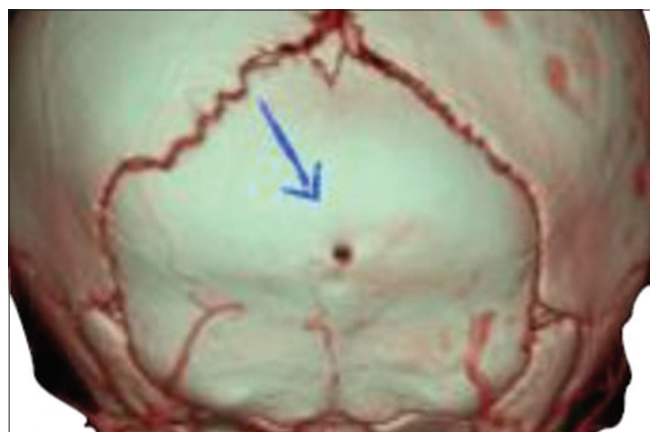


Figure 1: Emergency 3D-CT scan showing occipital fistula (blue arrow).

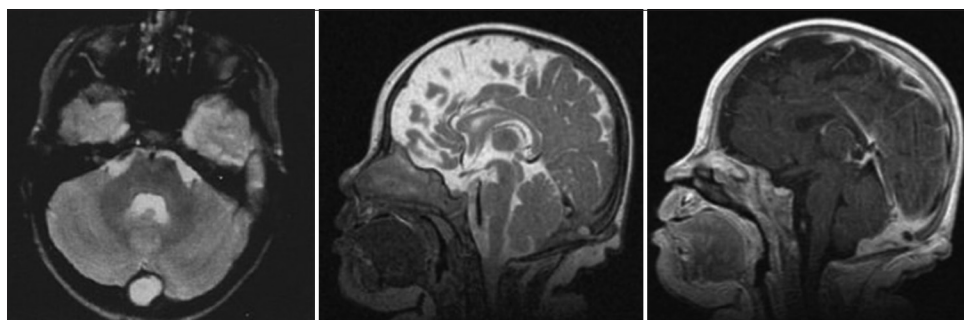


Figure 2: Mid-sagittal MRI showing a cystic subtentorial mass with ring enhancement of the cystic walls.

encapsulated, and hairy cystic mass was performed. The cyst was adherent to the dura and to the confluence of sinuses [Figure 5].

Bacterial investigation revealed methicillin-sensitive *Staphylococcus aureus*. Histological examination confirmed the diagnosis of dermoid tumor [Figure 6]. Therefore, antibiotic treatment was switched to Ampicillin IV 50 mg/kg every 6 h and was administered for 7 weeks after surgery.

The patient's physical condition and neurological symptoms improved rapidly. After 15 days, she was discharged without any deficit. At a 36-month follow-up, there was no evidence of recurrence [Figure 7].

DISCUSSION

Classification

Intracranial dermoid cyst represent a rare condition mostly recognized in the pediatric population with few cases described in the literature.^[2-14,16,17,20-33] Logue and Till^[15] have classified posterior fossa dermoid cysts into four groups based on the presence of extradural/intradural cyst and degree of development of the dermal sinus: (1) extradural dermoid cyst with a complete dermal sinus, (2) extradural dermoid cyst without a dermal sinus, (3) intradural dermoid cyst with an incomplete dermal sinus, and (4) intradural dermoid cyst with a complete dermal sinus. In their review of intracranial congenital dermal sinuses, French^[5] found that 85% were located near the external protuberance of the occipital bone, 11% in proximity of the nasion, and 5% in the posterior parietal area. About 89% of the reported cases were associated with inclusion tumors, mostly described as dermoid cysts except three classified as epidermoid cysts. Our case would be classified as a first group and was associated not only with dermoid sinus and cyst but also with a cerebellar abscess. This peculiar condition accounts for only a minority of reports, with abscess formation only described in 14 cases of intracranial dermoid cyst, with an estimated incidence of 14%.^[9,10,13,17,20-22,30,33]

Instrumental diagnosis

Dermoid cysts usually present as an oval or round defect on plain skull X-ray films. While they could be both small or moderate in diameter, rarely exceed 20 mm. Commonly reported are the sclerotic margins and the preferred midline location.^[13,27] Some cases are associated with vertebral deformities such as hemivertebrae, spina bifida, or Klippel-Feil deformity.^[24,28,31] Brain CT scan shows thinning or interruption of the compact bone of the skull and presence of calcification. It is not uncommon the presence of a homogeneous hypodensity of the cyst content even after contrast administration if the cyst is not infected, otherwise a peripheral enhancement is usually encountered.

Smith *et al.*^[27] have analyzed brain MRI in seven patients with ruptured dermoid cyst. In all reported cases there was a thick, viscous, greenish-brown fluid made of lipid metabolites and liquid cholesterol derived from decomposed

epithelial cell. This liquid component could therefore explain the hyperintensity on T1 sequences, with lower signal on T2.^[27] In the case of a superimposed infection a central



Figure 3: Preoperative evidence of non-purulent subcutaneous nodule with skin fistula.

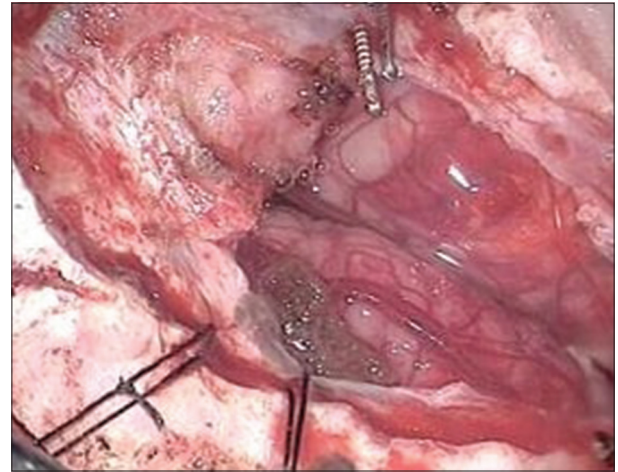


Figure 5: Evidence of cystic adhesion to dura and confluence of sinuses.

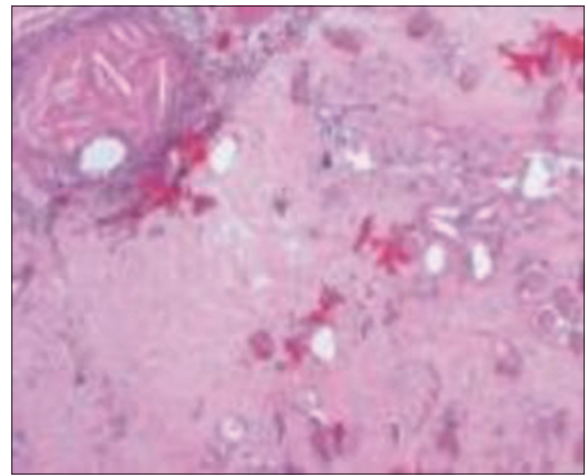


Figure 6: Histological examination confirmed the diagnosis of dermoid tumor.



Figure 4: Excision of the entire abscess capsule.

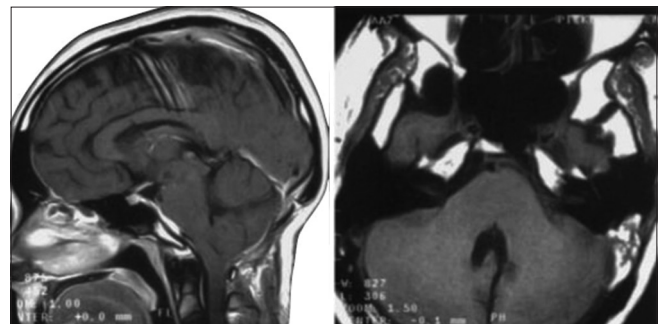


Figure 7: No signs of recurrence at 36 months follow-up evaluation.

hyperintense area can be seen.^[4,6,18,20] Fistulography through the dermal sinus should be avoided because of potential risk of infection and iatrogenic distention or rupture of the cyst.^[6] Cerebral angiography usually shows an avascular mass.^[34] In our case, the head CT showed the occipital skull interruption while the MRI demonstrated the peculiar oblique stalk that links cyst with the skin. After contrast administration a peripheral homogeneous enhancement was present.

Surgical treatment

Treatment of dermoid cysts consists in microsurgical excision and antibiotic therapy.^[8,10,18,34] A total removal of the cyst is not always possible because of firm adhesion to surrounding structures.^[1] Indeed a connection among the dermal sinus, dermoid cyst, and venous confluence can be present.^[26,32] The choice of treatment depends on the clinical status of the patient, as well as size, location, and presence or absence of the capsule. When hydrocephalus is present, external drainage is associated with more favorable operative outcomes.^[10,28,30] Recurrence of posterior fossa dermoid cyst after complete resection is virtually nonexistent.^[4,7,9,15,30] Only recently, Hashmi and Jones^[9] have reported recurrence of a cerebellar abscess 20 years after excision of dermoid cyst with bilateral abscesses.

CONCLUSION

Posterior fossa dermoid cyst should be suspected in children with an occipital skin lesion with concomitant dermal sinus. Neuroradiological investigation is necessary to define dermoid cyst site and any associated disorders. An early neurosurgical treatment should be performed to prevent the development of severe infections, such as bacterial meningitis and cerebellar abscess.^[4,7,9,10,15,30] Mortality and morbidity increase if meningitis develops. Better results were obtained in cases of early diagnosis and surgical treatment.^[30]

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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