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RESEARCH ARTICLE

CEREBELLAR ABSCESS SECONDARY TO DERMAL SINUS ASSOCIATED WITH DERMAL CYST IN CHILDREN: REVIEW OF THE LITERATURE AND REPORT OF A RARE CASE

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ABSTRACT

Introduction: Dermal sinus occur in the cranial vault, and the occipital region. Rarely, is localized in the posterior fossa, particularly 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 (0,1 to 0,7%).

Methods: A 24-month old girl was admitted to our institution with a cutaneus fistula in the midline of the occipital region. A CT scan and brain MR showed a sottotentorial intradiploic cyst with peripheral enhancement and edema. The mass was hyperintensity on T1-weighted sequences, with lower signal on T2-weighted images. A suboccipital craniotomy was performed with evacuation of the abscess and an excision of the visible capsule with a total removal of a 3 cm whitish, midline, encapsulated cystic mass with hair component.

Results: The histologic examination confirmed the diagnosis of abscess associated with dermal cyst and dermal sinus. Post-operative course was uneventful. The patient experienced a sudden improvement of cephalalgia and 15 days after the microsurgical excision, was discharged. On postoperative RM imaging, it was found the total removal of the lesion. A 36 month follow-up no evidence of recurrence.

Discussion: Posterior fossa dermoid cyst should be considered in all children with a cutaneus fistula. Early neurosurgical treatment of these benign tumours should be performed to prevent the development of severe intracranial infection. Best results were obtained in cases of early diagnosis and the complete removal of the abscess. The reported case passes through a review of similar cases riported in the literature confirm the rarety of the case report.

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INTRODUCTION

The authors present an unusual case of posterior fossa abscess secondary to a dermal sinus associated with intracranial dermal cyst. Intracranial dermoid cysts are rare (0.1 to 0.7% of all intracranial tumors) (Guidetti, 1977; Lunardi *et al.*, 2002; Roberts, 1958; Yasargil, 19899). They mostly occur in the posterior fossa, particularly in the midline position in the vermis or adjacent meninges, or in the cavity of the fourth ventricle (Groen *et al.*, 1995; Higashi *et al.*, 1995; Logue, 1952). In the cranial vault, dermoid cysts are often found at the anterior fontanel and the occipital region (Martinez-Lage *et al.*, 1997). More rarely, dermoid cysts may communicate with the skin through a narrow tract lined by epithelium (dermal sinus), which, therefore, contains the glandular architecture of skin, encouraging infection (Akhaddar *et al.*, 1999; Logue, 1952).

These cysts rarely cause abscess formation or formation of daughter abscesses within the cerebellum (Smith, 1959; Starinsky et al., 1988). At present are reported only 16 cases with posterior fossa dermoid cysts causing cerebellar abscesses (Akhaddar et al., 1999; Erdem et al., 1994; Goffin et al., 1993; Groen et al., 1995; Guidetti et al., 1977; Hashmi et al., 1998; Hayek et al., 2001; Higashi et al.,1995; Hsu et al., 1998; Lepindre, 1970; Logue, 1952; Lunardi et al., 2002; Martens et al., 1987; Martin et al., 1943; Martinez-Lage et al., 1992; Martinez-Lage, 1997; Matson, 1951; Roberts, 1958; Rubin, 1989; Schijman, 1986; Smith, 1989; Tekkok, 1996; Tytus, 1956; Vinchon, 1995; Wiemer, 1988; Yasargil, 1989; Wright, 1971; French, 1990; Guidetti, 1977; Matson, 1969; Davidson et al., 1985; Smith et al., 1991).

Case report

A 24-month old girl with a 2-month history of psychomotor retardation signs and weight loss with unremarkable medical history was admitted to our institution.

Physical examination revealed mild confusion. She present horizontal nystagmus without sensory nor motor deficit. The lesion appeared to communicate with the skin and intracranial space. Emergency CT scan (Fig.1; blue arrow) and brain MR (Fig. 2) showed a cystic subtentorial mass with ring enhancement of the cystic walls. During surgery, a 5 mm small subcutaneous nodule was seen with a small skin fistula opening without pus (Fig. 3) A suboccipital craniotomy was performed.

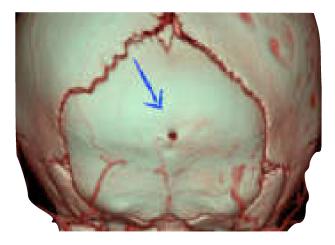
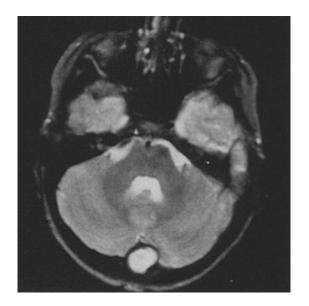


Fig. 1: Emergency head CT scan showing point of communication (blue arrow) between dermoid cyst and skin through the dermal sinus



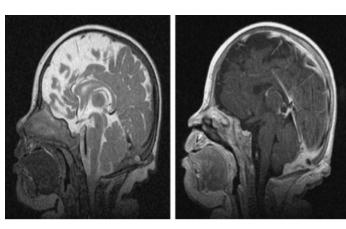


Fig. 2. brain MR showing cystic subtentorial mass



Fig. 3. Skin fistula

The cerebellar abscess was evacuated, followed by excision of the entire capsule (Fig.4, 5). Total removal of a 3 cm whitish, midline, encapsulated with hair cystic mass was carried out. The cyst was adherent to the dura and to the torculary venous sinus (Fig. 6, 7).

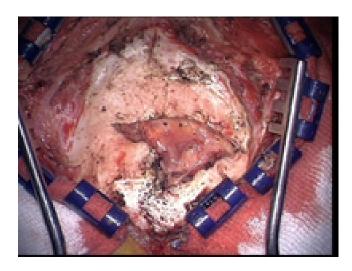


Fig. 4. Intraoperative image showing epicranial fistula identificationn

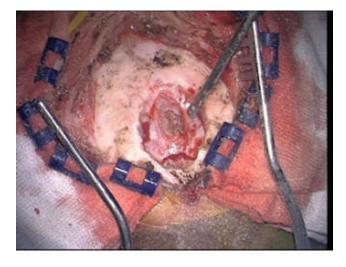


Fig. 5. Intraoperative image showing epicranial fistula isolation

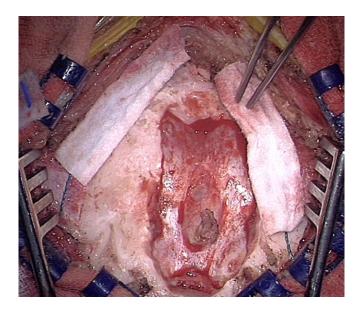


Fig. 6. Intraoperative image showing dural communication

Bacterial investigation revealed Staphylococcus aureus. Histological examination confirmed the diagnosis of dermoid tumor (Fig. 8, 9, 10, 11). The patient's general physical condition and neurological symptoms improved rapidly. After 15 days, she was discharged without any deficit. After a 36 month follow-up, there is no evidence of recurrence (Fig. 12).

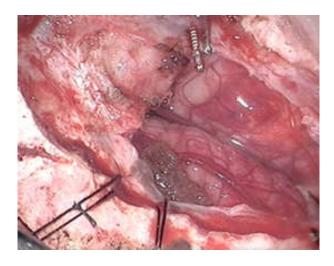


Fig. 7. Complete excission of the fistula

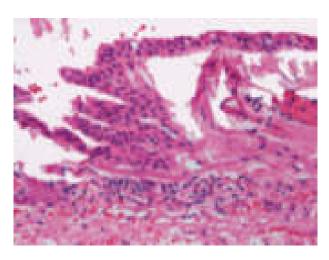


Fig. 8.

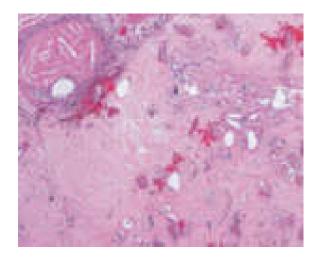


Fig. 9.

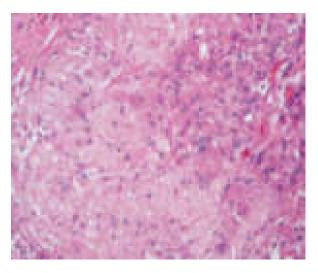


Fig. 10.

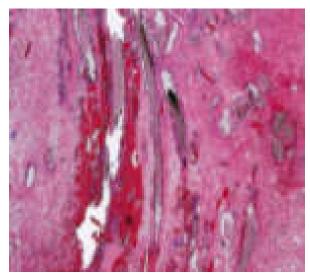


Fig. 11.

Fig. 8-9-10-11. histological examination

DISCUSSION

Classification

Logue and Till (1952) 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

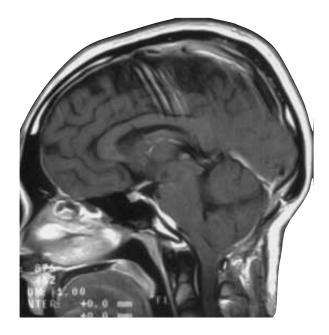




Fig. 12. Post-operative head MR with no evidence of recurrence

dermal sinus, (2) intradural 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. French (1977), reviewing the cases of intracranial congenital dermal sinuses, found that 85% of the dermal sinuses were located near the external protuberance of the occipital bone, 11% at the nasion, and 5% in the posterior parietal area. 89% of the reported cases were associated with inclusion tumors and that tumors were dermoid cysts except three classified as epidermoid cysts. Our case comes under the Logue e Till first group and was associated not only with dermoid sinus and cyst but also with a cerebellar abscess.

Strumental diagnosis

Dermoid cyst, at plain skull films often it present as an oval or circular defect, small or moderate in size (less than 20 mm in diameter) with a sclerotic margin, localized along the midline, just below the inion. If it is very small, it could not be visible on plain radiographs (Davidson, 1985; Smith, 1991). Some cases are associated with vertebral deformities such as hemivertebrae, spina bifida, or Klippel-Feil deformity (Roberts, 1958; Smith, 1959; Vinchon, 1995).

Brain CT scan detects thinning or interruption of the skull tables, and presence of calcification and can reveal the homogeneus hypodensity of the cyst content; after contrast administration, if the cyst is infected, an homogeneus periferical enhancement is present. Smith *et al.* (1985) have analyzed the brain MR in seven patients with ruptured dermoid cyst. In all reported cases there was a thick, viscous, greenish-brown fluid composed of lipid metabolites and liquid cholesterol from decomposed epithelial cell; these liquid product can explain an hyperintensity on T1 sequences, with lower signal on T2 (Matson, 1969; Davidson *et al.*, 1985; Smith *et al.*, 1991).

If the dermoid cyst is infected, the density values in the central area of the lesion are higher (Erdem et al., 1994; Goffin et al., 1993; Lunardi et al., 2002; Martinez-Lage et al., 1997). Fistulography through the dermal sinus should be avoided because of the potential risk of infection and iatrogenic distention, or rupture of the cyst (Goffin et al., 1993). Cerebral angiography usually shows an avascular mass (Yasargil, 1989). In our case, the head CT showed the occipital skull interruption while the MRI demonstrated the tipically oblique stalk or tract that links cyst and skin and after contrast administration a periferical homogeneus ovalar enhancement was present.

Surgical treatment

Treatment of dermoid cysts consists in micro-surgical excision and antibiotic therapy (Guidetti, 1977; Hayek et al., 2001; Lunardi et al., 2002; Yasargil et al., 1989). A total removal of the cyst is not always possible because of firm adhesion of its (Akhaddar, 1999). Indeed a connection among the dermal sinus, dermoid cyst, and venous confluence can be present (Schijman et al., 1986; Wiemer, 1988). The choice of treatment depends on the clinical status of the patient, as well as n the size, location, and the presence or absence of the capsule. When hydrocephalus is present, external drainage promotes more favorable operative conditions and perhaps decreases the likelihood of permanent CSF diversion (Hayek et al., 2001; Smith, 1959; Tekkok et al., 1996). For the presence of a communication between external and intracranial space due to the dermal sinus, we did an emergency intervention and a local and systemic antibiotic therapy was initiated.

Conclusion

Posterior fossa dermoid cyst should be suspected in children with an occipital skin lesion in presence of a dermal sinus. Neuroradiological investigations are necessary to define the 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 of the cyst (Erdem, 1994; Groen et al., 1995; Hashmi, 1998; Hayek et al., 2001; Logue et al., 1952; Tekkok et al., 1996). No recurrence of a posterior fossa dermoid cyst after surgery has been reported n the literature (Erdem et al., 1994; Groen et al., 1995; Hashmi et al., 1998; Logue, 1952; Tekkok et al., 1996). Only recently, Hashimi and Jones (Hashmi, 1998) have reported recurrence of a cerebellar abscess 20 years after excision of a dermoid cyst wit bilateral abscesses. Mortality and morbidity increase if meningitis develops. Better results were obtained in cases of early diagnosis and surgical treatment (Tekkok et al., 1996).

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