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CLINICAL CASE REPORT

Recurrent cerebellar abscess secondary to cranial dermal sinus associated with dermoid cyst in children

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Abstract

Background: Posterior fossa dermoid cysts are rare, benign lesions whose diagnosis can be quite challenging because of their slow growth and subsequent paucity of symptoms. We present herein an unusual case of recurrent cerebellar abscesses induced by an adjacent extradural dermoid cyst with a complete occipital dermal sinus.
Methods: The authors report the case of a 20-month-old girl who presented with signs of acutely raised intracranial pressure and whose head scans showed a left cerebellar hemisphere abscess associated with obstructive hydrocephalus. The patient was treated initially with an external ventricular drain, followed by burr-hole aspiration of the abscess and long-term antibiotics. Since the cerebellar abscess recurred, a posterior fossa craniotomy was performed and gross total resection of the lesion along with the dermal sinus tract and abscess contents was achieved. Histopathological analysis confirmed a dermoid tumor.
Conclusions: The occurrence of recurrent cerebellar abscesses must always rise up the suspicion of an associated dermoid cyst. Neuroradiological scans should be carefully evaluated in search for this lesion. Once the diagnosis is established, radical resection of the cyst, sinus tract and infectious components is the treatment of choice.

Key words: Cerebellar abscess, Dermal sinus tract, Dermoid cyst, Posterior fossa.

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Background

General overview

Intracranial dermoid cysts are benign tumors that arise from the inclusion of ectodermal components within the neural tube during the third and fifth week of embryonic development, when the neural fold closes [1, 2, 7]. Their slow growth results from progressive epithelial desquamation and gland secretion within the cyst [1]. In this sense, dermoid cysts can be difficult to diagnose, since the clinical features are usually very mild. They are also extremely rare in children, with very few cases reported in the medical literature [1, 21-30].

However, especially when associated with dermal sinus tracts, they are a predisposing factor for infections [7]. Therefore, patients with recurrent meningitis or abscesses should always undergo a

careful radiological assessment in search for these lesions. This is particularly important for children with posterior fossa abscesses, provided that they hardly ever occur spontaneously.

We conducted a literature review of eighteen pediatric cases of occipital dermal sinus tracts associated with cerebellar abscesses and posterior fossa dermoid cysts that were confirmed histologically (Table 1). The clinical and radiological presentation varied substantially. Herein, we summarize the literature data and present an unusual clinical case of a recurrent cerebellar abscess associated with a cranial dermal sinus.

Pathophysiology and clinical features

Overall, intracranial dermoid cysts are very rare, accounting for 0.1 to 0.7% of all intracranial tumors [1, 14]. When located in the posterior fossa, they occur most often in the midline, around the vermis, or inside the fourth ventricle [13, 19, 28]. They are the result of inclusion of ectodermal elements into the neural tube before its closure, and thus are frequently associated with a dermal sinus tract [28]. These tracts are the primary manifestation of the defective closure, and usually terminate in connective tissue, but sometimes they might extend into the craniospinal axis [3, 15, 19].

Cranial dermal sinus tracts can be situated anywhere in the cranial vault, but are commonest in the occipital region [1, 6, 28]. They communicate with the dermoid through a small pinhole in the occipital bone [28]. The inner walls of both the sinus tract and the dermoid cyst are overlaid by dysplastic epithelium, whose structure resembles skin and contains sebaceous and mucous glands [13], and whose secretions will ultimately lead to its growth in size.

With regards to surgical implications, Logue and Till [13] studied 32 cases (including adults) published in the literature and divided posterior fossa dermoid cysts into four groups depending on their anatomical situation (intra or extradural) and on the degree of development of the dermal sinus, whether absent, partial, or complete: (1) extradural dermoid cyst with a complete sinus, (3) intradural dermoid cyst without a dermal sinus, (3) an intradural dermoid cyst with an incomplete dermal sinus, and (4) intradural dermoid with a complete dermal sinus. Clearly, a dermal sinus tract communicating the inner aspect of the cyst and the skin would provide direct access for bacteria to enter the intracranial compartment and therefore generate infections such as meningitis, abscedation of the dermoid cyst itself or surrounding abscesses within the cerebellum [1].

According to Martens et al [19], the onset of closed posterior fossa dermoids occurs usually between the second and the fifth decade of life, when the cyst has increased in size considerably. Signs and symptoms, thereby, are similar to other posterior fossa tumors, such as headaches, vomiting, gait disturbances and dysmetria [28]. In younger children, a small dimple appears on the skin of the occipital region, and might be ignored if there is hair coverage. If infection develops, the dimple may become red and swollen, and the patient will present with typical infectious signs, like pyrexia, malaise, and neck stiffness. Since meningitis is the most frequent presentation, these signs will help differentiate dermoids from other posterior fossa tumors [1, 21].

Radiological studies are very accurate in confirming the diagnosis of dermoid cyst. Plain skull X-rays frequently demonstrate the bony defect, which can be oval or circular with a sclerotic margin just below the inion [1]. Obviously, small tracts might not be visible. Computerized tomography (CT) scans often reveal precisely the location of the cyst, and the extent of bony abnormalities and calcifications, if present. Dermoids typically appear on CT as non-enhancing, round, homogeneously hypodense cysts. It can be very difficult to confirm the presence of an associated infection, even though destruction of the diploe seems to be a very specific finding [13,14].

Magnetic Resonance Imaging (MRI) scans provide optimal visualization of the cyst and increase diagnostic sensitivity [13]. Curiously, few cases in the literature have been studied with MRI [1]. Dermoid tumors exhibit high signal on T_1 and T_2 weighted images and low intensity on STIR images [19]. MRI scans give full anatomic details of the lesion, including its relationship to surrounding structures, and the characteristic oblique dermal sinus tract can be visualized [28]. Increased signal or extended pericystic edema might indicate an associated infection [28]. Any type of fistulography should be avoided because of the risks of further introducing infectious agents and iatrogenic distension or rupture of the infected cyst [1].

Management

Surgical excision of the dermoid cyst, the dermal sinus and associated components is the treatment of choice [1, 28]. It should be performed as soon as possible to confirm the histopathological diagnosis but mainly to avoid the occurrence of secondary infections and abscess formation, as well as to reduce mass effect [3, 15, 28].

The goal of the operation should be total excision of the dermoid and associated sinus tract components [28]. However, complete resection is not always possible due to strong adhesions of the tumor capsule to surrounding structures; also, the tract might be connected to the posterior venous sinuses, such as the torcula or occipital sinus, and major bleeding needs to be anticipated [16, 17].

Staphylococcus aureus is often implicated as the infecting pathogen, though infection with other pathogens has been described. In one review of the literature, *Staphylococcus aureus* was isolated from

Table 1 - Summary of cases of cerebellar abscesses associated with dermoid tumors of the posterior fossa and occipital dermal sinuses

M=male; F=female; m=months; yrs=years; CT=computerized tomography; MRI=magnetic resonance imaging; US= ultrasonography; NA= not available

Authors & year/reference no.	Age/ Sex	Clinical presentation	Scans	Agent	Outcome
Groen & van Ouwerkerk 1995 [8]	7.5 m/F	hydrocephalus, acutely raised intracranial pressure	CT / MRI	*Staphylococcus aureus*	Good
Martínez-Lage et al. 1997 [20]	27m /F	progressively enlarging occipital mass	CT	*Staphylococcus aureus*	Good
Akhaddar et al. 2001 [1]	14m /M	acutely raised intracranial pressure, seizures, meningitis	CT	*Staphylococcus aureus*	Good
Hayek et al. 2001[10]	18m /F	acutely raised intracranial pressure	CT	NA	Good
Hayek et al. 2001[10]	2yrs /F	hydrocephalus, acutely raised intracranial pressure	CT / MRI	NA	Good
Costa et al. 2004[4]	12m /F	fever, vomiting, irritability	US / CT / MRI	*Staphylococcus aureus*	Good
Youklif at al. 2006 [29]	13y/ F	headache and vomiting	MRI	*Staphylococcus aureus*	Good
Cai et al. 2008 [2]	2yrs /F	headache and vomiting	CT / MRI	*Staphylococcus epidermidis*	Good
Douvoyiannis et al. 2008 [5]	18m /M	lethargy and meningitis	MRI	*Propionibacterium acnes*, *Prevotella loescheii*, and other mixed anaerobic species not further identified	Good
Yang et al. 2008 [28]	26m /F	headache and vomiting	CT	Sterile	Good
Yang et al. 2008 [28]	19m /M	headache and vomiting	CT	*Staphylococcus aureus*	Good
Yang et al. 2008 [28]	30m /F	headache and vomiting	CT / MRI	*Klebsiella pneumoniae*	Good
Yang et al. 2008 [28]	18m /F	headache and vomiting	CT / MRI	Sterile	Good
Karagöz Güzey et al. 2007 [13]	22m /M	fever, inability to hold head and walk, vomiting	CT / MRI	*Streptococcus* species	Good
Karagöz Güzey et al. 2007 [13]	14m /F	ataxia, vomiting	CT / MRI	Sterile	Good
Mann et al. 2009 [18]	20m /F	fever, vomiting, irritability	CT / MRI	*Propionibacterium avidum*, *Peptostreptococcus*, *Enterococcus faecalis*	Good
Ramzan et al. 2011 [22]	5yrs /F	discharge from dermal sinus, torticollis, cerebellar signs	CT / MR	*Staphylococcus epidermidis*	Good
García Galera et al. 2013 [6]	18m /F	occipital lump, fever, vomiting, irritability	MRI	Sterile	Good

the abscess, cerebrospinal fluid, or drainage fluid in 64% (9 of 14) of the patients [18].

There remains some controversy whether surgery should be performed in the presence of cyst infections or surrounding abscesses. Some authors advocate primary total removal including the infectious components [18]; others recommend treatment of the infection with systemic antibiotics and repeated abscess aspirations, if necessary, so as to perform tumor resection with the patient in a more stable clinical condition.

Yang at al. [28], report two cases in which this management strategy was adopted, with excellent results.

Two rare clinical conditions have been reported in the literature in association with posterior fossa dermoid tumors and occipital sinus tracts [19, 20, 21]. The first one is Mollaret (or aseptic) meningitis, which has been hypothesized to occur due to episodic releases of aseptic dermoid material into cerebrospinal fluid spaces [19]. Also, Klippel-Feil anomaly has been linked to posterior fossa dermoids, since the development of this lesion is believed to hamper the intrauterine movements of the cervical spine, which is quintessential for its normal anatomy [20]. Therefore, this hindrance may give rise to Klippel-Feil anomalies and possibly various other malformations of the craniovertebral junction [21].

Case illustration

A 20-month-old girl was admitted to our emergency unit with a 5-month history of vomiting, fever and headache and a cluster of generalized tonic-clonic seizures that had started one day before admission. She was, at that time, in deep coma and unresponsive to painful stimuli.

An emergency CT scan showed a 2.4 X 2.5 X 3.2 cm cystic mass in the left cerebellar hemisphere with enhancement of the cystic walls alongside some edema and a multiloculated mass on the cerebellar vermis. This lesion was compressing the fourth ventricle rendering triventricular obstructive hydrocephalus. Due to the signs of raised intracranial pressure,an external ventricular drainage was inserted.

After clinical stabilization of the child a posterior fossa burr-hole for primary aspiration of the cerebellar abscess was initially performed. Microbiological studies of the aspirated pus revealed oxacillin-sensitive *Staphylococcus aureus*, so the patient received oxacillin for 4 weeks.

In the 23rd day of the antibiotic therapy the patient become drowsy and lethargic. Repeated MRI confirmed recurrence of the cerebellar abscess. A second aspiration was performed and the course of oxacillin was extended to six weeks.

Control CT scans showed a multiloculated abscess. Further investigation with MRI revealed a mass in the cerebellar vermis with a dermal sinus tract from the lesion to the skin and an occipital bony calvarium defect (Fig. 1).

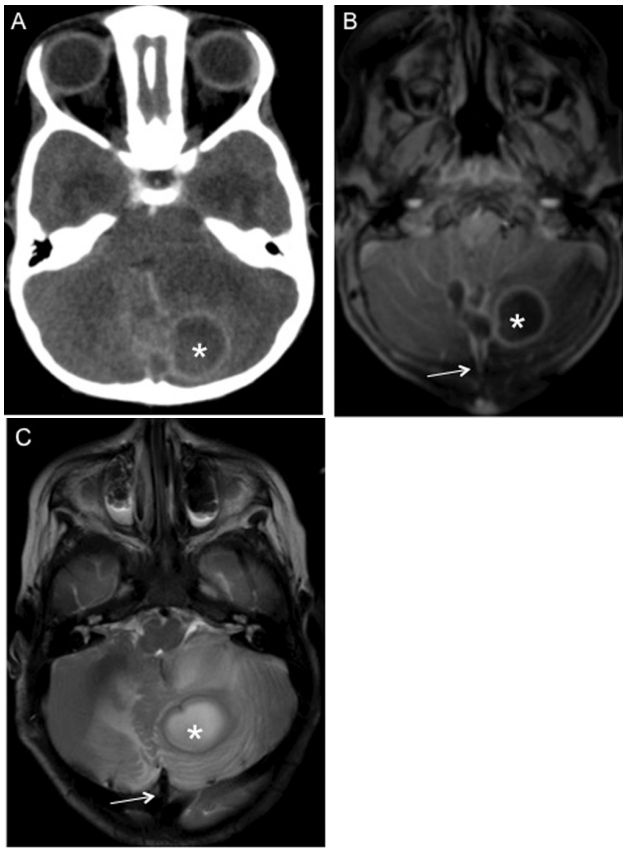

Figure 1 - Cerebellar abscess in a child associated with a cranial dermal sinus. **a** Preoperative CT scan after contrast showing a left-hemisphere cerebellar abscess dislodging the brainstem and fourth ventricle (*) **b** Axial T_1-weighted after gadolinium and **c** axial T_2-weighted magnetic resonance (MR) images showing a space-occupying lesion (asterisks) associated with cerebellar edema and an occipital dermal sinus tract (arrows).

A meticulous clinical examination of the occipital region after shaving revealed a pinhole on the skin of the occipital region at the level of the external occipital protuberance. A midline suboccipital craniotomy was performed. Total removal of a 4 cm whitish, midline, encapsulated cystic mass with hair and purulent contents was observed. The dermal sinus tract was also excised (Fig. 2). The multiloculated cerebellar abscesses were evacuated

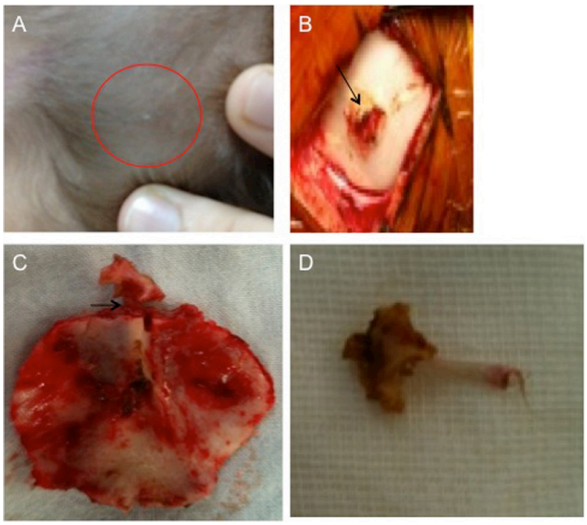

Figure 2. A Photograph depicting a small skin pinhole over the occipital region, in keeping with a dermal sinus tract. Intraoperative photographs show; **B** the dermal sinus tract associated with a bony erosion (black arrow); **C** craniotomy along with the tract (black arrow) and, **D** after its excision.

Histopathological examination confirmed a dermoid cyst (Fig. 3). A post-operative MRI scan did not show any remnants.

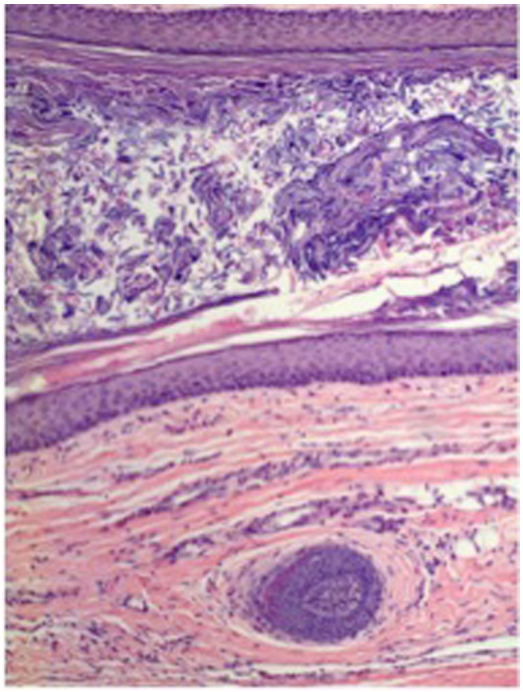

Figure 3 - Histopathological examination showing a dermoid cyst with a deep collagen layer and well formed stratified squamous epithelium. Hair follicles and sebaceous glands can also be seen (H & E, original magnification x200).

The patient had an uneventful postoperative course. She was discharged two weeks later, after completion of the antibiotic therapy.

Conflict of interest

The authors declare that they have no conflict of interest related to this article.

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