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Dermoid Cyst of the Posterior Cerebral Fossa Associated with A Congenital Dermal Sinus in A Child:

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Abstract: *The dermoid cyst of the central nervous system is a very rare entity. It is a benign tumor that can be associated with dysraphic disorders, mainly in the dermal sinus. Hydrocephalus and surinfection are the main complications of the dermoid cyst of the posterior cerebral fossa. Neuroradiological examinations (especially CT and MRI) are necessary for diagnosis, followed later by anatomopathological confirmation. The curative treatment is surgical. We present a case of a 04-year-old child, without any notable pathological history, who was consulted for acute isolated intracranial hypertension, with a cerebral CT showing an obstructive hydrocephalus that complicates a dermoid cyst of the posterior cerebral fossa associated with an occipital dermal sinus.* ***Purpose:*** *Consider the posterior dermoid cerebral fossa cyst in children with a median cystic lesion on sectional imaging, especially if it is associated with a complete or incomplete occipital dermal sinus.*

Keywords: dermoid cyst, dermal sinus, posterior cerebral fossa, tumor.

Introduction

The dermoid cyst of the central nervous system is a very rare entity. It is a benign tumor that can be associated with dysraphic disorders, mainly in the dermal sinus. Hydrocephalus and surinfection are the main complications of the dermoid cyst of the posterior cerebral fossa. Neuroradiological examinations (especially CT and MRI) are necessary for diagnosis, followed later by anatomopathological confirmation.

We report an observation of a dermoid cyst of the posterior cerebral fossa associated with an occipital dermal sinus, in a cephalalgic child, with the contribution of imaging in the diagnosis of this anomaly.

Patient and Observation

Female child, 04 years, without any notable pathological history, having intense headaches for four days of progressive aggravation, associated with vomiting and visual disorders, evolving in a context of preservation of general state and apyrexia. The general somatic examination found a conscious patient, neurologically and hemodynamically stable, normocardial, apyretic at 36.5°C. Neurological examination found a firm neck, no motor or sensory deficit, no signs of cranial pair damage, and no cerebellar syndrome.

Faced with isolated intracranial hypertension syndrome, a cerebral computed tomography (CT) scan was performed before and after contrast, showing a subtentorial cyst, medial vermian, rounded in shape, well limited, with hypodense content (density greater than CSF) containing a few very hypodense punctiform inclusions (of fatty density), measuring 53x35mm in axial diameters. This lesion does not change after contrast, is not surrounded by peri-lesional edema, and is responsible for an active triventricular hydrocephalus, by compression of the fourth ventricle**. (Figure.1).**

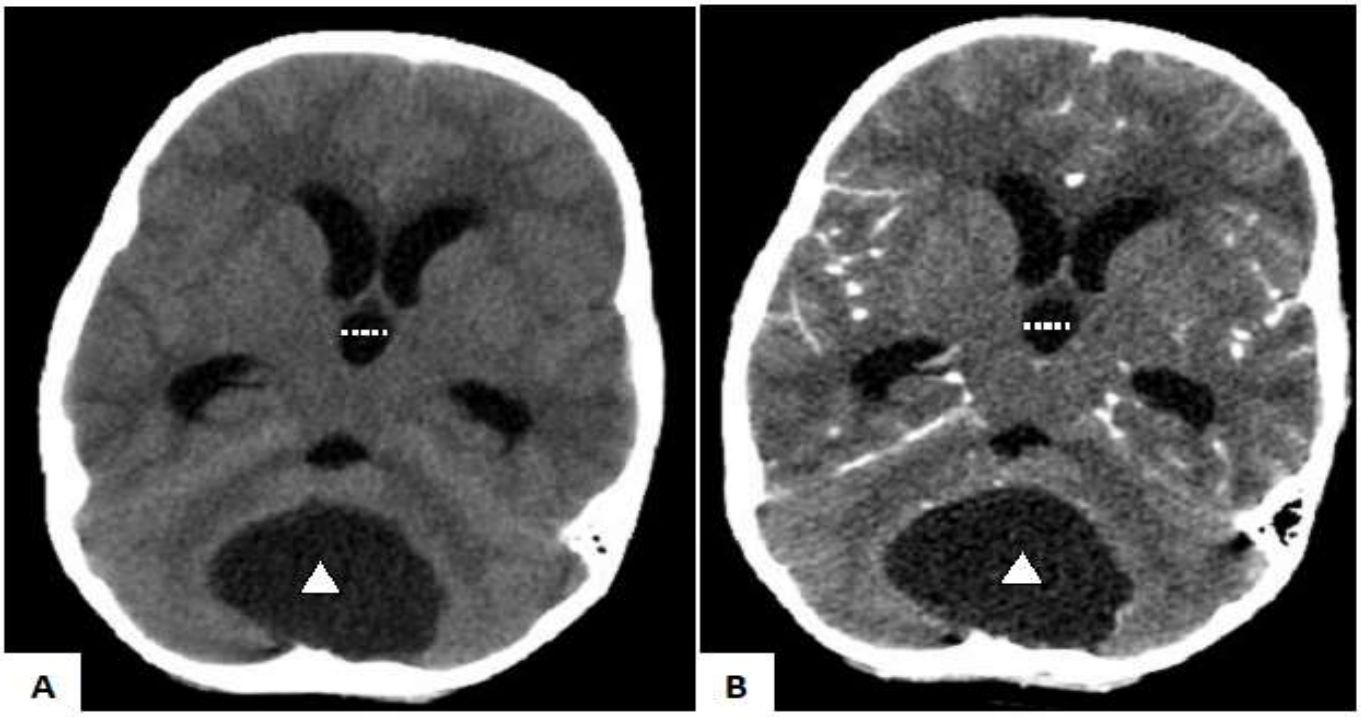

Figure 1: Cerebral CT scan (A : before contrast) and (B : after contrast) showing a subtentorial dermoid cyst (white triangle), as a median vermian lesion, thin-walled, hypodense (density greater than that of the CSF) containing a few very hypodense punctiform inclusions (of fatty density).

This cyst compresses the 4th ventricle, responsible for an active tri-ventricular hydrocephalus (white dotted line).

There is a small subcutaneous medial occipital cystic lesion of 5 mm long axis, without communication with the cyst described above. **(Figure.2)**

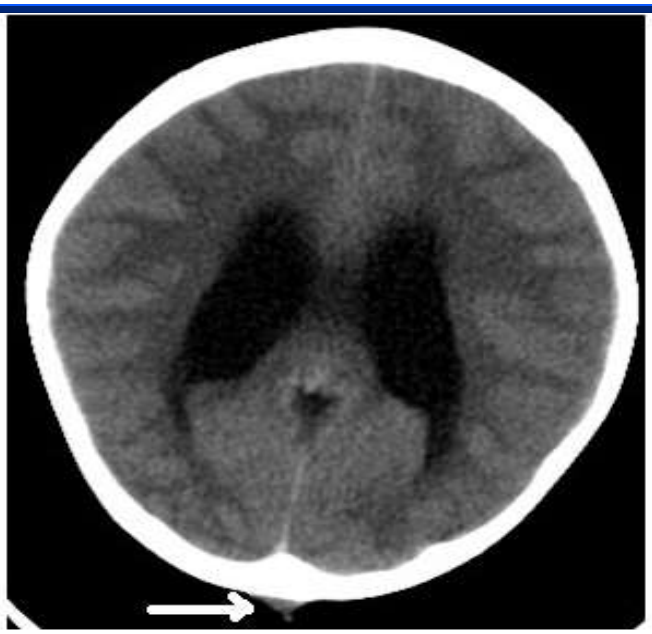

Figure 2 (same patient): Cerebral CT scan: showing a small medial occipital subcutaneous lesion, with a liquid density of 5 mm in length, without any mass effect on the adjacent bone, nor communication with the above-mentioned cyst, related to a partial dermal sinus (white arrow)

In front of this clinicoradiological picture, a dermoid cyst of the posterior cerebral fossa, associated with a partial dermal sinus, was first evoked. The patient was taken to the emergency operating room. A medial suboccipital approach allowed complete removal of the extra parenchymal lesion, whose macroscopic examination, after opening its thin wall, found the presence of a brown viscous liquid and hairs, thus confirming our radiological diagnostic suspicion. The patient had a good postoperative evolution, without the need for external ventricular drainage, and was discharged on the fourth postoperative day. Histological examination revealed a stratified squamous epithelium with fatty debris and hair follicles. The diagnosis of dermoid cyst was retained with certainty.

Discussion:

Intracranial dermoid cyst is a rare entity. It represents 0.1-0.7% of all brain tumors [1] and occurs mainly in early adolescence [2]. They are benign congenital tumors that are known to progressively evolve with slow growth due to progressive epithelial desquamation and secretion of glands in the cyst. The cyst originates from the inclusion of ectodermal elements in the neural tube during its closure between the third and fifth weeks of embryonic development. [3,4]
The frequent location of dermoid cysts is most often at the cisternal level, mainly in the cerebellopontine angle and the parasellar cistern. The location in the posterior cerebral fossa is exceptional. At this level, they usually develop in the midline, mainly in the vermis, but also in the fourth ventricle [5]. Lateral localizations have also been described in the cerebellar hemispheres, although rare. In the latter position, these tumors can extend asymmetrically into the cerebellar hemisphere, the bulk of the tumor overlying the vermis or penetrating the outlet of the fourth ventricle [6]. These tumors can be associated with dysraphic disorders [7]. This explains the frequent association of dermoid cysts with the dermal sinus, myelomeningocele, and sometimes with Klippel-Feil skin syndrome [8]. The dermal sinus can be located anywhere in the midline from the nasion to the coccyx, but is more common in the lumbosacral and occipital region. [9,10] The last case corresponds to our observation.
Through this sinus there is a permanent risk of infection of the central nervous system, such as recurrent bacterial meningitis in childhood or cerebellar abscess [11].

Logue and Till2 [12] classified posterior fossa dermoid cysts, according to whether they are extradural or intradural, and according to the degree of development of the dermal sinus, absent, partial, or complete, into 4 groups: extradural dermoid cyst with complete dermal sinus; intradural dermoid cyst (without dermal sinus); intradural dermoid cyst with incomplete dermal sinus; and intradural dermoid cyst with complete dermal sinus. According to this classification, our case is type 3.

The clinical manifestations of posterior cerebral fossa dermoid cysts are related to their complications. The main symptoms found are intracranial hypertension syndrome with headache, nausea, vomiting, decreased visual acuity, papilar edema, or cerebellar signs such as ataxia, seizures, and cranial nerve paralysis (usually nerves VI and VII).[1,13]
The dermoid cyst can be complicated by an upstream hydrocephalus, by compressive mass effect, generating an increase in intracranial pressure. This occurs when it is present in the posterior cerebral fossa, which is the case in our patient. The second complication is rupture in the subarachnoid or intraventricular spaces, causing aseptic meningitis, recurrent meningitis, subdural empyema, and intraparenchymatous abscess. [14]

Radiologically,
On CT, dermoid cysts appear as a well-limited solitary lesion formation, hypodense (fat density of -20 to -140 HU, depending on their lipid content), not enhanced after contrast (the enhancement would rather suggest an epidermoid than a dermoid cyst). Parietal calcifications are often present.
Sometimes they appear hyperdense, mimicking a hemorrhage, or are related to the presence of powdery intracystic microcalcifications. The cyst may be round, oval, or bilocular and varies from a few millimeters to several centimeters in diameter. [15]
On magnetic resonance imaging, which is the key examination, the dermoid cyst is characterized by a T1 hypersignal explained by a high cholesterol density in the cyst, and varies from hypo to T2 hypersignal, with generally a FLAIR hypersignal, not restrictive in diffusion. Fat saturation sequences erase the lesional signal, confirming the lipidic character. It does not improve after Gadolinium injection [16].
The main differential diagnoses on imaging are epidermoid cyst, pure cystic craniopharyngioma, and arachnoid cyst. Epidermoid cysts are more variable in location than dermoid cysts and are usually nonmedial. Magnetic resonance features allow differentiation between these different entities. [17 ,18]

The curative treatment is exclusively surgical. [19] It consists of microsurgical evacuation of the cyst contents and complete removal of the capsule. However, in a certain number of cases, surgical excision is incomplete, leaving a tumor residue in place due to the sometimes frequent adhesions of the capsule to adjacent structures, the complete release of which carries a risk of neurological sequelae and death. In the presence of hydrocephalus, external ventricular drainage promoted more favorable operative conditions and reduced the risk of hypotonic residual ventricular dilation. [20]

In our case, tumor resection was complete with a medial occipital approach, without external ventricular drainage. The immediate evolution was favorable.

Conclusions

In summary, a posterior cerebral fossa dermoid cyst should be considered in children with a midline cystic lesion, especially if it is associated with a complete or incomplete occipital dermal sinus. Neuroradiological examinations (especially CT and magnetic resonance imaging) are necessary for diagnosis, followed later by pathological confirmation. Once diagnosed, surgical excision should be performed early to prevent intracranial complications such as obstructive hydrocephalus and intraventricular or subarachnoid rupture causing recurrent meningitis.

CONFLICT OF INTEREST

None.

CONSENT TO PUBLISH:

Written informed consent was obtained from the parents. They consented to the submission of the case report to the journal.

References :

1. LUNARDI P, MISSORI P, GALIARDI FM, FORTUNA A. Dermoid cyst of the posterior fossa in children: report of nine cases. Surg Neurol 1990; 34: 39-42.
2. GUIDETTI B, GAGLIARDI FM. Epidermoid and dermoid cysts: clinical evaluation and late surgical results. J Neurosurg 1977; 47: 12-18.
3. SHARMA MS, SHARMA BS, YADAV A, KHOSLA VK. Posterior fossa dermoid in association with Klippel-Feil syndrome: a short report. Neurol. India. 2001; 49: 210-212.
4. DIAZ A, TAHA S, VINIKOFF L, ALLAOUI M, COMOY J, LERICHE B. Kyste dermoïde des ventricules latéraux associée à un sinus dermique éthmoïdal : à propos d'un cas. Neurochirurgie. 2000; 46: 39-42.
5. BENZAGMOUT ET AL; Posterior fossa dermoid cyst... Neurosciences 2011; Vol. 16 (2) .153-155.
6. NAJIB J 1 LAGHMARI Mi 1 LMEJJATI M 1 ANIBA K 1 GHANNANE H 1 AIT BENALI S1 Les kystes dermoïdes de la fosse cérébrale postérieure ajns-2010-vol-29-no-2 .
7. GELABERT-GONZALEZ M. Intracranial epidermoid and dermoid cysts. Rev Neurol. 1998; 27: 777-782.
8. HINOJOSA M, TATAGIBA M, HARADA K, SAMII M. Dermoid cyst in the posterior fossa accompanied by Klippel-Feil syndrome. Childs Nerv Syst. 2001; 17: 97.
9. AKSOY FG, AKSOY OG, GOMORI JM. Klippel-Feil. syndrome in association with posterior fossa suboccipital dermoid cyst. Eur Radiol. 2001;11: 142-144.
10. Cai, C.-Q., Zhang, Q.-J., Hu, X.-L., & Wang, C.-X. (2008). Dermoid cyst of the posterior fossa associated with congenital dermal sinus in a child. World Journal of Pediatrics, 4(1), 66–69. doi:10.1007/s12519-008-0014-x
11. GOFFI N J, PLETS C, VAN CALENBERGH F, WEYNS F, VAN HAVENBERGH T, EECKELS R, ET AL. Posterior fossa dermoid cyst associated with dermal fi stula. Childs Nerv Syst. 1993; 9: 179-181.
12. LOGUE V, TILL K. Posterior fossa dermoid cysts with special reference to intracranial infection. J Neurol Neurosurg Psychiatry. 1952; 15: 1-12.
13. Eustathios Vlachakis, George A. Alexiou, Kalliopi Stefanaki,1 George Sfakianos, and Neofytos Prodromou. Posterior fossa dermoid cyst ; Journal of Pediatric Neurosciences 7(1):79
14. Abrar A. Wani, MS, mch, Uday S. Raswan, MB, MS, Nayil K. Malik, MS, mch, and Altaf U. Ramzan, MS, mch Posterior fossa ruptured dermoid cyst presenting ; Neurosciences 2016; Vol. 21 (4)
15. BROWN JY, MOROKOFF AP, MITCHELL PJ, GONZALES MF. Unusual imaging appearance of an intracranial dermoid cyst. AJNR Am J Neuroradiol 2001; 22: 1970-1972.
16. HAYEK G, MERCIER P, FOURNIER HD, MENEI PH, POUPLARD F, GUY G. Sinus dermique avec kyste dermoïde de la fosse cérébrale postérieure révélé par un abcès à propos de 2 cas pédiatriques avec revue de la littérature. Neurochirurgie. 2001; 47: 123-127.
17. W.B. Gormley, F.J. Tomecek, N. Qureshi Craniocerebral epidermoid and dermoid tumors-a review of 32 cases Acta Neurochir (Wien), 128 (1994), pp. 1115-1121
18. M. Caldarelli, L. Massimi Intracranial midline dermoid and epidermoid in children J Neurosurg, 102 (2005), pp. 135-137
19. Dange N, Bonde V, Goel A, Muzumdar D. Posterior fossa dermoid cyst in a patient with Goldenhar syndrome. Pediatr Neurosurg 2007;43:522-525.
20. Tekkök IH, Baeesa SS, Higgins MJ, Ventureyra EC. Abscedation of posterior fossa dermoid cysts. Childs Nerv Syst 1996;12:318-322.