Cranial dermal sinuses in pediatrics: Presentation, diagnosis, complications, and management. Experience at a tertiary care children's hospital

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ABSTRACT

Introduction. A cranial dermal sinus is a tubular tract resulting from the incomplete separation of the epithelial ectoderm from the neuroectoderm which may lead to infectious complications. There have been isolated reports of this condition.

Objective. To describe a series of patients with cranial dermal sinus, its presentation, diagnosis, management, and complications.

Population and methods. Observational, descriptive study of a series of pediatric patients with cranial dermal sinus treated at a tertiary care children's hospital between 2014 and 2019.

Results. A total of 18 patients were included. The clinical presentation was a specific lesion on the scalp in 12 cases, intracranial hypertension in 4, ataxia in 1, and a specific lesion with fistula tract in 1. Half of patients had symptoms of infection. The lesion was located in the midline of the occipital bone in 13 cases; in the midline of the frontal bone in 3 cases; and in the interparietal midline in 2 cases.

The dermal sinus was associated with an extracranial image in 5 cases and an intracranial complication in 11 cases. No patient had recurrence and only one surgery was performed in all of them.

Conclusions. In this series, dermal sinuses presented as specific lesions on the scalp. The most common site was the occipital midline, and more than 50% of these extended through the bone. The treatment of choice was complete resection of dermal sinus and associated lesions. An emergency surgery was performed when the dermal sinus was symptomatic or associated with intracranial lesions.

Key words: *dermal sinus, complications, infections, imaging studies, pediatrics*.

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INTRODUCTION

A cranial dermal sinus is a tubular tract lined by stratified squamous epithelium resulting from the incomplete separation of the epithelial ectoderm from the neuroectoderm during the first weeks of gestation.[1] It extends between the superficial dermal layers and the deepest cranial structures and may end at the subcutaneous cellular tissue and the dura mater, and even reach neural structures, favoring a potential communication between the skin and the central nervous system, which may result in severe infectious complications.[2,3]

To date, there have been isolated cases of dermal sinuses reported in the international literature.[4-8]

The objective of this study was to describe a series of patients with cranial dermal sinus, its presentation, diagnosis, management, and complications.

POPULATION AND METHODS

This was a descriptive, observational study that included all the patients with a histopathological diagnosis of cranial dermal sinus operated at Hospital de Pediatría Juan P. Garrahan between January 2014 and August 2019, and who had at least a magnetic resonance imaging (MRI) or computed tomography (CT) scan test from the time of diagnosis. If there was suspicion of infection, a study with a contrast agent had to be performed. Patients with incomplete data in the medical record or a follow-up of less than 12 months were excluded.

For purpose of the analysis, information was collected about the patients' sex and age and presenting symptoms; patients who were admitted with symptoms of infection (markers of infection include fever, meningeal syndrome, and purulent material discharge through the defect) were identified. The functional status scale (FSS) was administered to all patients before the surgery and at discharge.[9]

Imaging studies were done to assess the site and depth of the lesion (subcutaneous cellular tissue, bone, dura mater or intracranial) and the type and presence of extracranial or intracranial lesions (cyst, infected cyst, or abscess). Contrast enhancement and intracranial lesion restriction were studied. The presence of hydrocephalus was also investigated.[10]

The time in days from diagnosis to surgery was recorded and patients were classified based on whether or not they had infection at the time of surgery. It was considered that patients had an infection at the time of surgery if they had symptoms of infection before performing the surgical procedure. During surgery, the sinus depth was recorded and the specimen was submitted for histopathological examination. If the patient had an intracranial lesion, histopathological findings and the type of resection were recorded. Post-operative complications were recorded along with the need to place a ventriculoperitoneal shunt up to 30 days after surgery.

The length of stay (in days) was also recorded. If the patient had not undergone surgery during the period in hospital when the diagnosis was made, the total number of days of hospitalization was added up. Patients were followed up for at least 12 months. Surgical failure was defined as a patient requiring a second surgery or developing new symptoms during follow-up.

Statistical analysis

Continuous variables were described as median and interquartile range, whereas categorical variables were represented by absolute frequency.

Ethical considerations

This study was conducted in accordance with the principles of the Declaration of Helsinki. It was approved by the Research Ethics Committee of Hospital Garrahan. Information was kept confidential, and all study participants were asked to give their informed consent.

RESULTS

TABLE 1. *Baseline characteristics of the 18 patients included in the series*

Case	Sex	Age (months old)	Diagnostic symptom	Infection	Site	Extracranial complication	Intracranial complication	Surgery type	Complications	Length of stay (days)	Resolution	Follow-up (months)
1	M	17	SD and F	1	FML	1	1	Intracranial surgery	Yes	30	Yes	12
2	M	9	SD	0	OML	1	0	Tract resection	No	2	Yes	12
3	M	12	Fe and P	1	FML	1	1	Intracranial surgery	No	4	Yes	26
4	M	96	Fe	1	OML	0	1	Intracranial surgery	No	60	Yes	15
5	M	28	SD	0	IPML	1	0	Tract resection	No	3	Yes	12
6	M	170	SD and Fe	1	OML	0	1	Intracranial surgery	No	2	Yes	25
7	M	163	SD	0	IPML	0	0	Bone resection	Yes	2	Yes	20
8	M	157	Fe and M	1	OML	0	1	Intracranial surgery	No	35	Yes	18
9	M	111	SD and IHT	0	FML	1	1	Intracranial surgery	No	6	Yes	12
10	F	2	SD	0	OML	0	1	Intracranial surgery	No	7	Yes	30
11	F	45	IHT and Fe	1	OML	0	1	Intracranial surgery	Yes	352	Yes	24
12	F	25	SD	0	OML	0	0	Bone resection	No	7	Yes	19
13	M	125	SD	0	OML	0	0	Dural closure	No	2	Yes	37
14	M	185	SD and P	1	OML	0	0	Dural closure	No	2	Yes	12
15	M	1	SD	0	OML	0	0	Dural closure	No	3	Yes	13
16	F	16	SD, Fe, and P	1	OML	0	1	Intracranial surgery	No	57	Yes	15
17	M	29	A	0	OML	0	1	Intracranial surgery	No	29	Yes	12
18	F	20	SD, IHT, and M	1	OML	0	1	Intracranial surgery	No	48	Yes	12

SD: specific deficit; F: fistula; Fe: fever; P: purulent material; M: meningitis; IHT: intracranial hypertension; A: ataxia; OML: occipital midline; FML: frontal midline; IPML: interparietal midline.

A total of 21 patients with cranial dermal sinus operated on at Hospital Garrahan between January 2014 and December 2019 were identified. Two patients were excluded because they did not have brain imaging studies and 1 had been operated on at a different facility.

Eighteen patients with cranial dermal sinus were reviewed. *Table 1* describes the main characteristics of each patient. The study population included 13 males; the median age was 28.5 months (16-125) ranging from 1 month to 15 years. The median length of stay was 6.5 days (2-35) and the median follow-up of patients was 15 months (12-24).

Lesion presentation and site

Out of the 18 patients studied, 13 had a specific lesion on the scalp compatible with the diagnosis of dermal sinus (*Figure 1 A-E*); 1 had a fistula with cerebrospinal fluid (CSF) discharge through the defect. Half of patients had symptoms suggestive of infection, including fever in 5 cases, meningitis in 2 cases, and discharge of purulent material from the specific defect in 3 cases. Other forms of presentation included symptoms of intracranial hypertension, such as vomiting, headache, and altered sensorium in 4 cases, and ataxia in 1 case. The median FSS score before surgery was 6 (6-7), ranging from 6 to 30.

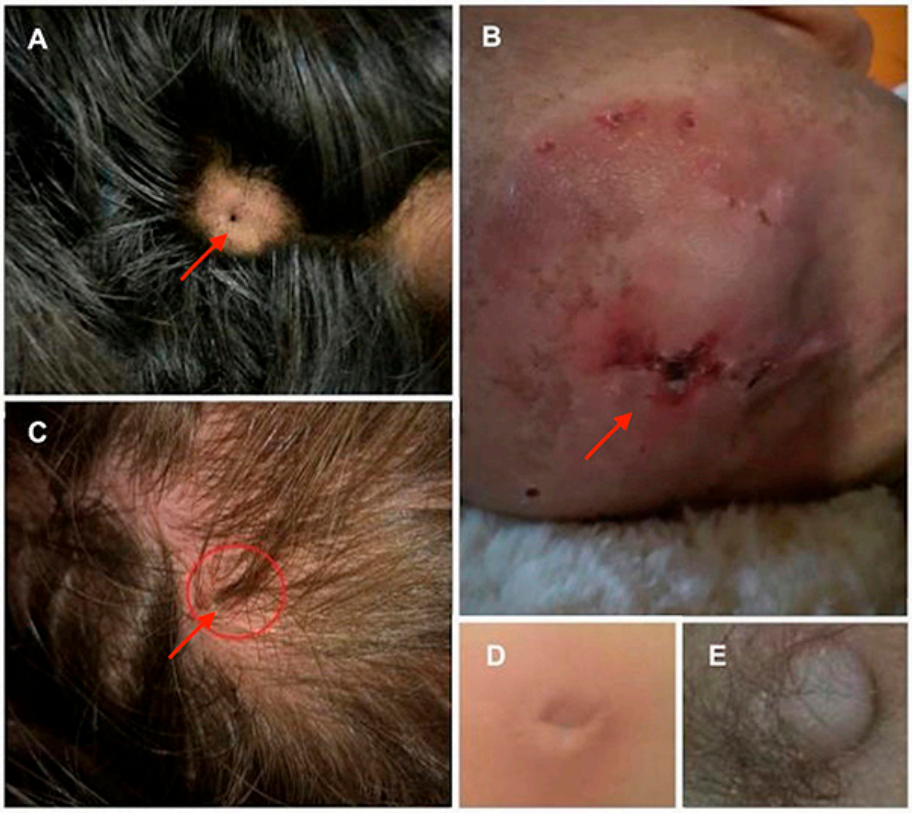
The lesion was located in the midline of the occipital bone in 13 cases; in the midline of the frontal bone in 3 cases; and in the interparietal midline in 2 cases. No dermal sinus tracts were found outside the midline.

Neuroimaging

In the imaging studies it was found that the tract ended at the level of the subcutaneous cellular tissue in only 1 case, it reached the bone in 3 cases, the dura mater in other 3 cases, and the central nervous system (CNS) in 11 cases either to the brain in the case of supratentorial sinuses or to the cerebellum in those of the posterior fossa.

An extracranial image was also found in

FIGURE 1. *Scalp lesions suggestive of dermal sinus. A: Specific lesion in the frontal midline (red arrow), B: Edematous occipital lesion with infection and scabby surface (red arrow), C: Specific lesion with small hair follicle coming out of the defect (red arrow), D: Small skin indentation, E: Elevated lesion containing fluid*



5 cases. Four of them corresponded to a dermoid/epidermoid cyst, and 1, to an infectious collection. An intracranial lesion was present at the time of diagnosis in 11 patients. Four of these corresponded to an infected dermoid cyst; 3, to a dermoid/epidermoid cyst (*Figure 2 A-E*); and 4, to an abscess (*Figure 3 A-C*). The intracranial lesion was located in the cerebellum (1 cyst, 2 superinfected cysts, and 2 abscesses), in the occipital lobe (1 cyst, 1 superinfected cyst, and 1 abscess), and in the frontal lobe (1 cyst and 1 superinfected cyst).

Most intracranial lesions had contrast enhancement (7/11) and 5/11, restriction in diffusion (*Figure 4 A-F*).

The diagnostic imaging study showed that 4 patients had hydrocephalus; they corresponded to the patients who had intracranial hypertension at the onset and to 1 patient with asymptomatic ventriculomegaly.

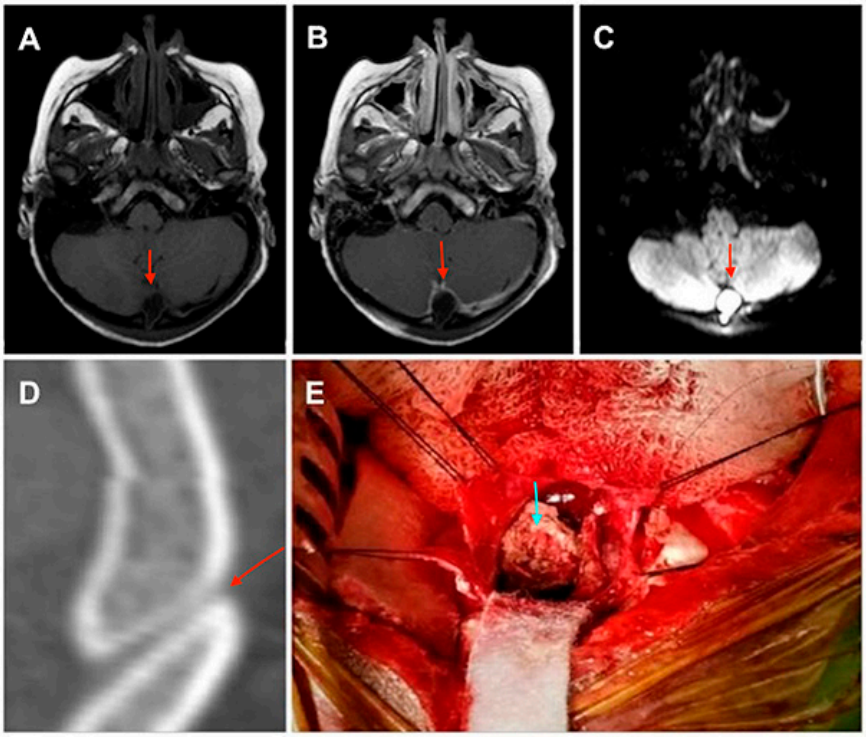
Surgery

A median of 37 days (13-102) elapsed between diagnosis and surgery. In 4 patients, surgery was performed while they had an infection, whereas the procedure was delayed until the infection had resolved in 5 cases.

In 2 cases, resection of the fistulous tract was sufficient, in other 2 cases bone enlargement was required; in 3 cases the dura mater had to be sutured or ligated; and in 11 cases the dura mater had to be opened to treat an intracranial lesion (*Figure 5 A-D*). The dermal sinus ended close to a venous sinus in 2 patients.

Intra-operative findings showed that the tract ended at the level of the subcutaneous cellular tissue in only 1 case, it reached the bone in 1 case; the dura mater, in 5 cases; and the CNS,

FIGURE 2. *Patient with dermal sinus and complication due to intracranial epidermoid cyst. A: Brain magnetic resonance imaging (MRI), T1 images without contrast, axial section showing rounded interhemispheric lesion in the posterior fossa which appears to extend from the bone (red arrow); B: Brain MRI, T1 images with contrast, axial section showing the same lesion, no wall enhancement; however, the dura mater area in contact with the lesion is enhanced (red arrow); C: Brain MRI, diffusion image, axial section showing a uniform restriction of the lesion leading to a suspected diagnosis of epidermoid cyst (red arrow); D: Brain computed tomography (CT) scan, sagittal section, bone window showing solution of continuity of the bone due to dermal sinus (red arrow); E: Intra-operative image showing the posterior fossa dura mater is open and retracted using anchors; pearly white material, similar to typical characteristics of an epidermoid cyst is observed inside (light blue arrow)*



¿Figura en color?

FIGURE 3. *Patient with dermal sinus and supratentorial abscess complication. A: Brain magnetic resonance imaging (MRI), T1 images without contrast, axial section showing occipital lesion pressing and displacing midline structures and causing the collapse of the temporal horn; B: Brain MRI, T1 images with contrast, axial section showing the same occipital lesion described before, where ring contrast enhancement is observed, suggesting an infectious process; C: Brain MRI, diffusion image, axial section showing peripheral restriction in the cystic lesion, compatible with an abscess; in addition, the left temporal horn shows restriction, suggesting it had opened inside the ventricle and caused a pyocephalus*

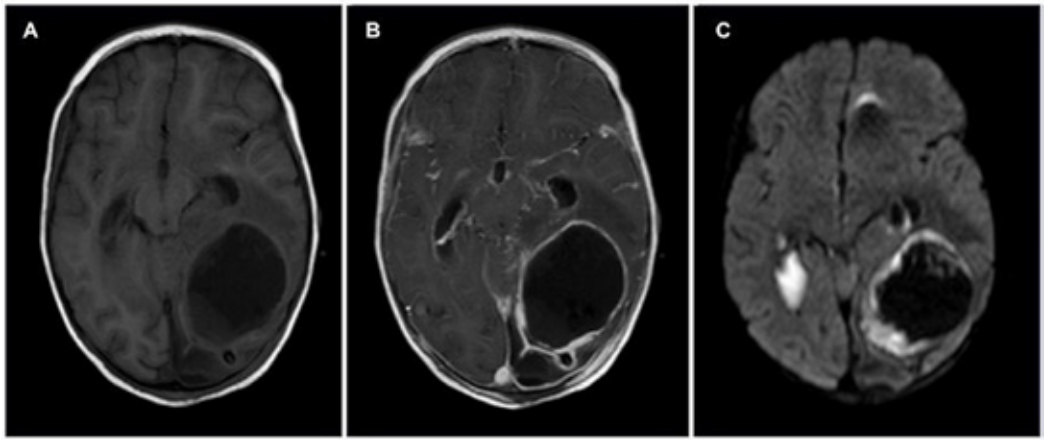
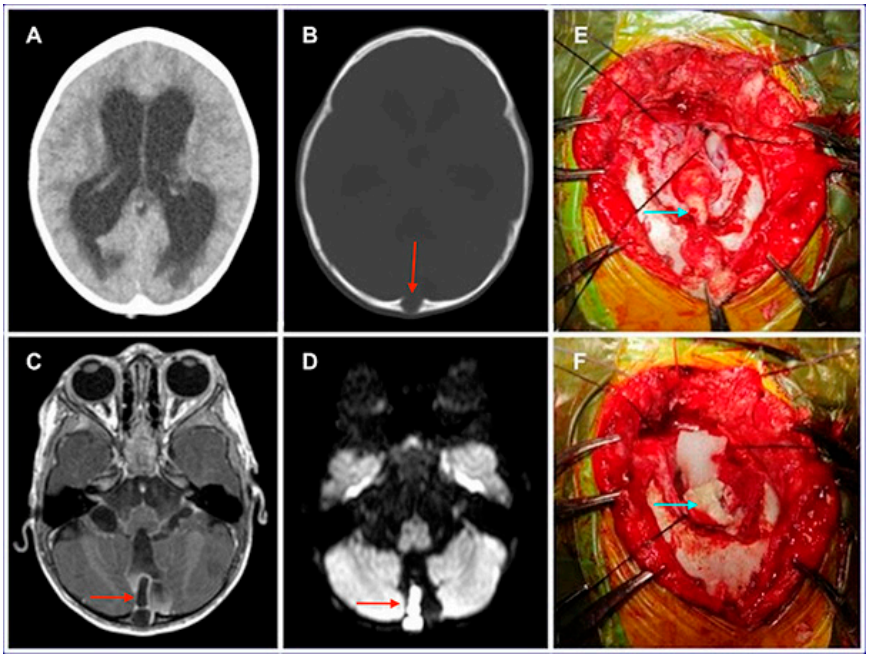


FIGURE 4. *Patient with dermal sinus and complication due to infected dermoid cyst and hydrocephalus. A: Brain CT scan, axial section showing enlarged lateral ventricles, reduced cortical subarachnoid spaces, and transependymal edema compatible with hydrocephalus; B: Brain CT scan, bone window showing solution of continuity of the occipital bone, where the dermal sinus enters (red arrow); C: Brain MRI, T1 images with contrast, axial section showing interhemispheric lesion in the posterior fossa, where ring contrast enhancement is observed (red arrow), suggesting an infected cyst; D: Brain MRI, diffusion image, axial section showing a uniform restriction of the lesion (red arrow), suggesting an infectious process; E: Intra-operative image showing occipital bone craniotomy, which allows the dermal sinus to enter the dura mater (light blue arrow); F: Intra-operative image of the site where the dura mater was opened and repaired showing purulent material coming from the intradural space (light blue arrow)*



in 11 cases.

Complete resection of the 11 intracranial complications was performed. The histopathological report indicated the presence of a dermoid cyst in 1 case; an infected dermoid cyst, in 4 cases; an epidermoid cyst, in 2 cases; and an abscess in 4 cases. *Staphylococcus epidermidis, S. aureus, Peptoniphilus asaccharolyticus*, and *S. lugdunensis* were isolated in cultures.

The median post-operative FSS score was 6 (6-6), better compared to the pre-operative score.

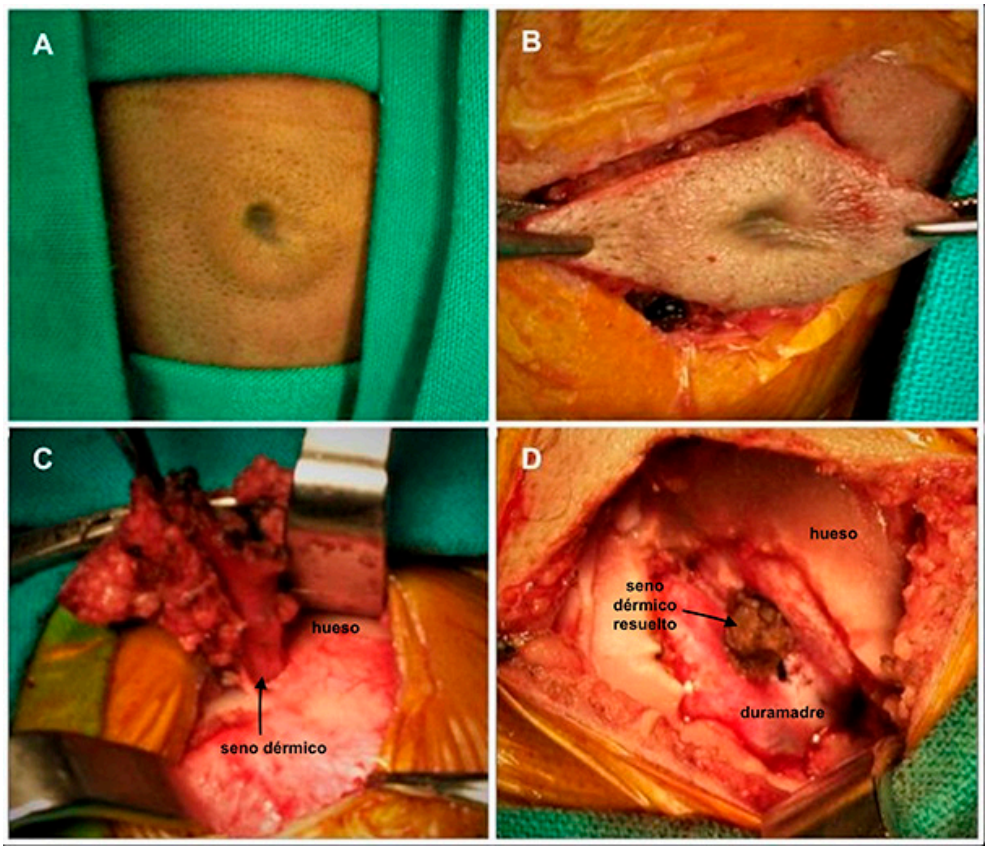
Post-operative complications were found in 3 patients: 1 fistula, 1 superficial infection, and 1 death, corresponding to a patient who was in a very poor condition at the time of surgery, with an FSS score of 30. Out of the 5 patients with ventriculomegaly before surgery, 3 required the placement of a ventriculoperitoneal shunt in the postoperative period. No patient relapsed, and all were operated only once.

DISCUSSION

A dermal sinus results from the failure of neuroectoderm to separate from the cutaneous ectoderm, thus creating a local communication between the developing nervous system and the skin. A dermal sinus may reach the subcutaneous cellular tissue, the bone, the dura mater or the SNC, depending on the level at which fusion stopped.[7]

Histopathological results show that dermal sinuses contain ectodermal and mesodermal derivatives. A dermal sinus is composed of a stratified squamous epithelium lining and specialized adnexal structures that may contain hair follicles, pilosebaceous glands, and smooth muscle.[11] Dermoid/epidermoid cysts or tumors developed from dermal sinuses result from the cellular desquamation of the elements lining the tract.[12]

FIGURE 5. *Intra-operative image of dermal sinus with intracranial extension. A: Skin defect; B: Lozenge resection of specific defect; C: Dermal layers opening to see dermal sinus reaching the intracranial space through the bone defect; D: After craniotomy, the dermal sinus is resected at the level of the dura mater and the defect is covered with clotting material*



Although dermal sinuses may be located in any site along the neuraxis, the most common site is the skull, in the occipital region (85%), whereas 11% are in the frontal region and 5%, in the posterior parietal region.[2] In this series of patients, distribution was similar to that described in the bibliography, with a predominance in the occipital bone and to a lesser extent in other sites. There have been isolated reports of lateral dermal sinuses, close to the external auditory canal in the temporal region, although these are extremely rare.[4]

Dermal sinuses are more common among children younger than 5 years and have a similar distribution in both males and females. Given that this is a congenital lesion, in most cases, diagnosis depends on the time when the patient develops symptoms. In the case of asymptomatic lesions, the only sign is usually a specific defect, which may or may not be accompanied by skin discoloration or a tuft of scalp that exits through the defect. In this study, over 50% of patients had a specific lesion in the scalp, which may have aided with the diagnosis.

In the case of symptomatic patients, symptoms vary depending on whether it is a simple dermal sinus or is accompanied by an intracranial or extracranial lesion. Among the latter, symptoms of infection are more common, which may range from skin redness to fulminant meningitis.[13,14] Half of patients in this study had signs of infection, and more than half had an associated intracranial lesion. Among these, prevailing symptoms were related to an infection in more than 50%, followed by symptoms of intracranial hypertension, ataxia, and fistula.

Inclusion tumors are associated with 89% of dermal sinuses. Dermoid cysts are the most common type. In 82% of cases, they extend until reaching the subdural space or go deeper, while 18% are exclusively located in the extradural space.[12-15]

These lesions are diagnosed using MRI and CT scans. The most important information derived from these studies is the extension of the dermal sinus and whether it is accompanied or not by an intracranial or extracranial lesion. A CT scan shows the relationship between the dermal sinus and the bone in greater detail; in addition, it is useful if a dermoid cyst is suspected because these lesions may appear as hypodense in the CT due to their high fat content. An MRI allows to see the dermal sinus in greater detail at the level of the subcutaneous cellular tissue, especially in T2 and FLAIR sequences. Besides, an MRI is the study of choice if the dermal sinus is associated with an intracranial or extracranial lesion. The most common differential diagnoses include dermoid cyst, epidermoid cyst or abscess. The signal intensity of epidermoid cysts is similar to that of CSF in T1 and T2 weighted images and do not usually show contrast enhancement; however, the diagnosis is confirmed using diffusion weighted imaging (DWI) because they have a typical homogeneous restriction. Dermoid cysts, on the other side, tend to show a hyperintense signal in T1 and a hypointense signal in T2 images, and do not usually have contrast enhancement. Hyperintensity usually disappears in the fat suppression sequence. If contrast enhancement or restriction in DWI is observed, superinfected of the cyst should be suspected. In addition, abscesses usually show ring-enhancement with contrast material besides restriction in DWI.[16] In our series, 3 of the 4 infected cysts showed contrast enhancement and restriction in DWI. The 2 cases of epidermoid cyst showed restriction in DWI and no contrast enhancement, whereas the 4 abscess cases showed contrast enhancement and 3 of them, restriction.

Intracranial lesions are managed with complete resection, including the dermal sinus and any associated lesion.[7,8,12] Incomplete resection may result in recurrent meningitis and the development of new tumor lesions.[17] For surgery, the incision of the skin is made using the buttonhole technique; divulsion of the muscle planes is done protecting the tract because this is the only way to determine the lesion depth. Once the site of the tract insertion is defined (bone, dura mater or CNS), ligation is performed above and below the incision, so that the tract can then be resected without complications. The distal end is cauterized and a specimen is submitted for histopathological examination. If the patient has tract infection, meningitis or hydrocephalus, these conditions should be managed before surgical resection, unless the patient's general status prevents this.[18] In our series, 4 patients underwent surgery while receiving antibiotic therapy due

to dermal sinus infection. The urgency of the surgery was associated with the severity of symptoms. The complication rate of this type of surgery is usually low if operated at an early stage, but it is higher when there is intracranial involvement or infections.[19] If an intracranial lesion is present, surgery should be immediately done to prevent a permanent neurological deficit. The only patient who died had been admitted to the hospital in a very poor condition. Death occurred during the post-operative period. The other complications were mild.

We believe that these lesions should be managed aggressively and a complete resection of all lesion components should be considered. In our study, we did not observe recurrence of symptoms of the lesion in any patient, and the complication rate was low.

It is worth noting the importance of preventing these lesions because preventive surgery usually entails minimal morbidity and mortality. In our cohort, the 4 patients who underwent surgery due to asymptomatic lesions did not have any complications during the post-operative period and were discharged 3 days after the surgery. For this reason, the systematic screening of specific defects along the neuraxis is recommended during routine pediatric checkups. If a dermal sinus is diagnosed in an asymptomatic patient, they should be referred immediately to a specialist (pediatric neurosurgeon), who will request imaging studies and schedule surgery as soon as possible.

CONCLUSIONS

In this series of patients, the most common presentation of dermal sinuses corresponded to specific lesions on the scalp; almost 50% of patients showed symptoms of infection. The most common site was the midline of the occipital bone, and more than 50% of these extended through the bone. The treatment of choice was complete resection of the dermal sinus and associated lesions. If an asymptomatic lesion is diagnosed, surgery may be scheduled; however, in the case of a dermal sinus presenting with symptoms or in association with intracranial lesions, an emergency surgery should be performed. ■

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