

Congenital Spinal Dermal Sinuses Long-Term Analysis at King Hussein Medical Center .

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ABSTRACT

Objectives: The aim of this study is to investigate the corresponding variables: age, sex, the reason for consultation, clinical manifestations, location, methods of diagnosis, time of surgery, type of surgery, histological diagnosis, the presence of infection and subsequent evolution, with baseline comparison of our findings to those reported in the literature.

Methods: A retrospective analysis carried out of the medical records of all the patients presented with spinal dermal sinuses cases treated surgically in our setup over a period of 10-years. From January 2005 to January 2015, at King Hussein Medical Center. The mean follow up was 87 months (14-109 months).

Results: The study group included 53 consecutive patients who were managed in a single center. Male : female ratio was 28:25. Mean age was 3.4 years, ranging from one month to 15 years. The mean follow up was 87 months (14-109 months). Anatomically the majority of the treated sinuses occurred in lumbar region 37-cases, representing 69.8 % of cases. 83% progressed satisfactorily.

Conclusion: Spinal dermal sinuses are a rare entity that may progress to a serious neurological condition. Early detection and management have good outcomes. The Paediatrician has a crucial role in diagnosis. Surgery aims to excise the sinus tract completely, excisable skin tags, and correct the dysraphic state in the same setting.

Keywords: Congenital, Inclusion tumour, Spinal dermal sinus.

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Introduction

Spinal dermal sinus is a rare congenital tubular tract anomaly, product of the incomplete separation between the neural ectoderm and the epithelial ectoderm during the first gestational weeks. It is a variety of dysraphism that allows a potential communication between the skin and the central nervous system ⁽¹⁾ The inward extent of the tract traverses the underlying structures is variable, it may extend beyond the dura and terminate in the spinal cord or conus medullaris. It is located mostly in the midline, although apara-median location reported ^(2,3). It can originate at any level of the neuraxis,

from the first cervical vertebra to the coccyx, most frequently in the lumbar region. The cutaneous stigmata that accompany the dermal sinus are a small hole or dimple, sometimes almost imperceptible, it may harbor suppuration, which may be accompanied by cutaneous angioma, hypertrichosis or subcutaneous tumor. All of these findings are contemporaneous from birth ⁽⁴⁾ An inclusion tumor (dermoid cyst, Epidermoid cyst, or lipoma), can develop at any point of the length of the tract, more usually at the intra-dural level. These are produced by anomalous implantation of ectodermal cells during neural tube closure

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between the third and fifth weeks of embryonic life ^(4,5) As the intra-spinal portion of the sinus or the inclusion tumor is attached to the nervous structures, sinus ascends with them during the cephalic migration of the spinal cord, creating a long intra-dural trajectory between the cephalic end of the tract or cyst and the skin stigmata. Despite its harmless external appearance, they can produce serious neurological deficit and the treating physician has a crucial role in its early diagnosis. The objective of our work was to describe forms of presentation, diagnosis, and treatment of patients with dermal sinuses.

Methods

We carried out a retrospective analysis of the medical records of all the patients presented with spinal dermal sinuses cases treated surgically in our setup over a period of 10-years. The study group included 53 consecutive patients who were managed in a single centre, from January 2005 to January 2015. The purpose of this current study was to investigate the corresponding variables: age, sex, the reason for consultation, clinical manifestations, location, methods of diagnosis, type of surgery, histological diagnosis, presence of infection and subsequent evolution.

Results

In our series of 53 treated patients, male: female ratio was 28:25. Mean age was 3.4 years, ranging from one month to 15 years. The mean follow up was 87 months (14-109

months). All our patients presented with an orifice which was the major complaint, other presentations are shown in Table I. Anatomically the majority of the treated sinuses occurred in lumbar region 37-cases, representing 69.8 % of cases, 9 cases in the cervical region and 7 cases in the dorsal region. Topographic localization was compared to reported literature Figure1. Stigmata associated with the spinal dermal sinuses shown in Table II. The histological diagnosis was of pure dermal in 39.6% of cases (n =21), 26.4% (n = 14) were associated with epidermoid cysts, 17% (n = 9) with dermoid cysts and 17% (n = 9) with lipomas. All patients were clinically monitored with an average follow-up of 3.9 years, with MRI performed for follow up in those with surgically verified intra-dural component, inclusion tumor, or change in clinical picture. All evolved favorably with neurological examination, except 6-cases, that persisted with neurogenic deficit. Three cases developed para-paresis. The study group included 53 consecutive patients who were managed in a single center. Male: female ratio was 28:25. Mean age was 3.4 years, ranging from one month to 15 years. The mean follow up was 87 months (14-109 months). Anatomically the majority of the treated sinuses occurred in lumbar region 20-cases, representing 37.73 % of cases. 83% progressed satisfactorily.

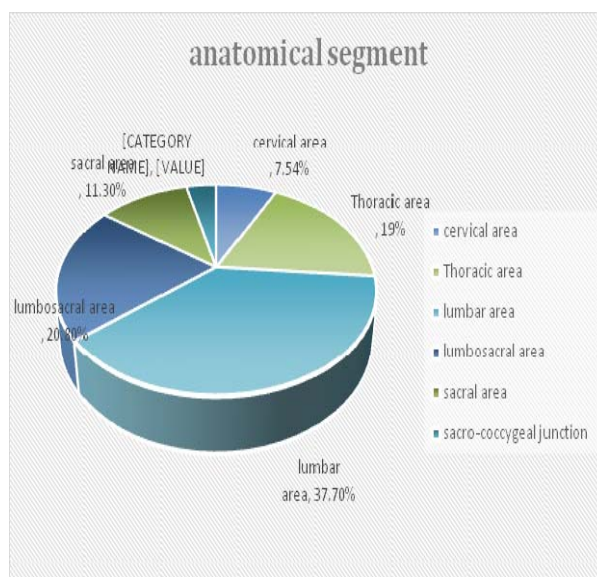
Table I : Clinical characteristics of all patients at presentation

Manifestations	Number of cases	Percentage
Orifice	53	100%
Asymptomatic	34	64.2%
Neurological deficits	19	35.8%
Infection	4	7.5%

Table II: Cutaneous findings associated with dermal sinus.

Findings	Patients	Percentage
Sinus ostea with a cephalically oriented tract	53	100%
Angiomata	5	9.4%
Hypertrichosis	15	28.3%
Skin tags	12	22.6%
Abnormal pigmentation	11	20.8%
Subcutaneous lipomas	8	15.1%

Figure.1:Topographic localization of spinal dermal sinus



DISCUSSION

The spinal dermal sinus is a rare entity; estimated incidence about one in every 2500 live births, it is always above the sacro-coxygeal level ^(4,7) It must be differentiated from another frequent entity; the sacro-coxygeal sinus, which is present in 2 to 4% of all newborns at the upper end of the fold and ends in the cases of maximum extension in the periosteum of the coccyx. These sinuses end blindly with no extension so it is not necessary to perform a radiological evaluation. The operation only needed if the background of the sinuses not displayed, to prevent future infections ⁽¹⁾ Many publications on spinal dermal sinuses, emphasized that despite their benign external appearance, these lesions are capable of producing serious neurologic deficits disease due to meningitis, spinal cord compression, and infected dermoid cysts, as occurred in a large part of our cases ⁽⁵⁻⁹⁾ Medical records of our patients with dermal sinuses were analyzed and revealed 53-cases. All had a cutaneous orifice in the mid line Figure 2 . Regarding the anatomical segment involved our results compared to what has been reported in the literature ^(3,10,15) Results are shown in the chart Figure1. In terms of radiological evaluation, cutaneous stigmata and, or congenital anomalies often prompt screening, in all patients. Plain radiographs of the spine performed as the primary imaging modality (where dysraphy was not always found). All patients underwent magnetic resonance imaging, while in 7cases computed tomography (CT) was performed to further



Figure 2: showing a lumbar spinal dermal Sinus Associated with massive lipoma

assess bone deformities ⁽¹⁶⁻¹⁹⁾ Magnetic resonance imaging showed the pathology, congenital anatomical abnormality and the tract in all cases Figure3. The surgery aimed to excise the sinus tract completely, excisable skin tags, and correct the dysraphic state in the same setting ^(2,20,21) Surgical steps were common for all the cases. The procedure included tracing, sinus tract through the subcutaneous tissue and muscle layer still its attachment with the neural axis. Tract excised completely. In cases where sinus was intradural, dura opened for further complete resection. Associated Intraspinal epidermoid, myelocele, pathologies; lipomeningomyelocele, abscess, arachnoid adhesion, tethered cord were dealt with accordingly ^(21,24) Those patients presenting with infectious complications were managed with appropriate antibiotics, after recovery surgery for resection of sinus, and correction of associated anomalies was performed. Our diagnostic and therapeutic algorithm specified in Figure 4 applied. Clinical outcome in most patients was excellent, 83% progressed satisfactorily regarding neurological examination, and on the other hand, 6-cases persisted with a neurogenic deficit. In addition, 3-cases developed para-paresis. Detailed clinical outcomes showed in Table III.



Figure 3: Thoraco-lumbar MRI, T2-wighted, sagittal view, showing a sacral spinal dermal sinus

Table III: Patient clinical outcome post-operatively

Patient Outcome	cases	percentage
Neurologically intact pre- and postoperatively	34	39%
Had complete neurological recovery	10	17%
Neurologically unchanged	6	11.3%
Neurologically worsened	3	5.7%

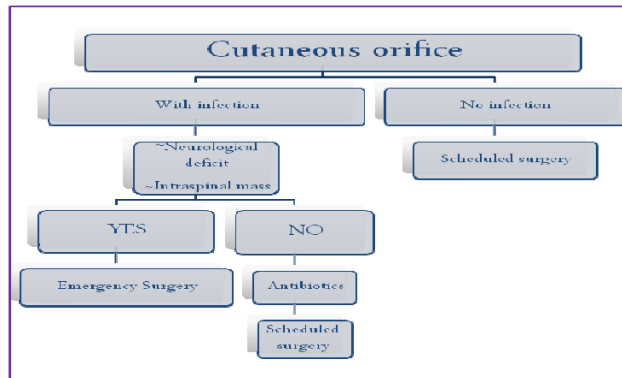


Figure 4: Diagnostic and Therapeutic Algorithm

Conclusion

Spinal dermal sinuses despite their rarity and innocent appearance, may progress to a serious neurological condition. Early detection and management have favorable results. The Pediatrician has a crucial role in diagnosis. The surgery aims to excise the sinus tract completely, excisable skin tags, and correct the dysraphic state in the same setting.

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