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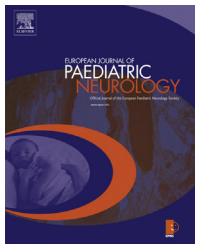

Original article

Spinal dermal sinuses and dermal sinus-like stalks analysis of 14 cases with suggestions for embryologic mechanisms resulting in dermal sinus-like stalks

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ABSTRACT

Background: Spinal dermal sinuses consist of an epithelium-lined tract extending from the skin towards the spinal cord, often resulting in infections or tethered cord syndrome. Recently, a variant called dermal sinus-like stalk was described as an analogous tract but not containing an epithelium-lined lumen.

Aims: We aimed to describe the findings in our patients, subdivide our specimens into both conditions, compare the characteristics of both groups and search for possible embryologic mechanisms of dermal sinus-like stalks.

Methods: We performed a retrospective analysis of all patients operated in our hospital for both conditions between 1996 and 2012.

Results: 14 patients were operated upon for spinal dermal sinuses ($n = 5$) and spinal dermal sinus like-stalks ($n = 9$). Patients were mainly referred from other hospitals due to skin abnormalities and were evaluated at mean age of 7 weeks and operated upon at mean age of 1 year and 2 months. Primary reason for referral was skin abnormalities in both groups, though there were two cases of meningitis in dermal sinus patients and 2 of recurrent urinary tract infections in dermal sinus-like stalk patients. Consistent with previous findings, dermal sinus-like stalk patients do not have a history of meningitis, lack dermoid or epidermoid tumours along their tract, and are histologically of pure mesodermal origin. Dermal sinus-like stalks might result from interposition of mesenchyme during primary or secondary neurulation.

Conclusions: We consider dermal sinus-like stalks as a rare but currently under diagnosed condition with different clinical, pathological and probably also embryologic characteristics compared to spinal dermal sinuses.

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1. Introduction

A spinal congenital dermal sinus tract (DST) is an uncommon form of spinal dysraphism, resulting in a cutaneous defect along the midline of the spinal axis. Its hallmark is a hollow epithelium-lined tract extending over a variable distance towards the spinal cord, often continuing to the intradural space, where it is mostly attached to the spinal cord.[1–4] Frequently dermoid or epidermoid tumours can be found over the course of these tracts.[1,4] Due to the open connection to the skin, infections are frequent, either superficial or deep with possible meningitis and/or abscess formation.[1,5] Dermal sinuses in the lumbar or lumbosacral region are usually seen in combination with low-positioned conus medullaris, and can be associated with tethered cord syndrome.[1,2,4]

In 1972 similar tracts but lacking an epithelium-lined lumen and dermoid or epidermoid tumours were given the name 'meningocoele manqué'.[6] It was not until 2009 that these tracts were reconsidered as being a histologically, clinically and probably also embryologically different entity and named 'dermal sinus-like stalks' (DSLS) by van Aalst.[7]

This study is a retrospective analysis of 14 cases of DST and DSLS that have been operated by the Neurosurgery department of the Leuven University Hospitals. The aim of our study was to describe the findings in our patients, to subdivide our specimens into DST and DSLS, to compare the characteristics of both groups and to search for possible embryologic mechanisms resulting in DSLS.

2. Methods

We retrospectively analysed the medical records of all paediatric (aged 16 years or less) patients with a clinical diagnosis of DST in whom intra-operatively a tract from the skin towards the spinal cord was seen between 01-01-1996 and 31-10-2012. Simple coccygeal pits and children that have not been operated upon in our hospital were excluded. Epidemiological and clinical data, imaging, surgical and pathological descriptions and follow-up data were collected and if necessary completed by telephone interrogation of the parents. Approval of the local ethics committee was obtained.

Skin abnormalities were systematically listed according to the study of Schropp[8] and urological abnormalities according to that of Radmanesh.[1]

All specimens were re-evaluated by a senior neuropathologist to detect the type of tissue and the presence of an epithelium-lined lumen within the tracts.

Statistical analysis was performed using SPSS Statistics 19.0 for Mac (IBM Corporation, Armonk, New York, United States). Unpaired Student's *t*-test, Wilcoxon two-sample signed rank tests and Fisher's exact tests were applied. A value of $P < 0.05$ was considered significant.

3. Results

3.1. Demographics

Fourteen patients were included, 6 boys and 8 girls. Mean age of birth was 39 weeks PMA (range 34–40 w). Mean birth weight was 3000 g (range 1900–5000 g), at P23 according tot the Flemish birth record.[9] Mean birth length was 49.0 cm (range 44.0–50.5 cm) and mean birth head circumference was 34.3 cm (range 32.0–36.5 cm). Half of the patients were born by a caesarean section due to various reasons, including foetal stress,[2] pelvic dystocia,[2] known spina bifida,[1] breech presentation[1] and the umbilical cord entanglement (Table 2).[1]

3.2. Referral

Mean age at time of first contact with the paediatric department was 7 weeks (range 7 d–8 y4 m). The primary reasons for referral were abnormalities of the skin (in 10 patients), recurrent urinary tract infections (in 2 patients) and meningitis (in 2 patients).

3.3. Skin abnormalities

Skin abnormalities are listed in Table 1. Skin lesions were located at the midline in 7 patients, leftward in 3 and rightward in 4. All lesions were in the lumbar or lumbosacral region. Of note, two patients had two dimples; one coccygeal "pit", and a more rostrally located porus. All patients had at least one skin abnormality, 11 had 2 or more (Figs 1a and 2a).

3.4. Clinical examination

On neurological examination reflexes and motor function were evaluated. Age of ability of independent sitting and walking was registered, as well as foot abnormalities such as pes cavus. Scoliosis (Cobb's angle $\geq 10^{\circ}$) and self-reported pain were noted.

3.5. Continence and urological examination

Urinary and faecal continence were asked for, and age of continence was noted. Technical examinations consisted of

Table 1 – Skin abnormalities in DST and DSLS patients.

Skin abnormality	DST ($n = 5$)	DSLS ($n = 9$)
Subcutanous lipoma	1	4
Skin tag	2	4
Deviation gluteal fold	1	4
Coccygeal pit	1	1
Porus	3	4
Skin dysplasia	1	0
Cigarette burn	0	2
Cutaneous hemangioma	0	1
Vascular naevus	1	2
Depigmented macula	1	0
Hairy patch	0	1
Hypertrichosis	1	1