Letters to the Editor

NEUROSURGICAL FORUM

Congenital spinal dermal sinus

TO THE EDITOR: I read with the greatest interest the authoritative article by Tisdall et al.8 (Tisdall MM, Hayward RD, Thompson DNP: Congenital spinal dermal tract: how accurate is clinical and radiological evaluation? J Neurosurg Pediatr 15:651–656, June 2015). The authors report on 74 patients diagnosed with congenital spinal dermal sinuses who were submitted to operation and to histopathological verification of the tracts in all instances.8 They report clinical and radiological findings with the aim of assessing their value for diagnosis. The removed tracts were described as consisting of variable combinations of epidermal, dermal, meningeal, and neural tissues, with the relative proportion of cutaneous and neural elements being what differentiates the stalk of a dermal sinus tract (DST) and the tracts of so-called limited dorsal myeloschisis as defined by Pang et al.4 This last entity refers to a distinctive form of spinal dysraphism characterized by a focal closed midline defect with a fibroneural stalk that connects the skin lesion with the underlying spinal cord.4 As suggested in previous papers from our institution,2,3 it appears that there is some confusion in the current literature for differentiating these 2 entities (spinal DST and limited dorsal myeloschisis) and, consequently, they are currently documented as mixed together in many reports.3,5,6 The distinction we propose is not only of semantic or academic interest, especially from a practical point of view.

With the Editor’s kind permission we would like to introduce some reflections on the importance of differentiating these 2 conditions. Our aim is not to detract from the authoritative work of Tisdall et al. but to respectfully draw their attention, and that of the journal’s readers, toward the significance of both types of congenital spinal tracts because we believe this difference is of the utmost interest, especially from a practical point of view.

In 2011, we published a short series of 20 cases involving patients who shared the characteristic of presenting with a midline skin dimple or orifice on their back.3 We classified the patients into 2 groups: Group 1 comprised 8 patients with “true DST,” and Group 2 (n = 12) included patients with cutaneous dimples resembling DST that we called “pseudo-DST.” The classification we proposed arose from the direct observation (with the surgical microscope) of the spinal stalks during the operations, and it was supported by histological verification performed in all instances.

The following features contributed to categorizing these 2 groups:

1) Patients with true DSTs had a hollow tract that could be identified during the surgery, with a lumen that was proven histologically to be lined by squamous epithelium. In addition, many instances of true DST contained or ended in a dermoid cyst. In contrast, the stalk in cases of pseudo-DSTs appeared as solid during the surgical exploration. Microscopically, these solid tracts were mainly composed of fibrous tissue, although it might be intermingled with epidermal elements and with fat, nerve bundles, and meningeal or neural remnants.

2) The skin opening in a true DST usually consisted of a minute pore, sometimes surrounded by a superficial cutaneous angioma. These tiny orifices may exude caesous debris, hairs, or purulent material. In contrast, the cutaneous opening of the pseudo-DST was more complex and usually consisted of a dimple, crater, congenital scar, or bullous lesion that almost invariably was covered by a thinned or translucent membrane.

3) From all of the above, it is evident that true DSTs may be complicated by an intraspinal infection of variable severity, as they represent a pathway for communication with deep structures and may give rise to meningitis, subcutaneous or intraspinal abscesses, or arachnoiditis, as happened in 7 of our 8 cases.

4) In contrast, none of our patients with pseudo-DSTs developed an infectious complication. Pseudo-DSTs usually were brought to our attention either by the presence of the cutaneous mark or by the clinical manifestations of spinal cord tethering.

5) In our opinion, cases of true DSTs ought to be managed with prompt evaluation and surgery, while cases of pseudo-DSTs can be managed more conservatively and surgery can be appropriately scheduled.

The existence of true DSTs and pseudo-DSTs as different entities has also been supported by other researchers’ work.10 There has also been some discussion about the nomenclature referring to these tracts.1–3 Van Aalst et al.9 suggest using the term “spinal dermal-sinus-like stalk,” while Pang et al.4 prefer using the name “limited dorsal myeloschisis.” Other terms to designate the stalks of pseudo-DSTs include tethering bands, hamartomatous...
or meningotheelial tracts, rudimentary meningocele, and meningocele manqué.2–4,7 We suggested using the term “pseudo-DST” for denoming these solid tracts.3

We agree with the comments of Dr. Tisdall et al. regarding the difficulties in the use of MRI for visualizing the trajectories of both types of tracts, especially in their intradural course, and also for depicting the associated dermoids when they exist.8 The intravenous administration of contrast medium during MRI studies seems to be of help for depicting signs of infection and for visualizing the coexistent dermoid cyst.

The authors comment that there is no animal experimental investigation regarding the development of these congenital tracts.8 For completeness, we would also like to draw attention to the existence of the work of van Aalst et al. on the experimental production of spinal-dermal-sinus-like tracts in a chick embryo model.10

Finally, we wish to remark that it is not the name that matters most but the concepts underlying the true clinical significance of these lesions. From our limited experience, it seems advisable to proceed promptly in instances of true DSTs and we suggest inclining the balance toward the side of surgical exploration in cases in which, after diagnostic work-up, a reasonable doubt about the true nature of the tract persists.

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DISCLOSURE
The author reports no conflict of interest.

References

Response
We thank Dr. Martínez-Lage for his interest in our article. We are of course mindful of his publication and its contribution to the understanding of the subject.1

We entirely agree that the pathological entities previously encompassed under the term “dermal sinus tract” are perhaps more diverse, both in clinical presentation and prognosis, than previously realized. It is generally thought that while incomplete dysjunction of the cutaneous from the neuroectoderm underpins this group of disorders, the histological constitution of the connecting tract is variable, and this has implications for mode of clinical presentation, prognosis, and indeed, nomenclature.

We would also agree that the presence of an obvious cutaneous punctum, particularly one that has leaked CSF or has been responsible for an episode of local infection or bacterial meningitis, is a clear indication for surgery.

In Dr. Martínez-Lage’s paper, the primary discriminator was the infection risk: he observed that only the 8 patients who had presented with superficial or deep infection had tubular epithelium-lined tracts and furthermore, that all of these had a characteristic cutaneous signature comprising a small punctum or opening. These were termed true DSTs to distinguish them from the 12 patients who had solid, fibrous tracts and a translucent, meningeal cutaneous opening; the latter were termed pseudo-DSTs.

In our experience we have not observed such a distinct clinicopathological delineation. Some patients with a “true punctum” had tracts that were tubular and epithelial in their cutaneous course but gave way to a more solid, fibrous core in the deeper layers. Furthermore, we have observed epithelial elements in cases in which the cutaneous signature was more akin to the meningeal cutaneous opening or “cigarette burn” lesion. Similarly, while we have observed dermoids more commonly in the context of DSTs that have been infected, this was not exclusively the case, and dermoid formation occurred in both types of cutaneous lesions.

It is perhaps pertinent to emphasize that infection is not the only pathophysiological mechanism of neurological deterioration for this group of patients. Dr. Martínez-Lage reported a low conus in 11 of their 12 pseudo-DST patients and neurological or urological abnormalities in over half of this group. Similarly, we too have observed neurological deficits and the emergence of a “tethered spinal cord syndrome” in children with a cutaneous signature more in keeping with pseudo-DSTs.

One of the findings of our study was the apparent limitation of MRI in detecting not only the extent of the tract but also the presence of inclusion cysts along the course of the tract. Thus we would caution against relying too heavily on imaging criteria in deciding whether or not to offer surgery.

Dr. Martínez-Lage makes the point that the distinction between pseudo-DST and true DST cannot be made from superficial examination of the cutaneous abnormality and instead requires “direct observation (with the surgical microscope) … during the operations … supported by histological verification.” As such, this distinction can only be made after the decision to pursue an operative course has