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, as we consider that these 2 types of tracts behave differently and that the distinction justifies a different clinical and surgical approach.

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 comprehensive 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-dermal sinus tracts."³ 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 caseous 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.^{1,7,9} 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 meningotheial tracts, rudimentary meningocele, and meningocele manqué.^{2-4,7} We suggested using the term “pseudo-DST” for denominating 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.

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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, membranous 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 membranous 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

been taken and may therefore be of limited value in deciding on a management strategy.

In summary, we fully acknowledge Dr. Martínez-Lage's argument that the true tracts likely represent a greater risk for infective complications. However, in recognizing that both types of tracts are clearly capable of causing neurological damage (albeit by differing mechanisms) and, in acknowledging the limitations of MRI, and clinical examination, in anatomical delineation of the lesion, we believe the arguments lie in favor of surgical exploration with excision of the tract and untethering of the spinal cord in all cases. Surgery, particularly in those children with minimal or no deficit at the time of the operation, can be carried out with low risk and with extremely favorable prognosis.

Our current policy is to follow these children until such time as normal or stable motor function can be demonstrated and normal continence is attained and then discharge them from follow-up. If a nonoperative policy were to be pursued then children would, at the very least, require ongoing clinical and perhaps radiological surveillance and would additionally incur the risks of late complications—of not only infection but also of tethering and inclusion cyst formation.

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Onchocerca lupi infection

TO THE EDITOR: It was with great interest that we read the paper by Chen and colleagues² (Chen T, Moon K, deMello DE, et al: Case report of an epidural cervical *Onchocerca lupi* infection in a 13-year-old boy. *J Neurosurg Pediatr* 16:217–221, August 2015). Our understanding of this complex emerging pathogen is evolving, and this case report adds to that understanding. However, we are concerned by several points made by the authors that could be misleading or wrong. The first statement of concern is that *O. lupi* is the only species other than *O. volvulus*, the parasite that causes river blindness, that has been reported to infect humans. This is incorrect, as several other zoonotic species have been reported to infect humans.⁵ It is, however, the only zoonotic onchocercal infection reported to have involved the spine. This report increases the number of patients with spinal involvement to three.^{4,5}

Later in the paper the authors discuss options for medical therapy for the parasite. They state that doxycycline therapy was not effective in their patient because re-exploration and debulking of the original mass was required when it became inflamed 10 weeks after the initial surgery. They mention the case of the 22-month-old girl who was treated with doxycycline followed by ivermectin. First, to the best of our knowledge the girl was not treated with doxycycline.⁵ Second, recommendations for medical therapy for *O. lupi* are based primarily on what is known about the treatment of *O. volvulus*. Based on what is known, it would be premature to make any statement on the efficacy of either therapy. Ivermectin kills the microfilariae (juvenile form) of *O. volvulus*, preventing the symptomatic manifestations of river blindness and reducing the probability that black flies will become infected and maintain the cycle of transmission.¹ There are limited data that treating an individual with ivermectin 4 times a year will accelerate the sterilization and death of the adult female parasites; however, this effect takes up to 5 years or more.³ Doxycycline is known to kill the adult form of *O. volvulus* because of its effects on the *Wolbachia* endosymbiont, which is required for reproduction and the long-term survival of the parasite.⁶ However, this effect takes 21–27 months.^{6,7} The patient discussed by Chen and colleagues has not been on ivermectin therapy long enough for it to have resulted in the death of adult parasites, nor has enough time elapsed to determine the efficacy of doxycycline. Because we do not currently have a noninvasive test, medical therapy is primarily directed at the prevention of symptoms due to parasites not found in the resected biopsy.

Although Chen and colleagues state that they believe their patient may have been reinfected, there are other potential explanations. One possible explanation for what occurred could be that the biopsy killed the adult parasite(s) in the nodule but did not result in complete resection of it (them). The expected inflammatory response to the dead parasite(s) may have been delayed by the use of dexamethasone, so the inflammation surrounding the dead parasite(s) did not become symptomatic until 10 weeks after the initial surgery. Importantly, the case report illustrates that there may be a role for corticosteroids in patients with spinal nodules due to *O. lupi*, although more information will be needed to determine if their use can reduce the need for additional surgical interventions.

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DISCLAIMER The findings and conclusions in this report are those of the authors and do not necessarily represent the views of the CDC.

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Response

We appreciate the insightful comments and expertise of Dr. Cantey and colleagues on this topic. It is correct that the 22-month-old girl who was also infected with *Onchocerca lupi* was, in fact, initially treated with albendazole (rather than doxycycline) for 12 days before being transitioned to ivermectin.² Thank you for raising this point. It is also true that the treatment of spinal *O. lupi* infection is based on what we know of treating human *O. volvulus* infections from the limited number of reported cases documenting such medical therapy. Given the delayed effect of doxycycline, it is not surprising that our 13-year-old male patient had local disease recurrence upon cessation of the therapy after a 6-week course of treatment.^{1,3,4} This recurrence reinforces the need for long-term medical therapy, although the ultimate duration of treatment remains unknown.

We agree that multiple explanations could be possible

for the symptomatic disease recurrence in our 13-year-old male patient. Dr. Cantey and colleagues suggest that resection potentially did not result in complete removal of the adult parasite(s), and that delayed inflammation surrounding these parasites may have become symptomatic once the patient was taken off corticosteroids. Although this is a possibility, it would be unlikely for the enhancing lesion to have increased significantly in size, as seen on the patient's MRI scan, secondary to inflammation alone. Purulence and strandlike material were observed intraoperatively and were subsequently confirmed pathologically to be *O. lupi*. Corticosteroids are essential in the perioperative period to reduce the effects of inflammatory products released into the CSF; however, corticosteroids are unlikely to be a substitute for surgical intervention if a recurrent lesion has increased in size, demonstrated mass effect on the spinal cord, and/or caused new neurological deficits.

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