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LETTER TO THE EDITOR

Pseudo-dermal sinus tract or spinal dermal-sinus-like stalk?

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Dear Editor,

We read with great interest the paper "Spinal dermal sinus and pseudo-dermal sinus tracts: two different entities" by Martinez-Lage et al. [1]. In fact, the term "sinus" may be used for (chronically) infected tracts connecting a deep-seated infection to the skin, and discharging pus to the surface, except for those tracts connecting two epithelialized surfaces, which are called "fistula".

A spinal dermal sinus (DS) is a developmental anomaly of the dorsal midline axis, in which a hollow, epithelium-lined tract extends inward from the skin surface for a variable distance [1–3]. A key feature of a DS is an open skin defect; however, this may be minute and, as such, easily overlooked during routine neonatal examination [1]. Occasional discharge from this orifice is a common finding, however, even in case of connection with the spinal cord, this never is cerebrospinal fluid [3, 4]. Another key feature of a DS is the accumulation of (epi)dermoid remnants, often presenting as tumors inside the tract or even inside the spinal cord. Such tumors may produce manifestations of spinal cord or cauda equina compression [1]. Moreover, due to an open connection with the skin surface, many cases present with meningitis and intraspinal, intramedullary, or more superficial intra-epidermoid abscesses, a potentially life-threatening condition, rather than with cord tethering (recurrent cases no longer connected to the skin surface) [1, 3].

A spinal dermal-sinus-like stalk (DSS) [4] or pseudo-dermal sinus tract as proposed by Martinez-Lage et al. [1] is a developmental anomaly of the dorsal midline axis, in which a solid tract extends outward from the intradural space to the skin. Key features of a DSS are a closed skin defect (a dimple, cigarette burn, or blister, all without an orifice) usually more obvious than in case of a DS, the absence of a lumen, and the absence of (epi)dermoid remnants inside the tract [1, 4]. Consequently, a DSS will never present with meningitis, but rather with cord tethering [1, 4]. To illustrate this point, we have included several photographs highlighting the essential surgical steps in case of a DSS, which, in this particular case, was attached to a low-lying conus medullaris (Fig. 1a–f).

In case of a DS, stratified squamous epithelium inside the lumen, surrounded by dermal tissue, suggests a nondisjunction of cutaneous and neural ectoderm with inward dragging of the epidermis. In case of a DSS, however, mesenchymal and neural elements inside the stalk suggest an embryologic development in the opposite direction (inside-out rather than outside-in). This would explain the absence of a skin orifice, the occasional dorsal tenting of the dura, and the presence of glioneural tissue inside the stalk. Thus, a DSS would represent an atrophic mesodermal-neural stalk [4] and, as suggested by Martinez-Lage et al., may indeed represent the spinal counterpart of an atretic encephalocele [1].

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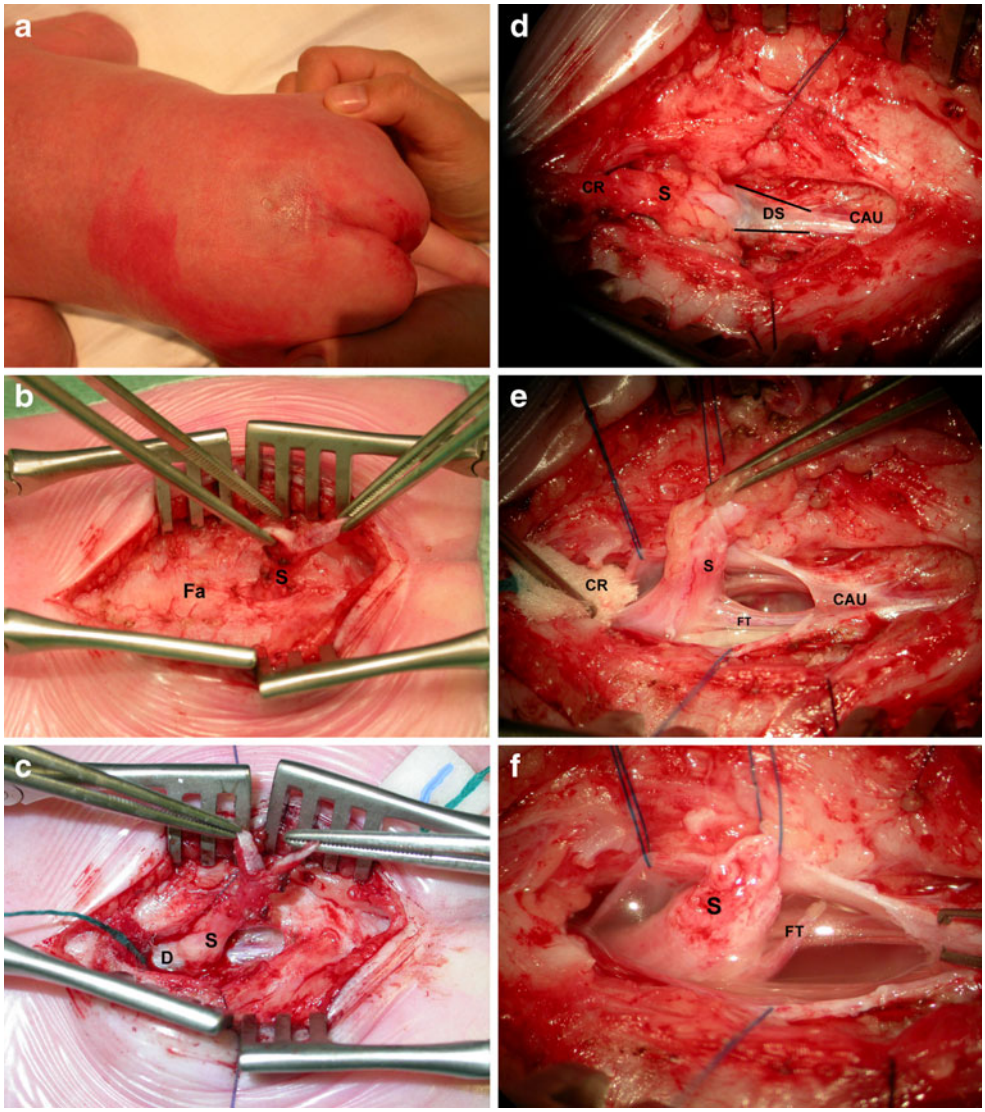

Fig. 1 **a**–**f** Illustrates a typical DSS case born at a time DS and DSS were not yet recognized as different entities. The child was scheduled for early operation which, for unrelated causes, was deferred until the age of 3 months. Essential surgical steps are illustrated. *CAU* caudal, *CRA* cranial, *D* dorsal, *DS* dural sleeve, *Fa* lumbar fascia, *FT* filum terminale, *S* stalk. **a** Skin defect (dimple) typically seen in case of a DSS: above the gluteal fold, in the midline, and without an orifice. **b** Dimple and surrounding skin are excised, and the attached stalk is followed through the lumbar fascia to its ending, which in case of a DSS should be intradurally. **c** The stalk is followed to the point where it penetrates the dura. **d** When the stalk is reflected cranially, the dural sleeve surrounding the stalk where it penetrates the dura is seen. **e** A thick, fibrous stalk, surrounded by some extradural fat, is attached to the conus medullaris (in this particular case close to the junction with the filum terminale). **f** The stalk has been cut approximately 1 cm below the lowest nerve roots branching off the spinal cord. The filum terminale has been cut as well. Histological examination revealed a solid stalk composed of connective tissue with some intermingled nervous and muscle tissue

In conclusion, DS and DSS are clearly different clinico-pathological entities that share some common cutaneous and neuro-imaging findings [1, 4]. The former requires (semi) urgent surgical correction mainly to prevent infection, the latter allows calm clinical evaluation and surgical correction to release tethering [1, 4] or whenever in doubt about its true nature. Martinez-Lage et al. have made an excellent contribution to the literature, giving a detailed description of 8 children with a DS and 12 children with as DSS. However, as long as neuroradiologists, clinicians, and pathologists use different eponyms to describe both entities [1], the confusion will continue, and affected patients may be given inappropriate care. Therefore, we strongly advocate to use a uniform nomenclature, and suggest the term spinal dermal sinus (a true sinus), and spinal dermal-sinus-like stalk (a stalk rather than a sinus) [4].

References

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