

Dermal Sinus Tract associated with Type I and Type II Split Cord Malformation

Abstract

The dermal sinus tract of the spine is associated with other occult spinal dysraphisms, such as the split cord malformation (diastematomyelia) in a 40% of the cases and embryologically is not clearly defined if the dermal sinus and split cord malformation have origin in gastrulation or late primary neurulation, but the most accepted theory of the dermal sinus tract consists in early incomplete disjunction, which explains the relation with other spinal dysraphisms. Here, we present two cases, with a dermal sinus tract of the spine associated with Type I and Type II split cord malformation.

Keywords: *Dermal sinus, diastematomyelia, dysraphisms*

Introduction

The dermal sinus tract of the spine is a rare spinal dysraphism which occurs in approximately 1 in 2500 newborns.^[1,2] The dermal sinus tract includes a stratified squamous epithelium, with variable depth, and it can end in any structure including the dural sac.^[3,4] The dermal sinus tract is associated with other occult spinal dysraphisms, as demonstrated by Gupta *et al.*, that found an association of 11.34% with other dysraphisms.^[5] The dermal sinus tract is associated with split cord malformation in 40% of cases.^[4,6] In addition to this, terminal syringomyelia is associated with split cord malformations although its incidence and natural history are not very clear.^[7]

We present two cases with dermal sinus tract associated with split cord malformation.

Case Reports

Case 1

A 10-month-old male presented with a delay in motor development and abnormal findings involving lower limbs.

At physical examination, him was noted a dorsal column deviation with left convexity and lumbosacral bluish spot, a thoracic midline cutaneous appendix at T10 level and another it at the right knee, in the left

foot with presence of paraxial polydactyly by duplication of the first toe and in the right foot with third and fifth toe hypoplasia and absence of the fourth toe with appendix skin replacing it. Anal sphincter tone was decreased, and lower limb distal flaccid paraparesis was found [Figure 1]. Spine X-ray [Figure 2] and magnetic resonance imaging (MRI) were made and showed a dermal sinus tract with Type II split cord malformation [Figure 3]. Somatosensory evoked potential studies has identified compromise of the motor and sensory pathway in the lower limbs.

Thoracic dermal appendix was identified at the T10 level, and laminectomy was performed at that level, finding the dermal sinus tract termination at the dural sac. Dura was opened observing arachnoid adhesions in relation to fibrous septum formed in the dermal sinus tract causing split of the spinal cord into two parts. The septum and the dermal sinus tract were removed, and sent to pathology studies, added to the release of spinal cord adhesions [Figure 4]. Postoperatively, MRI was performed [Figure 5] and pathology report showed a stratified squamous epithelium.

In the patient's postoperative follow-up, distal and sensitive motor deficit was identified, showing sphincter involvement.

Juan Esteban Muñoz Montoya, Marcial Anaya Jara¹, María Paula Vargas Osorio, Fernando Rueda Franco²

Departaments of Neurosurgery and Neurosciences, Hospital Militar Central, Universidad Militar Nueva Granada, Bogotá, Colombia, ¹Departament of Pediatric Neurosurgery, Instituto Nacional de Pediatría, ²Departament of Pediatric Neurosurgery, Chief of the Surgery Division, Instituto Nacional de Pediatría, Mexico City, Mexico

Address for correspondence:

Dr. Juan Esteban Muñoz Montoya, Departament of Neurosurgery, Hospital Militar Central, Universidad Militar Nueva Granada, Bogota, Colombia. E-mail: juanesteban1285@gmail.com

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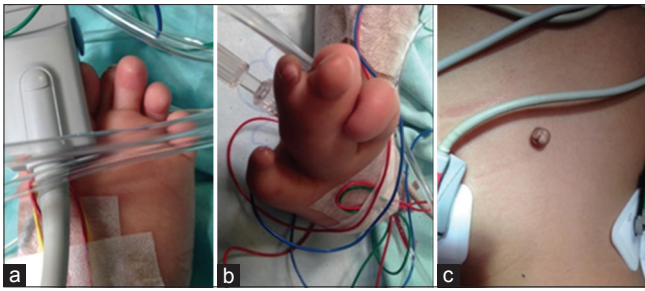


Figure 1: (a) Left foot polydactyly. (b) Third and fifth toe hypoplasia and absence of the fourth toe, (c) T10 dorsal column dermal appendix

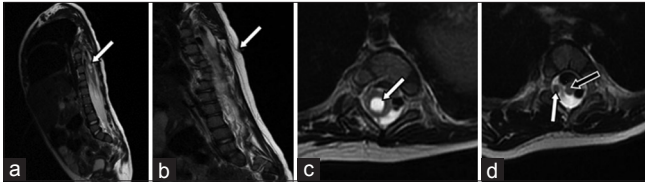


Figure 3: (a) Sagittal T2 magnetic resonance imaging sequence showing terminal syringomyelia (arrow), (b) Sagittal T2 sequence revealing dermal sinus tract (arrow). (c) Axial T2, syringomyelia is identified with dilation of the spinal cord's central canal (white arrow). (d) Axial T2 white and black arrows, double spine cord

Case 2

One year old female presented a thoracolumbar protrusion which it has grown progressively, associated with sphincter compromise and lower limb weakness. Physical examination revealed a soft mass in the thoracolumbar region measuring 12 cm in diameter, with the presence of an dimple at its center [Figure 6], sphincter involvement, flaccid paraparesis, and bilateral equinovarus foot.

Somatosensory evoked potential studies has identified compromise of the motor and sensory pathway in the lower limbs. MRI [Figure 7] showed a dermal sinus tract with type I split cord malformation, associated with intraspinal lipoma and a lobulated intralesional arachnoid cyst.

Surgery was performed for resection of the dermal sinus tract, opening of dural cavities, and resection of the osseous septum in relation to Type I split cord malformation [Figure 8]. Stratified squamous epithelium was confirmed by pathology. The patient after 1 month of follow-up dies from aspiration pneumonia.

Discussion

Split cord malformations (diastematomyelia) and dermal sinus tract belong to the group of closed spinal dysraphisms, which are very heterogeneous, and they usually appear with cutaneous marks identified at birth in more than 50% of the cases such as skin appendages, hypertrichosis, lipomas, hemangiomas, or infection signs located at the spinal dysraphism site.^[8]

Dermal sinuses can affect any neuraxis level, from the occipital to the sacral region, <1% affecting the cervical region, 10% thoracic, 40% lumbar, 12% lumbosacral,

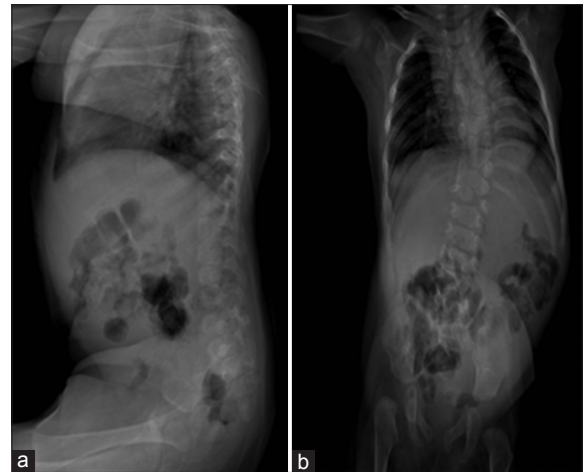


Figure 2: (a) Simple X-ray showing lumbar spine rectification. (b) Scoliotic curve with left convexity, butterfly-like vertebrae at T3, T4, T5, T6, T8, T11, L3, and T10 hemivertebrae

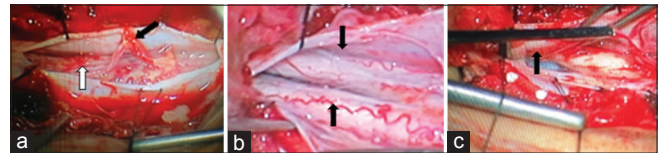


Figure 4: (a) Dermal sinus tract with termination in the dural sac (black arrow) and arachnoid adhesions (white arrow). (b) Opening of the dura with two split cord (black arrows) and (c) fibrous septum dividing spinal cord into two parts with unique dural sac (arrow)

23% sacral, and 13% sacrococcygeal.^[9] In addition, its tract can end in soft tissues in the superficial dorsal region (6%–7%), in epidural space (10%–20%), and the great majority (58%–60%) in the dural sac, of these 50% can develop adhesions to the conus medullaris, terminal filum, and cauda equina.^[1,9,10]

Embryologically, the dermal sinus tract origin is not clearly defined, but the most accepted theory consists in the incomplete early disjunction caused by a premature closure of the cutaneous ectoderm, and therefore, a permanent tract is formed between the skin and the neural tube, which occurs in late primary neurulation and in gastrulation phases.^[11,12] In split cord malformation, the defect is generated in the gastrulation phase, in which an endomesenchymal tract is produced by an accessory neuroenteric canal between the amnion and the yolk sac, that divides the notochord explaining the probable connection between split cord malformation with the dermal sinus tract.^[10,12,13]

Split cord malformation is a rare form of spinal dysraphism, which is divided into Type I: A fibrocartilage or bone spur divides the spinal cord and Type II in which there is a fibrous or cartilaginous tissue that divides the spinal cord; finding a female predominance,^[13,14] most of them are located in the thoracic and mid-thoracic upper regions and are associated with skin abnormalities and other spinal dysraphisms, including the dermal sinus tract.^[13,15] It has

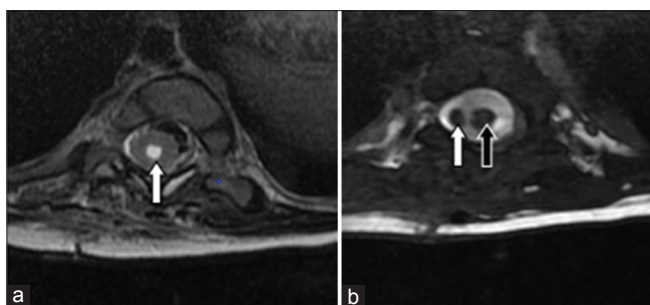


Figure 5: Postoperative magnetic resonance imaging. (a) Syringomyelia decrease with respect to the previous (arrow). (b) Two spinal cords with white and black arrows) without adhesions and without fibrous septum (white and black arrows)

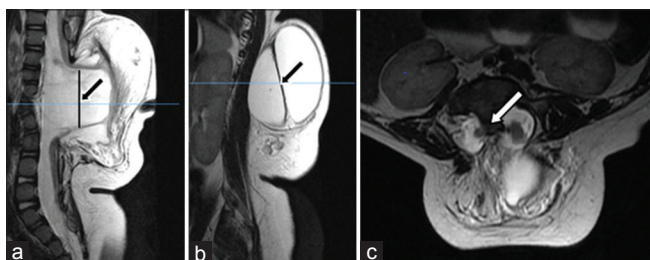


Figure 7: Thoracolumbar spine magnetic resonance imaging. (a) T2 sequence sagittal section identifying complex occult spinal dysraphism with absence of the posterior arch from T10 to L2 (line and black arrow) and spinal cord protruding out the spinal canal, underlying to the dermal sinus tract, (b) T2 sequence, sagittal view which evidences a septum dividing the dural sac in two cavities (arrow), (c) T2 sequence, axial section: Evidence of bone septum that divides both spinal cords (arrow)

been associated with low conus medullaris implantation in 83% of cases in addition to a thickened terminal filum.^[13] Syringomyelia is an associated finding that has been found in 29%–55% of the cases^[13,16] and tethered cord.^[17,18]

For the diagnosis of occult spinal dysraphisms, MRI is fundamental because it allows to adequately characterize them, as well as to detect associated malformations.^[4,19]

For surgical management, it consist on the exploration of the dermal sinus tract until the dural sac, since there are several reports, showing that incomplete exploration could finish in neurological deterioration which can be observed after surgery, with the need to reexploration it.^[1] In addition, arachnoid adhesions have been found intraoperatively, which contribute to tethered spinal cord in 33.3% of cases.^[1]

In split cord malformation, it has been hypothesized that bone spur resection could prevent tethered spinal cord and duraplasty could contribute to an improvement in the circulation of cerebrospinal fluid and it would also lead to resolution of the terminal syringomyelia.^[10,18]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The

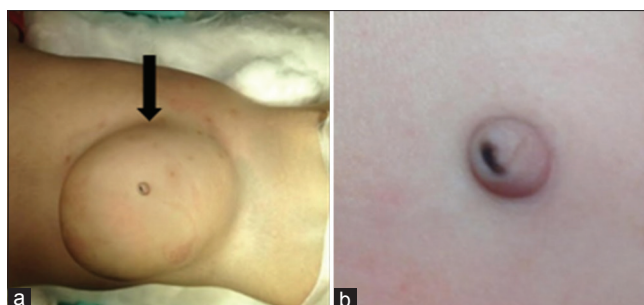


Figure 6: (a) Subcutaneous thoracolumbar 12 cm mass (arrow). (b) Dermal sinus in the thoracolumbar mass upper region

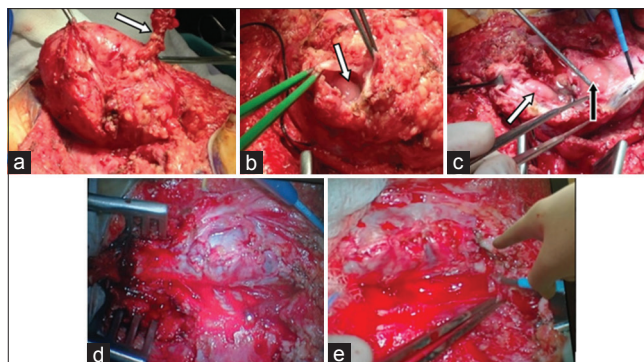


Figure 8: (a) Thoracolumbar subcutaneous mass dissection identifying dermal sinus and dermal sinus tract (arrow). (b) Subcutaneous mass opening and which to identify a cord inside. (c) Each mass with a spinal cord (white and black arrows). (d) Dural reconstruction was performed in a single sheath. (e) A bone spur distal to the dural closure was identified (arrow)

patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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