Conflicting Diagnosis of Dermal Sinus Tract and Tethered Cord

Chris J. Hong, Saleh A. Almenawer, Boleslaw Lach, Nina Stein, Benedicto Baronia, Sheila K. Singh

Can J Neurol Sci. 2013; 40: 889-891

Dermal sinus tracts (DSTs) are an uncommon form of occult spinal dysraphism that is attributed to incomplete neural tube closure during fetal development. Dermal sinus tracts are found along the midline neuroaxis from the nasion to the coccyx, but they most commonly appear in the lumbar region.[1] Dermal sinus tracts are more commonly associated with other developmental abnormalities such as skin tags, naevi, spinal dermoid cysts, meningocoele, lipomas and spinal cord tethering, and can be complicated by cerebrospinal fluid drainage, shedding of keratin from the epithelialized tract, and infection such as meningitis.[1]

We report a case of a radiologically diagnosed DST associated with multifocal dermal and subcutaneous hemangiomas. Surgical intervention demonstrated that despite high-resolution magnetic resonance imaging (MRI) showing intradural extension of the tract to the conus, intradural exploration revealed no such tethering tract. This unusual condition was successfully managed by surgical excision of pathological tissues.

Case Report

A 7-month-old girl was taken to her pediatrician for a routine "well baby" visit. Comprehensive history and examination were unremarkable in all aspects including her developmental milestones, limb movements, reflexes, and bowel and bladder function. However, the attending pediatrician confirmed a circular, one-centimeter skin defect over her mid-lumbar spine, larger than a typical dimple, which was identified by her parents two days after birth. The defect was surrounded by a reddish skin discoloration about four to five centimeters in diameter with irregular borders. The parents reported that the lesion had never leaked fluid nor discharged any material. The child was referred to the Pediatric Neurosurgery Clinic for further assessment of potential developmental abnormalities. Neurological examination revealed no abnormalities, and a MRI of the spine was undertaken.

The MRI study was performed as a routine spinal study with T2 weighted images with sagittal views of the whole spine complemented by additional axial T2 weighted images with fat saturation, axial T1 weighted images and axial Constructive Interference in Steady State (CISS) images of the lumbar and sacral spine, in attempts to better visualize the tethering structure identified in the spinal canal posterior to the cauda equina at the level of the spinal defect. Intravenous contrast was not used in this case as per routine protocol.

The MRI study revealed six lumbar-type vertebral bodies, and a thick T1 hypointense tract that originated at L5-L6 level suggestive of dermal sinus tract, with apparent intradural extension and termination at the tip of the conus, which was low-lying at the inferior end plate of L3 (Figure 1A-C). This tract demonstrated an unusual mild increased T2 signal. The sagittal T2-weighted images showed a conglomeration of small tortuous vessels surrounding this thick tract in the subcutaneous region. A small posterior midline defect was seen in the posterior aspect of the L5 spinous process. Otherwise, the vertebral bodies appeared normal in signal intensity and height at all levels, with unremarkable intervertebral disc spaces and posterior alignment. Furthermore, no evidence of fluid collection or fat was noted. There was also no evidence of syrinx or Chiari I malformation.

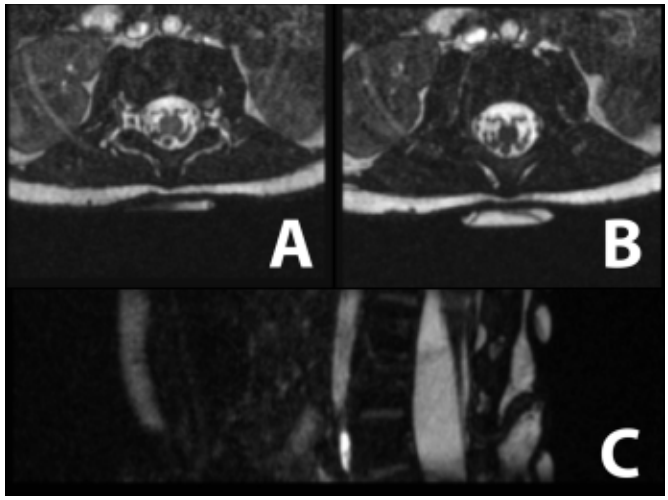

Figure 1: *Axial T2 MRI view showing intradural extension of DST that originated at L5-L6 level (A) and tethering of conus by DST (B). Sagittal MRI view also demonstrated intradural extension of DST and termination at the tip of conus, which was low-lying at the inferior end plate of L3 (C).*

From the Bachelor of Health Sciences (Honours) Program (CJH), Department of Pathology & Molecular Medicine (BL), Department of Radiology (NS), Division of Neurosurgery (SAA, BB, SKS), McMaster University, Hamilton, Ontario, Canada.

Received April 25, 2013. Final Revisions Submitted May 16, 2013.

Correspondence to: Saleh A. Almenawer, Division of Neurosurgery, Department of Surgery, McMaster University, 47 Caroline Street North, Suite 503, Hamilton, Ontario, L8R 2R6, Canada. Email: Dr_menawer@hotmail.com.

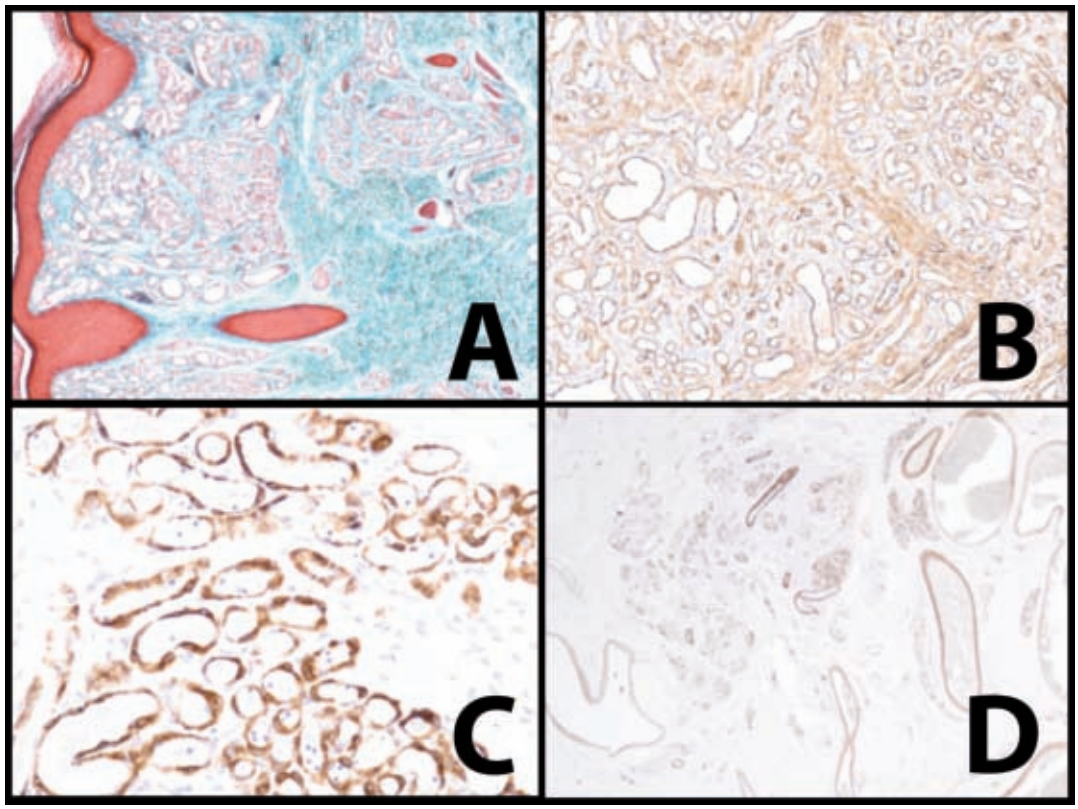

Figure 2: *Low power view of subepidermal collections of capillaries separated by keratinized ridges and collagenized dermis. Gomori trichrome elastic stain (A). The endothelium showed strong immunoreactivity for CD31 (B) and the subendothelial cells showed strong immunoreactivity for alpha smooth muscle actin (C). Low power view of deeply seated subcutaneous hemangioma with large arterial and venous vessels (D).*

An operative decision was made to untether the cord, prevent possible future cerebrospinal fluid leak, and to prevent meningitis. Surgical excision of the hemangioma and dermal sinus tract was recommended to the patient's family based on the radiological findings. In the operating room, after excision of abnormal cutaneous elements of the lesion, a fibrous tract was followed to a bony opening in the L5 lamina, and L3-L5 patital laminotomies were performed to allow for intradural exploration up to the level of the low-lying conus. However, the dermal sinus tract ended at the dura, and no intradural tethering bands or connection to the conus were identified. The patient made an uneventful recovery and was discharged three days after the surgery. All developmental milestones were met in the subsequent months of follow-up, with no new neurological deficits identified.

The grossly described area of skin elevation revealed multifocal nodular collections of tightly packed, occasionally dilated capillaries distributed throughout the dermis (Figure 2A). The lining endothelium was most often flattened without signs of atypia or mitotic activity. Few capillaries displayed early hyalinization of their walls. Factor VIII as well as CD31 and CD34 reactions were strong in all the endothelial cells (Figure 2B). Alpha smooth muscle actin displayed intense immunoreactivity in some endothelial cells as well as subendothelial pericytes and smooth muscle cells (Figure 2C). The intercapillary space harbored few factor XIIIA positive dendritic cells. Nuclear Ki 67 reaction was present in less than 1% of endothelial cells. Overlying epidermis showed slight keratosis.

Subcutaneous adipose tissue contained scattered nodules with identical morphological appearance, often with an additional component of large and histologically abnormal arterial and venous vessels (Figure 2D). Subcutaneous hemangiomas often incorporated small myelinated nerve branches positive for neurofilaments, membrane basic protein (MBP) and S-100 protein. Some of these nerves showed mild hypertrophic changes and multiple layers of epithelial membrane antigen (EMA) positive perineurium. In addition, the adipose tissue and collagenized dermis contained scattered dilated veins and arteries without accompanying hemangioma. Glial fibrillary acid protein (GFAP) and synaptophysin were entirely negative indicating an absence of heterotopic central nervous tissue in the specimen.

Discussion

The term "dermal sinus tract" was first coined by Walker and Bucy in 1934 to represent a form of congenital occult spinal dysraphism. Dermal sinus tracts are often identified by a skin dimple or tract, associated with a cutaneous manifestation

https://doi.org/10.1017/S0317167100016103 Published online by Cambridge University Press

ranging from hemangiomas to skin tags. The frequency of DST sites are: cervical (1%), thoracic (10%), lumbar (41%), lumbosacral (35%), and sacrococcygeal (13%).[1] Dermal sinus tracts have common histology consisting of a squamous epithelial-lined tract extending from the skin surface to the spinal fascia, dura mater, or spinal cord. In 6% to 7% of cases, the tract terminates dorsal to the spinal elements; in 10% to 20%, it terminates in the extradural space; and in about 60%, it terminates in the intradural space.[1]

The association between hemangiomas and DSTs is well documented throughout the literature. Many studies cite the incidence of hemangiomas to be 10% although the true incidence of infantile hemangiomas remains largely unknown. The likelihood of patients with hemangiomas having DSTs is reported to be about 40%.[2] About half of all DSTs are also accompanied by a mass lesion or tumor, dermoids being the most common.[3] Tethered cord is the functional condition most frequently encountered, occurring in up to 79% of cases.[3] Importantly, DSTs are shown to be well-correlated with meningitis, although there is poor awareness of this association especially at the primary care level. Based on our review of the literature, the association between DSTs and meningitis seems to be discussed significantly less compared to other associations. Radmanesh et al reported that 8 out of 35 children with DSTs presented with meningitis.[4] As a result of such potential concomitant conditions, early surgical intervention is suggested to avoid symptoms like nerve compression, tethered cord syndrome or meningitis.

On MRI, particularly on sagittal scans, DSTs are recognized as a thin hypointense stripe within the subcutaneous fat. They are generally found running obliquely and downward.[5] Tethered cords can be evaluated on MRI by determining the level of the conus as well as recognizing other common findings such as posterior displacement of the cord, lipoma or scar tissue within the epidural space, and increased thickness of the filum terminale.[5] Nevertheless, the intrathecal portion of the tract is often not detectable on MRI, making it difficult to assess the true extent of the tract itself and, particularly, whether it involves the dura and the central nervous system.

The case reported here is unusual in the sense that the child had clear radiological evidence of a tract but intraoperative exploration revealed no intradural extension or tethering of the apparent DST to the conus. Based on the radiological findings, surgery was aimed to untether the spinal cord and to prevent possible future meningitis due to DST. Both of these objectives were addressed through thorough surgical exploration and intervention. Despite the radiological and intraoperative discordance in this patient, we speculate that the intradural extension of the DST seen in the MR images could have represented blood vessels that would not have appeared anomalous intraoperatively. Another possibility is that the DST could have simply been obliterated upon dural opening and thus not visualized during the surgical procedure.

References

1. French BN. Midline fusion defects and defects of formation. In: Youmans JR, editor. Neurol Surg. Philadelphia, PA: WB Saunders Company; 1990. p. 1081-235.
2. Schumacher WE, Drolet BA, Maheshwari M, et al. Spinal dysraphism associated with the cutaneous lumbosacral infantile hemangioma: a neuroradiological review. Pediatr Radiol. 2012; 42(3):315-20.
3. Elton S, Oakes WJ. Dermal sinus tracts of the spine. Neurosurg Focus. 2001;10(1):e4.
4. Radmanesh F, Nejat F, El Khashab M. Dermal sinus tract of the spine. Childs Nerv Syst. 2010;26(3):349-57.
5. Tortori-Donati P, Rossi A, Biancheri R, Cama A. Magnetic resonance imaging of spinal dysraphism. Top Magn Reson Imaging. 2001;12(6):375-409.

https://doi.org/10.1017/S0317167100016103 Published online by Cambridge University Press