Unusual presentation of congenital dermal sinus: tethered spinal cord with intradural epidermoid and dual paramedian cutaneous ostia

Case report

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The authors present the first report of spinal congenital dermal sinus with paramedian dual ostia leading to 2 intradural epidermoid cysts. This 7-year-old girl had a history of recurrent left paramedian lumbosacral subcutaneous abscesses, with no chemical or pyogenic meningitis. Admission MRI studies demonstrated bilateral lumbar dermal sinus tracts and a tethered spinal cord. At surgery to release the tethered spinal cord the authors encountered paramedian dermal sinus tracts with dual ostia, as well as 2 intradural epidermoid cysts that were not readily apparent on MRI studies. Congenital dermal sinus should be considered in the differential diagnosis of lumbar subcutaneous abscesses, even if the neurocutaneous signatures are located off the midline.
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KEY WORDS • tethered spinal cord • epidermoid cyst • neural tube defect • congenital dermal sinus • dual ostia

CONGENITAL dermal sinus tracts of the spine are a rare form of spinal dysraphism, and are hypothesized to be the result of incomplete separation of the neuroectoderm from the cutaneous ectoderm during neurulation.^{1,21,27,43} The incidence of CDS has been estimated to be approximately 1 in every 2500 live births.³¹ The stratified squamous epithelium-lined sinus originates at the skin surface and may terminate superficially within the subcutaneous layers, or it may extend deep through the fascia through normal vertebrae or a bifid spine to communicate directly with the dura mater or intradural compartment.¹² The CDS tracts can be associated with several pathological findings, including inclusion tumors (for example, epidermoid, dermoid, and teratoma),^{2,5,12,21,29,30,35,39} split-cord malformations,³⁴ and tethered spinal cords.⁴⁰ An epidermoid or dermoid cyst may arise at any point along the course of the CDS tract. There have been only 5 reported cases of CDS with dual paramedian ostia, 2 of them associated with dermoid cysts. Association of these multiple paramedian CDS ostia with lumbosacral lipomas and vertebral anomalies such as hemivertebrae, block vertebrae, and accessory hemivertebrae are reported by Lee et al.²⁶

Abbreviations used in this paper: CDS = congenital dermal sinus; DW = diffusion-weighted.

Spinal congenital epidermoid cysts arise from epithelial inclusion during neural tube formation between the 3rd and 5th weeks of embryological development. Acquired intraspinal epidermoid cysts may occur iatrogenically following lumbar puncture.^{9,32} Congenital epidermoid cysts are uncommon, but occur more frequently than iatrogenic epidermoid cysts.²⁸ The incidence of intracranial epidermoids varies from 0.2% to 1%,³⁶ and is even less frequent in the spine. Epidermoids are classically noted to occur intracranially in a paramedian location such as the parasellar region or cerebellopontine angle. They are typically hypo- to isointense on T1-weighted MRI and hyperintense on T2-weighted MRI studies.²⁵ Because their signal intensity on MRI is similar to CSF, epidermoids can sometimes be difficult to diagnose. Some of these tumors appear iso- to hyperintense on T1-weighted MRI due to high lipid content, and hence are called “white epidermoids,” whereas those tumors having reduced lipid content on spectroscopy are called “black epidermoids.”¹⁶ Rarely, these epidermoid tumors have been reported to have varying signal intensities (for example, hyperintensity on T1-weighted images and hypointensity to mixed intensity on T2-weighted images).^{14,16,19,22,33,42}

The DW and FLAIR techniques are commonly used in MRI studies to differentiate epidermoid cysts from

arachnoid cysts. Most epidermoid cysts do not suppress completely on FLAIR images and restrict well on DW images, showing high signal intensity. Arachnoid cysts on the other hand commonly do not restrict on DW images. The CSF component of arachnoid cysts demonstrates a low or suppressed signal on FLAIR imaging. Chen et al.⁸ compared conventional MRI sequences to fast-FLAIR and echo planar DW MRI studies, and concluded that FLAIR was superior to other conventional MRI sequences in detecting epidermoids. However, some arachnoid cysts may also contain blood or proteinaceous material that can further complicate the radiological diagnosis. Likewise, rare white epidermoids are dense lesions with high protein content showing a reversal of signal intensity on MRI studies, with hyperintensity on T1-weighted and hypointensity on T2-weighted images.

Spinal epidermoid cysts may be asymptomatic or may produce a varied range of symptoms such as radiculopathy, motor deficit, or sphincter dysfunction. Gradual worsening of symptoms may suggest enlargement of the cyst with mass effect on abutting neural structures. A sudden onset of symptoms, especially headache, is often suggestive of cyst rupture. The rupture of an epidermoid cyst may introduce its inflammatory contents (keratin, cholesterol crystals, and desquamated squamous cells) into the CSF, resulting in chemical meningitis.

We report a child with 2 paramedian skin dimples and recurrent soft-tissue infections that were drained by general surgeons over 7 years. An MRI sequence of the lumbar spine showed a tethered cord, but 2 intradural epidermoid cysts were noted at surgery. We highlight the importance of CDSs with intradural extension in the differential diagnosis of recurrent lumbosacral soft-tissue infections, even in the absence of chemical or pyogenic meningitis.

Case Report

History and Examination. This 7-year-old girl presented with right lower-extremity radicular pain of inconsistent dermatomal distribution. She had a history of 4 recurrent paraspinal subcutaneous abscesses, all on the left of midline, which were treated at an outside hospital with incision drainage by pediatric surgery. The first episode had occurred at 2 months of age, and the subsequent 3 recurrences since the age of 6 years occurred over an 8-month period. Following the fourth surgical drainage she was treated with 4 weeks of trimethoprim-sulfamethoxazole for *Peptostreptococcus* cultured from the wound. She subsequently began to experience intermittent right lower-extremity radicular pain unaffected by posture or activity. Previously, she had no lower-extremity complaints. She had been born at term following an uncomplicated pregnancy. Her developmental milestones were normal. Examination was remarkable for 2 small lumbosacral dimples, one to the left and the other to the right of midline. Results of the neurological examination were normal.

Admission MRI studies showed a low conus terminating at the L-3 vertebral level. Two subcutaneous tracts

were identified coursing deeply to the thecal sac, one at the S-1 level on the left and the other at the S3–4 levels on the right. No clear evidence of intradural extension was identified (Fig. 1).

Operation. The patient underwent surgery for release of the tethered spinal cord, with neuromonitoring and exploration of the dermal sinus tracts. A midline lumbosacral incision was made between the 2 visible paraspinal dimples and opened sharply through the fascia to gain exposure of the S1–2 junction. An S-1 laminectomy was performed. Dermal sinus tracts were seen coursing through the dura on both sides, and were more prominent on the right than the left. The dura was opened in the midline and the filum terminale was identified under microsurgical magnification. A glistening yellow tumor was seen adjacent to the filum terminale rostrally, and a second yellow tumor was seen adjacent to the lower end of the filum (Fig. 2). Both tumors contained flaky white material on gross examination. There was abundant scarring and fibrosis surrounding each tumor. The scar was divided and the adjacent nerve roots were protected. The rostral tumor was resected (Fig. 2C), and then the caudal tumor was resected and the filum terminale was divided. The dura was closed and the dermal sinus tracts were excised completely.

Histopathological Findings and Postoperative Course. Histopathological examination of the resected tumor specimens demonstrated attenuated squamous epithelium and abundant lamellated keratinous material, confirming that both lesions were epidermoid cysts. Following surgery, the patient's lower-extremity pain was alleviated.

Discussion

Multiple paramedian CDS tracts and ostia are very rare; only a few cases have been reported in the literature.^{1,20,26} Lee et al.²⁶ proposed a hypothesis of "zipping error" to describe the pathogenesis of dual paramedian ostia associated with CDS. They described buckling of the gap between the ascending and descending closure areas of the neural tube resulting in bilateral redundancy in the neural fold, yielding 2 tracts with subsequent propagation as the closure process continues. Our patient had a tethered cord and 2 cutaneous sinus tracts with dual ostia communicating intradurally at different levels, each in a paramedian location, associated with intradural epidermoid cysts.

There are 5 reported cases of congenital dermal sinuses with unilateral and bilateral ostia at the same or a similar spinal level, 2 of them associated with dermoid cysts and lipomas but not epidermoid cysts.²⁶ Triple dermal sinuses of the lumbosacral region, of which 2 were paramedian and 1 midline, have been reported as an exceptional anecdotal case that is suggested to be the result of incomplete disjunction followed by division of 1 sinus into multiple extensions still retaining the attachment to underlying neural structures. The authors of that report, Ansari et al.,⁴ put forth another hypothesis: that the midline dermal sinus resulted from a defective dis-

FIG. 1. **A:** Sagittal T2-weighted MRI study of the lumbosacral spine obtained without contrast, demonstrating termination of the conus at the level of L-3. The rostral epidermoid is barely visible. **B:** Sagittal T1-weighted MRI study of the lumbosacral spine obtained without contrast, demonstrating a left dermal sinus tract with incomplete resection and resultant scar (arrow). **C:** Sagittal T1-weighted MRI study of the lumbosacral spine obtained without contrast, demonstrating a right dermal sinus tract (arrow). **D:** Axial T1-weighted MRI study of the lumbosacral spine at the S2–3 level showing a soft-tissue scar from the prior incomplete resection of the superior dermal sinus on the left side (arrow). **E:** Axial T1-weighted MRI study of the lumbosacral spine at the S3–4 level demonstrating the inferior dermal sinus on the right side (arrow), with intradural communication closer to midline.

junction developmentally, whereas the paramedian ones resulted from a later secondary insult during separation of the ectoderm from the underlying tissue. Double lipomas at different levels have been described even in the presence of split-cord malformations, but intradural lipomas in the setting of bilateral CDS ostia are rare.^{11,24,38} Paramedian congenital dermal sinuses in which there were multiple or laterally located dermal sinuses have also been reported in the literature.^{7,10,18}

The recommended treatment of CDSs with recurrent infections is resection. In the presence of communication with the thecal sac, intradural exploration must be performed. Our patient presented with radiographic evidence of a tethered spinal cord, but was also found to

have 2 intradural epidermoid tumors that were not seen clearly on routine MRI studies of the lumbosacral spine. The epidermoids were not adherent to nerve roots, but were firmly adherent to the filum terminale. Sharp dissection allowed for complete resection of the tumors and subsequent release of the tethered spinal cord. Similar to resection of intracranial epidermoid cysts, the aim of surgery in spinal epidermoids is complete resection and avoidance of aseptic meningitis.

Our patient had previously undergone 4 operations by general surgery at another hospital to drain recurrent left paraspinal abscesses beginning at 2 months of age and extending up to the latest recurrence at 7 years. The MRI sequence of lumbar spine showed that surgical inci-

FIG. 2. Intraoperative photographs. **A:** Two yellowish-white tumors (arrowheads) are identified adherent to the filum terminale. A dermal sinus tract (arrows) that was traced down from the superficial dimpled skin was in communication with the intradural compartment. A free nerve root is seen inferiorly. **B:** The filum terminale is separated from the tumor (arrowhead) to facilitate tumor resection. **C:** After the tumors have been resected, the intact filum is seen prior to its division.

sion and debridement had failed to obliterate the sinus tracts completely (Fig. 1D). Fortunately, the patient did not develop symptomatic pyogenic meningitis, despite evidence of intradural extension of the sinus tracts. In spite of the recurrent subcutaneous infections, our patient had no prior neurological symptoms until shortly after the fourth operation, when she began to experience right lower-extremity pain that prompted referral for neurosurgical evaluation. Recognition of neurocutaneous signatures along the midline of the back, such as dimples, hemangiomas, lipomas, or regions of hypertrichosis should prompt the clinician to consider the possibility of associated spinal dysraphism. Here we report that even when dimpling occurs in a paramedian location, such dysraphism should still be considered.

Sometimes it can be difficult to identify spinal intradural epidermoids because they appear similar to CSF on several MRI sequences. On rare occasions epidermoid contents can be heterogeneous, and can demonstrate a hyperintense signal on T1-weighted MRI studies with peripheral enhancement.⁶ The FLAIR sequences, and in particular, DW imaging, can be helpful in making the diagnosis.^{6,15,23,41} The MRI studies are sensitive in diagnosing the leakage of spinal epidermoid contents intradurally, which results in a chemical or aseptic meningitis. Pyogenic meningitis can occur with dermal sinus tract, previous surgery, or spina bifida aperta, often in a recurrent manner that mandates further imaging studies to rule out CDS tracts.³ Intramedullary abscesses of the spinal cord, albeit rarely associated with epidermoid cysts, have been reported in the presence or absence of dermal sinuses.^{13,17} There are anecdotal case reports of CDS-associated spinal intradural ependymal and neuroenteric cysts in the literature. The reported intradural ependymal cyst associated with CDSs was extramedullary in location and was noted in the cervical region.³⁷ Another lesion associated with CDS tracts is a neuroenteric cyst, which is often an intradural extramedullary lesion and histologically distinct from an ependymal cyst. The MRI modality is extremely helpful in characterization of these spinal lesions and their association with CDS.

Conclusions

To our knowledge this is the first report of bilateral CDS tracts with paramedian cutaneous ostia on either side of the midline and with epidermoid cysts. The patient had been treated for recurrent left paramedian soft-tissue infections, but never had meningitis or intradural abscesses. It is important to recognize that CDSs can be present off the midline because soft-tissue infections can result in intradural complications, including meningitis. The need for proper neurodiagnostic evaluation and neurosurgical exploration is emphasized.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: all authors.

Acquisition of data: all authors. Analysis and interpretation of data: Cohen, Cox, Manjila. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Administrative/technical/material support: Cohen, Cox, Manjila.

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