Case Report

Intramedullary abscess at thoracolumbar region transmitted from infected dermal sinus and dermoid through retained medullary cord

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ABSTRACT

Background: A retained medullary cord (RMC) is a relatively newly defined entity of closed spinal dysraphism that is thought to originate from regression failure of the medullary cord during secondary neurulation. A congenital dermal sinus (CDS) may provide a pathway for intraspinal infections such as repeated meningitis. Intramedullary abscesses are the rarest but most serious complication of a CDS.

Case Description: We treated a female infant with an intramedullary abscess in the thoracolumbar region, which was caused by infection of the CDS. Surgery revealed that the cord-like structure (C-LS) started from the cord with the intramedullary abscess, extended to the dural cul-de-sac, and further continued to the CDS tract and skin dimple. The boundary between the functional cord and the non-functional C-LS was electrophysiologically identified, and the entire length of the C-LS (the RMC) with an infected dermoid cyst was resected. As a result, the abscess cavity was opened and thorough irrigation and drainage of the pus could be performed. Histopathological examination of the C-LS revealed an infected dermoid cyst and abscess cavity with keratin debris in the fibrocollagenous tissue. The abscess cavity had a central canal-like ependymal lined lumen (CC-ELL), with surrounding glial fibrillary acidic protein (GFAP)-immunopositive neuroglial tissues.

Conclusion: We demonstrated that the transmission of an infection through the RMC was involved in the development of the intramedullary abscess. A good postoperative outcome was obtained because a terminal ventriculostomy for pus drainage could be achieved by excising the nonfunctional RMC.

Keywords: Cerebrospinal fluid culture, Chemical meningitis, Ependyma, Primary neurulation, Secondary neurulation

INTRODUCTION

A retained medullary cord (RMC) is a relatively newly defined entity of closed spinal dysraphism, which is thought to originate from an almost complete arrest of apoptosis during the last phase of secondary neurulation.¹⁷,¹⁸ Pang et al.¹⁷ first described seven patients with a redundant
nonfunctional “cord-like structure (C-LS)” originating from the true conus and extending to the dural cul-de-sac, which produced neurological deficits by spinal cord tethering. As nonfunctional RMC is indistinguishable from the functional conus on neuroimaging; moreover, in the intraoperative view, detecting the presence of a nonfunctional C-LS with intraoperative neurophysiological monitoring (IONM) is mandatory for the diagnosis of RMC.[4,6,9,11,12,14,17,18,20] The characteristic histopathological feature of RMC is the predominant presence of a central canal-like ependymal-lined lumen (CC-LELL) with surrounding glial fibrillary acidic protein (GFAP)-immunopositive neuroglial tissues, as a remnant of the medullary cord.[4,6,9,11,12-17,18,21] Some CC-LELLs show cystic dilatation, in which case, these are referred to as “cystic RMC” or “RMC of cystic type,”[4-6,14,18] though Pang et al.[17] did not describe cystic RMCs in their initial article.

A congenital dermal sinus (CDS) consists of a tract lined by stratified squamous epithelium, which is thought to result from a focal premature disjunction between the neuroectoderm and cutaneous ectoderm during primary neurulation.[8,10] Although seemingly innocuous, a CDS may provide a pathway for infections. This can cause discharging sinus, local erythema, and induration as well as repeated meningitis or neurological dysfunction associated with infected inclusion tumors such as dermoid or epidermoid cysts.[1,2,7,10,19] Spinal intramedullary abscesses are the rarest but most serious complication of a CDS, which may lead to permanent and serious neurological deficits.[1,2,7,10,19] Fifty cases of intramedullary abscesses due to a CDS in the pediatric age group were reported in 2019.[19] Recently, we treated a female infant with an intramedullary abscess in the lower thoracic and lumbar region caused by an infected CDS and dermoid cysts. We demonstrated that the transmission of an infection through the cystic RMC was involved in the development of the intramedullary abscesses. Surgical strategies for these complex pathologies have also been discussed.

CASE REPORT

This female infant, who was born at 39+5 weeks of gestation without any problems during pregnancy or delivery, was found to have a dimple at the lumbosacral region at the 2-week medical checkup. At the age of 22 days, she had a fever of 39.9°C and was admitted to the local hospital. She was drowsy and irritable. Her dimples showed no signs of infection and appeared to be blind [Figure 1a]. The numbers of cells and protein in the cerebrospinal fluid (CSF), taken by lumbar puncture, increased to 8533 mm$^3$ (predominantly neutrophils) and 853 mg/dL, respectively. Although the bacterial culture was negative, she was diagnosed with

![Figure 1](image_url)

**Figure 1:** (a) Photograph showing a dimple at the lumbosacral region, continuous from the gluteal cleft, which appeared to end up blind (yellow arrow). (b) Sagittal views of the T2-weighted magnetic resonance image (T2WI), performed on the 22nd day postpartum, show a cord-like structure (C-LS; arrow heads) continuous from the cord and extending to the dural cul-de-sac with spinal cord tethering, a characteristic finding for a retained medullary cord (b-1). A T2-prolonged intramedullary lesion is noted in the lower thoracic and lumbar cord (b-2). (c) T2WI on the 30th day demonstrates exacerbation of the intramedullary lesions. (d) Three-dimensional heavily T2WI image (3D-hT2WI) on the 35th day shows persistence of the intramedullary lesion with hydromyelia-like changes. (e) T1-weighted image with fat suppression (T1WI) fails to reveal the enhancing effect of the hydromyelic lesion following administration of gadolinium contrast medium (Gd). (f) 3D-hT2WI on the 45th day shows reduction of hydromyelic lesions. (g) T1WIs with the administration of Gd reveal the enhancing effect of the hydromyelic lesion (arrows, g-1) and distal part of the C-LS (arrow, g-2).
bacterial meningitis and underwent antibiotic therapy with ampicillin and cefotaxime. Magnetic resonance imaging (MRI) performed on the 23rd day revealed that C-LS continued from the cord and extended to the dural cul-de-sac with spinal cord tethering, which is a characteristic finding for an RMC [Figure 1b-1]. However, continuity between the C-LS and skin lesions was not apparent. In addition, a T2-prolonged lesion was noted in the lower thoracic and lumbar cords [Figure 1b-2]. Chiari malformation or hydrocephalus was not observed.

On the 25th day, her fever disappeared and her general condition improved. The number of CSF cells decreased to 1190 mm$^3$. However, on the 28th day, she suddenly had urinary retention and a urethral catheter was introduced. MRI performed on the 30th day showed an exacerbation of intramedullary lesions [Figure 1c]. With an additional diagnosis of myelitis, intravenous immunoglobulin therapy was administered for 5 days, in addition to antibiotic therapy with vancomycin and cefotaxime. On the 35th day, she experienced spontaneous urination after removal of the urethral catheter. MRI on the same day showed a reduction of hydromyelic lesions [Figure 1f]. However, the enhancing effect of Gd was observed on the hydromyelic lesion in the thoracolumbar cord [Figure 1g-1] and distal end of the C-LS [Figure 1g-2]. She had a transient fever and the CSF examination on the 51st day showed that the cell count and protein again increased to 117 mm$^3$ and 252 mg/dL, respectively. Thus, she was transferred to our hospital on the 52nd day for surgical intervention.

At the age of 55 days, semi-emergent surgery was performed. The dimple did not end at the blind end, and when the keratin-like substance at the bottom was excised, it continued to the deep sinus. The CDS tract began at the dimple and went through the myofascial defect below the level of the bifid S3. Laminoplastic laminotomy of L3-L5 and the dura opening revealed that the swollen C-LS, which partly appeared whitish on the surface, started from the cord, extended to the dural cul-de-sac, and continued to the epidural CDS tract and skin lesion [Figure 2a]. The border between the cord and the C-LS was determined at the L4-5 vertebral level with IONM by tracing the evoked compound muscle action potentials of the legs and anus with stimulation, beginning from the functional cord [Figure 2a, b, (1) and (2)] and proceeding to the nonfunctional C-LS [Figure 2a, b (3), (4), (5), and (6)]. The C-LS was first severed at the dural cul-de-sac and elevated to separate the nerve roots ventral to the C-LS. Second, a rostral incision was made on the dorsal surface of the electrophysiological border between the cord and C-LS, and pus flowed out from the intramedullary abscess cavity (white arrows), the C-LS is resected as a column. The numbers [1], [2], [3], and [4] indicate the position of the section of the resected nonfunctional retained medullary cord in Figure 3.
abscess [Figure 2c]. After irrigation of the pus in the abscess cavity, the C-LS was resected as a column [Figure 2d]. The abscess cavity at the resection edge of the cord was irrigated. Postoperatively, no de novo neurological abnormalities were observed and her fever reduced. Histopathologically, the epidural tract continuous from the dimple had a CDS lined by stratified squamous epithelium and contained keratin debris in the fibrocollagenous tract. The column of the resected C-LS was divided into four axial sections and designated, from the caudal to the rostral sides, as sections [1], [2], [3], and [4], respectively [Figure 2d]. Section [1] consisted of a dermoid cyst, which was lined by squamous epithelium and contained numerous neutrophils, in addition to keratin debris and hair shafts [Figure 3a and b], and GFAP-immunopositive neuroglial tissues with a small CC-LELL in the fibrocollagenous tissue [Figure 3c and d]. In Section [2], both the dermoid cyst and the abscess cavity in the fibrocollagenous tissue [Figure 3e] and GFAP-immunopositive neuroglial tissues were present at the periphery of the C-LS [Figure 3f]. In sections [3] and [4], a large abscess cavity, partially surrounded by GFAP-immunopositive neuroglial tissues with a small CC-LELL, was noted, while there was no squamous epithelium [Figure 3g-k]. Keratin debris was observed in the abscess cavity [Figure 3l]. No bacterial colonies were evident in these sections.

Bacterial culture of the pus confirmed that the causative agents were *Streptococcus anginosus*, *Prevotella bivia*, and *Enterococcus faecalis*, and antibiotics with meropenem and linezolid were administered for 6 weeks postoperatively. MRI performed on the 43rd postoperative day demonstrated that the hydromyelic lesion and intramedullary abscess disappeared and untethering of the cord were achieved [Figure 4a].

![Figure 3: Histopathological findings of the resected cord-like structure (C-LS).](image)

Figure 3: Histopathological findings of the resected cord-like structure (C-LS). The column of the C-LS is divided into four axial sections and designated, from the caudal to the rostral sides, as sections [1], [2], [3], and [4], respectively, as shown in Figure 2d (a). (c), (e) and (f), (g) and (h), and (i) and (j) are the lower magnification views of the sections [1], [2], [3], and [4], respectively, stained with hematoxylin and eosin (a, e, g, and i) and glial fibrillary acidic protein (GFAP)-immunostaining (c, f, h, and j), respectively. (b, d, k, and l) Higher magnification views of the area are indicated by the dashed squares in a, c, and g. (a-d). Section [1] consists of a dermoid cyst (Der), which is lined by squamous epithelium and contains numerous neutrophils (Neu), in addition to keratin debris (Ker) and hair shafts (Hair), and GFAP-immunopositive neuroglial tissues (Glia, red arrows) with a small central canal-like ependymal (Epn)-lined lumen (CC-LELL) in the fibrocollagenous tissue (e, f). Section [2] has both the dermoid cyst and abscess cavity (Abs) in the fibrocollagenous tissue. GFAP-immunopositive neuroglial tissues are present at the periphery of the C-LS (red arrows) (g-l). On sections [3] and [4], a large abscess cavity, partially surrounded by GFAP-immunopositive neuroglial tissues (red arrows) with a small CC-LELL, is noted, while there is no squamous epithelium component. In the abscess cavity, keratin debris is observed (l, blue arrows).
DISCUSSION

In the present case, the C-LS was neuroradiologically, electrophysiologically, and histopathologically consistent with RMC. Detailed histopathological findings of the C-LS showed GFAP-immunopositive neuronal tissues with CC-LELL in all four sections and a dermoid cyst in two sections of the caudal side, indicating that the CDS and the dermoid cyst were continuous from the skin dimple and extended to the caudal RMC. In addition, there were many neutrophils in the caudal dermoid cyst and keratin debris in the rostral abscess cavity. The abscess in the RMC was originally thought to be a cystic dilatation of CC-LELL, because part of the wall was surrounded by GFAP-immunopositive neuronal tissues with CC-LELL. Normally, the wall of the cystic RMC would be lined by ependymal cells, but it is possible that they were lost during abscess formation. These findings indicate that the bacterial infection from the CDS was transmitted first to the intradural dermoid at the caudal side of the RMC, then to the cystic RMC, and finally to the continuous central canal of the thoracolumbar cord, forming an intramedullary abscess [Figure 4b]. Section [2], in which, both dermoid and abscess cavities were found, may represent the transitional part from the infected dermoid and cystic RMC.

It is pathoembryologically unlikely that CDS of primary neurulation failure is associated with RMC, which is considered to be a secondary neurulation failure, at the same site; however, we have reported two similar cases. Since CDS was present on the distal side of the RMC, continuous from the skin lesion in all three cases (including this one), we can consider a mechanism similar to the mechanism by which CDS is dragged down to the stalk of limited dorsal myeloschisis (LDM) that occurs when CDS is associated with LDM of primary neurulation failure. In a study by Kim et al., 11 out of 28 LDMs were located below the S1-2 vertebral level, suggesting an error in the secondary neurulation. As a counterpart of the same spectrum, CDS and dermoid cysts are also found in the area of the secondary neurulation.

Intradural abscesses often leave permanent and severe neurological sequelae and require early surgical intervention. There were fortunately only mild neurological sequelae in this case, but the diagnosis of an intramedullary abscess was slightly delayed. One reason for this is that the response to antibiotic administration for the initial 2 weeks was good. In addition, since we initially decided that the dimple was a blind pouch and the repeated bacterial culture of CSF failed to identify the causative organism, we speculated the possibility of chemical meningitis associated with dermoid rupture. Intraoperatively, the C-LS was swollen further than estimated from the findings of the last preoperative MRI, indicating exacerbation of the infection during the waiting period for surgery. The diffusion-weighted images used for the MRI diagnosis of abscesses and dermoid cysts are often not useful for small intraspinal lesions in infants, which were the case in the present study.

Surgical treatments for intramedullary abscesses associated with CDS and dermoid cysts include complete removal of the...
CDS tract, which is the route of infection and drainage of the abscess.\textsuperscript{[11,12,17,19]} There is no objection to the former method, but the latter method may be invasive when a myelotomy is placed on the functional cord to drain the pus. In the present case, the boundary between the functional cord and the nonfunctional C-LS was electrophysiologically identified, the entire length of the nonfunctional RMC was resected, and the abscess cavity at the severed end of the functional cord was largely opened. As a result, a terminal ventriculostomy for thorough irrigation and effective drainage of the pus in the central canal could be performed [Figure 4c]. In addition, in the case of the coexistence of CDS and RMC, when the CDS is left at the excision margin of the RMC, intradural dermoid cysts may be generated in the future.\textsuperscript{[9-11]} Furthermore, retaining infected dermoid cysts can hinder the cure of the infection. In this respect, IONM is useful and it is important to histologically ensure that no squamous epithelium component remains on the excision margin.\textsuperscript{[9-11]}

**CONCLUSION**

We demonstrated that the transmission of an infection via the RMC was involved in the development of the intramedullary abscess. A good postoperative outcome was obtained because a terminal ventriculostomy for pus drainage could be achieved by excising the non-functional RMC.

**Ethics statement**

The authors confirm that written informed consent was obtained from the family of the infant described in this report. The authors declare that this work complies with the guidelines for human studies, and the research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent.

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**Conflicts of interest**

There are no conflicts of interest.

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