

# Unusual Presentation of Congenital Dermal Sinus: Tethered Cord With Paramedian Cutaneous Ostia

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**Abstract:** Congenital dermal sinus tract is a rare dysraphism with an incidence of 1/2500-3000 ratio. The sinus ostium located in the midline of the posterior of the body from the occiput to the lumbosacral region. Paramedian congenital dermal sinus tract with paramedian ostium are rare. Here, we describe a 2-year-old boy who presented with paramedian pigmented lesion with an ostium on the left lumbosacral region. During surgery we noted that sinus tract attached to thick lipomatous filum terminale.

**Key Words:** dermal sinus tract, intradural, paramedian

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Congenital dermal sinus tract (CDST) is a rare dysraphism with an incidence ratio of 1/2500 to 3000 that mostly occurs in the lumbosacral region.<sup>1,2</sup> The sinus ostium is located in the midline of the posterior of the body from the occiput to the lumbosacral region.<sup>3,4</sup> The most common causes for presentation are skin findings, infection, and space occupying lesions.<sup>3,5</sup> Paramedian CDST is rare. Herein, we describe a paramedian CDST, which ends in the intradural and is attached to lipomatous filum terminale.

## CASE REPORT

A 2-year-old boy presented with paramedian pigmented lesion with an ostium on the left lumbosacral region. The family stated that the pigmented lesion had been visible since birth. His neurological examination was normal. On palpation, there was thickening of the cutaneous tissue (Fig. 1A). Magnetic resonance imaging (MRI) of the spine demonstrated apparent lateral sinus tract originating from paramedian skin surface and extending down to the spinal canal (Fig. 1B) and syringomyelia with low-lying conus medullaris (Fig. 1C).

Excision of the sinus tract was planned. An incision was made around the cutaneous lesion, and the skin was elevated to

allow for dissection of the dermal sinus tract, which continued through the subcutaneous tissue until the level of the duramater. Under an operation microscope, the duramater was opened and we observed that the sinus tract attached to a thick lipomatous filum terminale (Fig. 2). The sinus tract was totally removed, and the lipomatous filum terminale was cut. Postoperative period was uneventful.

On histologic examination, the external dura was found to be surrounded by clusters of dense collagenous tissue that created thick and tight layers. In the inner part, capillaries and connective tissue cells were seen between numerous venous plexus (Figs. 3A, B). In the dural area, fibrous stromal tissue and neuroglial tissue were found to be fused to each other, and plenty of vascular structures with inflammatory cells were observed. Collagen fibers formed to compact collagenous tissue masses in a large region (Figs. 3C, D). Peripheral nerve fibers were found in a large number in the deep dermis with collagen fibers and connective tissue cells in between. Degeneration and vacuolization was observed in axons of the peripheral nerve and, acidophilic degeneration of myelin sheaths were quite evident in the cytoplasm (Figs. 3E, F).

## DISCUSSION

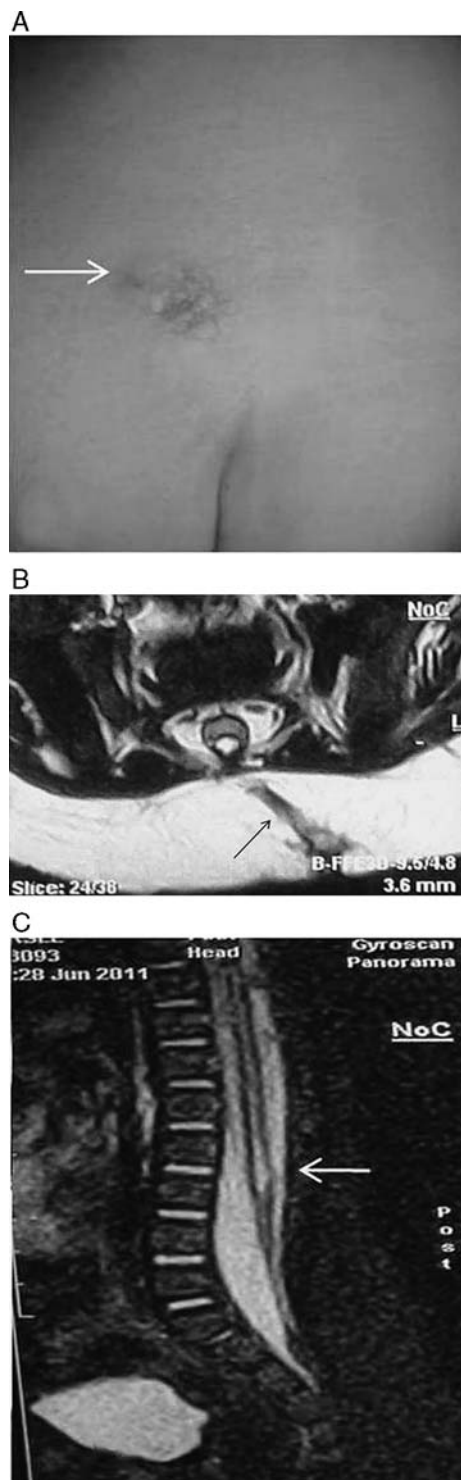
Congenital dermal sinus consists of a tract beginning from the skin and runs to underlying tissue at various lengths and is usually found on the midline anywhere from the nasal bridge to the coccyx. This pathology is thought to be because of neural ectoderm failing to completely separate from the cutaneous surface ectoderm in the third to fifth week of gestation.<sup>4,6</sup> Selçuki et al<sup>1</sup> reported that inhibition of apoptosis seemed to cause a considerable embryological error accounted for congenital dermal sinus tractus maldevelopment. However, why the dermal sinus tract formed distantly from the midline is still unclear. Paramedian CDST with paramedian ostium are rare. Ansari et al<sup>7</sup> reported triple ostium in a patient and 2 of them were bilateral paramedian. Cox et al<sup>2</sup> reported a patient with congenital dermal sinus who had bilateral paramedian ostium. In another article, Lee et al<sup>4</sup> reported 5 congenital dermal sinus patients with multiple ostium. Table 1 shows different case reports of paramedian spinal CDST with paramedian ostium in literature. Laterally located dermal sinuses have also been reported.<sup>3,6</sup> However, our patient had 1 paramedian ostium and we could not find any article in the English literature that reported one or more paramedian ostium other than those shown in Table 1.

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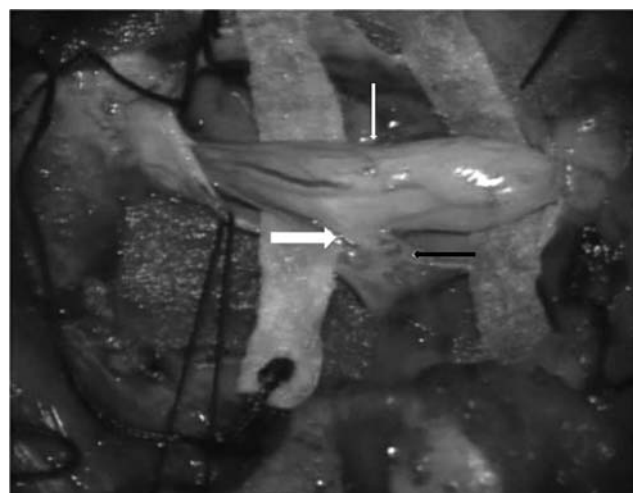


**FIGURE 1.** A, White arrow showing paramedian pigmented lesion and ostium on the left lumbosacral region. B, Black arrow showing apparent lateral sinus tract originating from paramedian skin surface and extending down toward spinal canal on magnetic resonance images. C, White arrow showing syringomyelia with low-lying conus medullaris.

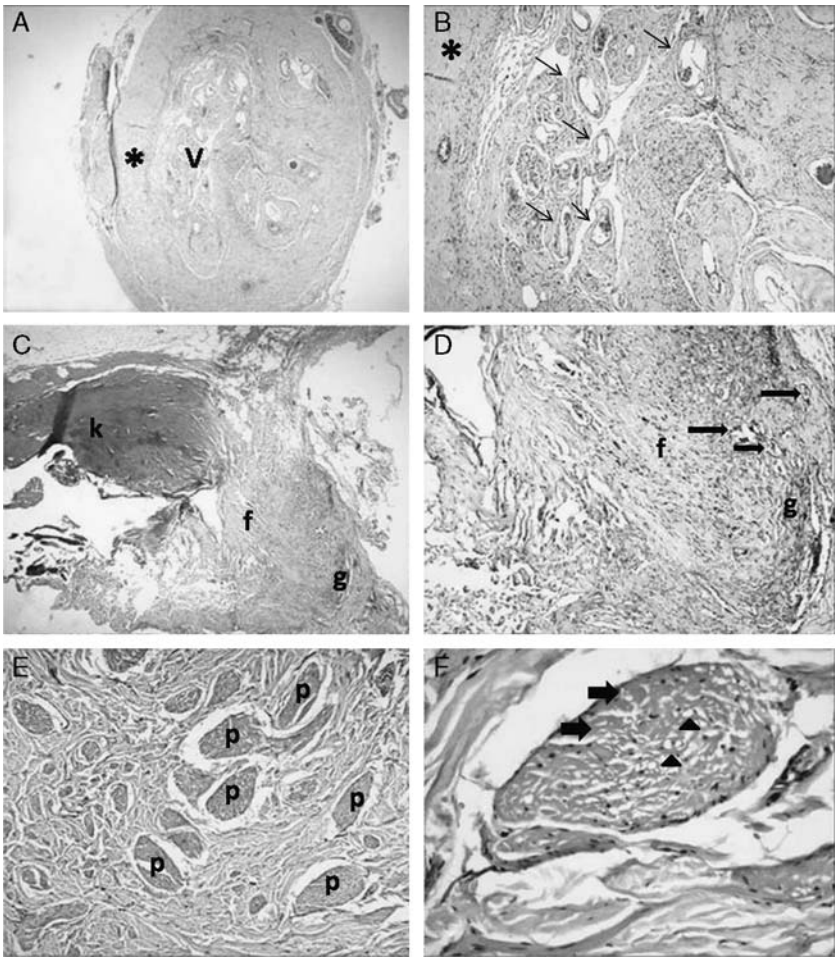
CDST have been associated with clinical manifestations ranging from asymptomatic cutaneous findings to infection, neurological deficit, urinary problems, and orthopedics deformities. Further, CDST may be accompanied with other pathologies such as lipomyelomeningocele, myelomeningocele, split cord malformation, tethered cord, and inclusion tumors.<sup>2,5</sup> In the present case, the surgical findings showed that the tract ascended and directed toward the midline, ending intradural, and attached to lipomatous filum terminale (Fig. 2). He had pigmented cutaneous lesion without clinical findings. In addition to attachment of lipomatous filum terminale to the duramater, dermal sinus tract may cause traction on the spinal cord resulting in a low-lying conus (Fig. 1C).

MRI is the first choice modality to diagnose CDST. It typically appears as a low-intensity tract that ascends in the subcutaneous tissue with high intensity on both T<sub>1</sub>-weighted and T<sub>2</sub>-weighted images.<sup>3</sup> However, only 40% of tracts were detected in the preoperative MRIs, and it is especially useful for determining the level of conus and detecting any other associated anomalies.<sup>5</sup> MRI demonstrated apparent lateral sinus tract originating from the paramedian skin surface and extending down toward the spinal canal and syringomyelia with low-lying conus medullaris (Fig. 1C).

Treatment of all dermal sinuses above the sacrococcygeal region should be explored operatively. The tract should be completely excised after intradural exploration to minimize the risk for neurological deterioration and reexploration.<sup>5</sup> We observed attachment of sinus tract to thick lipomatous filum terminale during surgery and performed a complete resection.



**FIGURE 2.** Thick white arrow shows the attachment point of sinus tract to lipomatous filum terminale. Thin white arrow shows the lipomatous filum terminale and black arrow shows the dermal sinus tract.



**FIGURE 3.** External duramater (A and B), internal region of the duramater (C and D), dermal sinus tract (E and F) (hematoxylin and eosin; A, C, E:  $\times 40$  magnification; B and F:  $\times 400$ ; D,  $\times 100$ ). f indicates fibrous stromal tissue; g, neuroglial tissue; k, compact collagenous tissue; p, peripheral nerve bundles; V, venous plexus; \*, collagen layer;  $\longrightarrow$ , veins;  $\searrow$ , vascular structures;  $\blacksquare$ , acidophilic myelin accumulation;  $\blacktriangle$ , degeneration of axons and vacuolization.

**TABLE 1.** Summary of the Different Case Reports for Paramedian Congenital Dermal Sinuses With Paramedian Ostium in the Literature

References	Age	Sex	Presentation	Internal Ending	Other Skin Stigmata	MRI	Associated Anomaly
Lee et al <sup>4</sup>	8 mo	M	Purulent discharge	Spinal rootlet; dura	Hemangioma	Dermoid tumor	Fatty filum
	15 y	F	Purulent discharge	Paraspinal muscle	None	Dermoid tumor	Currarino triad
	9 y	F	Purulent discharge	Dura	None	Tethered cord	Lumbosacral lipoma
	3 mo	F	Purulent discharge	Intradural lipoma	Subcutaneous mass	Tethered cord	Lumbosacral lipoma
Cox et al <sup>2</sup>	2 y	M	Keratin debris	Intradural lipoma	Subcutaneous mass	Tethered cord	Lumbosacral lipoma
	7 y	F	Subcutaneous abscesses	Intradural	None	Tethered cord	Epidermoid cysts
Ansari et al <sup>7</sup>	10 mo	F	Dimple	Intradural	Creamy dorsal surface	Dermoid tumor	Thick filum
This study	2 y	M	Keratin debris	Intradural lipomatous Filum terminale	None	Tethered cord	Thick fatty filum

**CONCLUSIONS**

- (1) Dermal sinus tract should be kept in mind in the presence of cutaneous lesion outside the midline, whether or not clinical and radiologic examination should be carried out for diagnosis.
- (2) If dermal sinus tract is detected, surgical treatment should be applied as soon as to prevent possible complications secondary to infection, tethered cord, or other pathologies.
- (3) Neurosurgical intervention should include total resection of the tract with intradural exploration.

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