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*Case Report*

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# Rare Association between Congenital Dorsal Dermal Sinus with Tethered Cord Syndrome: A Case Report

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**Abstract**

Congenital dermal sinus tract is an innocent appearing spinal dysraphism that may lead to devastating morbidities if not timely addressed. Congenital dermal sinus may be associated tethered cord syndrome. Here we report one patient who is presented at the age of eighteen months with lower limb weakness at Dr. MR Khan Shishu Hospital and ICH and was diagnosed as dorsal dermal sinus with teathered cord syndrome. Our aim is to illustrate the advantages of the early diagnosis of this rare treatable disorder and to prevent neurological sequelae. [Journal of National Institute of Neurosciences Bangladesh, July 2023;9(2):151-156]

**Keywords:** Congenital dermal sinus; tethered cord syndrome; neuroectoderm; spinal dysraphism; intradural dermoid cyst

## Introduction

Congenital dermal sinus (CDS) 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[1-2]. The incidence of CDS has been estimated to be approximately 1 in every 2500 live births[3]. It includes a tract lined by epithelium, which traverse for a variable depth into the underlying structures and in many instances, terminate within the thecal sac[4-5]. They are seen more frequently at the extremes of neuraxis with the majority of spinal dermal sinus tract occurring in the lumbosacral region[6-8].

Neural tube formation is the essential process of neurulation. Developmental errors during neurulation can lead to the formation of a myelomeningocele, meningocele, intraspinal lipoma, lipomyelomeningocele or dermal sinus tract[9-11]. Dermal sinus tract may have diverse and occasionally serious presentations; in fact, many cases come to clinical attention by neurologic deficit and/or infectious complications including life threatening conditions such as meningitis[6]. In addition, CDS are frequently associated with other anomalies of the central nervous system such as tethered cord, inclusion tumors, and split cord malformations (SCMs)[6]. So despite its benign external appearance, it may harbor great risks to the patients' health if not timely addressed.

Tethered spinal cord syndrome (TCS) is usually identified in childhood and is defined as stretch-induced functional disorder of the spinal cord with its caudal part anchored by an inelastic structure[12-13]. This disorder is usually associated with spinal defects such as myelomeningocele, diastematomyelia, lipomyelomeningocele, thickened filum terminale, intradural lipoma and dorsal dermal sinus tract with an incidence estimated at 0.25 per 1,000 births[14-15]. The cause of TCS is spinal cord traction, which leads to anatomic and metabolic disorders that are responsible for the clinical presentation[16]. Tethered cord syndrome (TCS) is also associated with neurological, gastrointestinal, musculoskeletal, and urinary dysfunction attributable to spinal cord traction[15]. Here we report one patient who is presented at Dr. MR Khan Shishu Hospital and ICH at the age of eighteen months with lower limb weakness and was diagnosed as congenital dorsal dermal sinus with tethered cord syndrome.

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**Case Presentation**

An eighteen-month old male child presented at Dr. MR Khan Shishu Hospital and ICH with sudden onset of lower limb weakness and low grade fever for 2 weeks. He had intermittent watery and purulent discharges from lower back and mother noticed a discharging sinus at lumbo-sacral region since birth. Baby also developed bowel and bladder incontinence after developing weakness of lower limb.

The boy is the 3rd issue of non-consanguineous parents and other siblings are healthy. The boy was delivered full-term by elective caesarian section. The prenatal, perinatal and postnatal periods were uneventful. His motor and cognitive developmental skills were achieved in appropriate age. Mother informed he could sit independently from lying state at 7 months of age, started walking independently at 11-month age. Speech and communication skills achieved at appropriate age.

On examination, the child was very irritable, vital signs were within normal limit. On skin survey there was a discharging sinus on lumbo sacral region & the surrounding area was wet with watery discharges (Figure I). Signs of meningeal irritation were absent.

The patient was conscious, oriented. Cranial nerves were intact. Upper limb examination revealed no abnormality. In examination of lower limbs, muscle bulk and tone were normal. Muscle power was 3/5. Knee jerks were normal but ankle jerks were absent, plantar was nonresponsive. Sensory functions could not be evaluated properly.

Other systemic examination findings revealed no abnormality. We did X-Ray of lumbosacral spine which revealed normal findings. No pre or para-vertebral soft tissue shadow was noted (Figure II).

MRI of whole spine including brain showed teathered cord (S3 level) with congenital dorsal dermal sinus complicated with intraspinal abscess formation within lower part of spinal cord up to L1 level & intramedullary inflammation above L1 upto C7 (Figure III).

We started broad spectrum injectable antibiotics for controlling infections and patient was being prepared for surgery. The patient underwent surgery under general anaesthesia for release of the tethered spinal cord and exploration of the dermal sinus tracts. A one and half inch linear incision was given in lower lumber

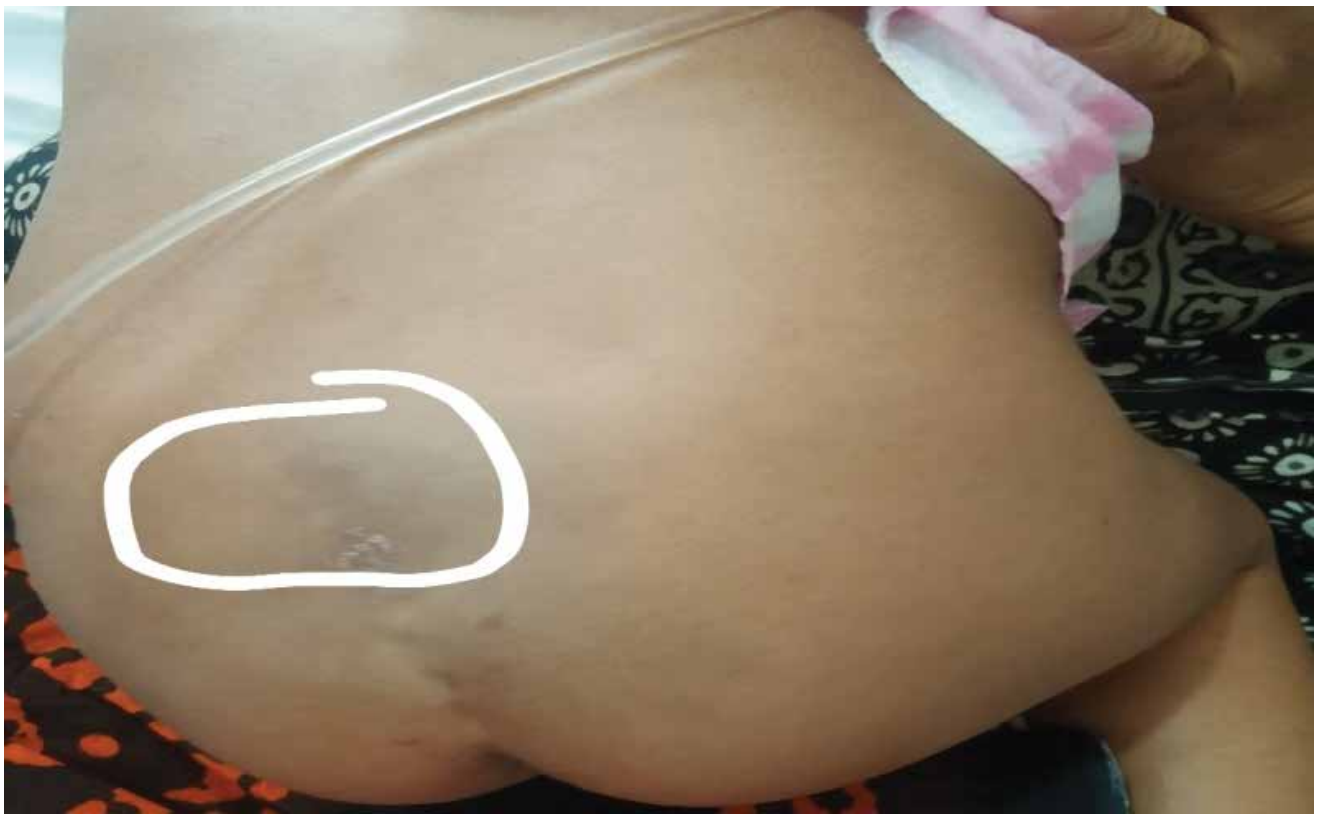

Figure I: Discharging Sinus in Lower Back


Figure II: X-Ray of Lumbo-sacral spine both view

Figure III: MRI of whole spine including Brain

region keeping the opening of dermal sinus in middle. Careful dissection was done up to the opening of sinus in spinal dura (Figure 4). It was tied and dissected out. Wound was then closed in layers. Another one and half inch incision was given at L ¾ level (Figure IV) which was dissected up to lamina on both side. A small incision was made on dura. Pus came out under pressure. Pus was cleaned. A small catheter was passed through the dural opening and intradural space was carefully washed with normal saline till clear fluid came out. Dural opening was closed by suture. Wound was then closed in layer. Pus was sent for culture and sensitivity. His postoperative period was uneventful and was discharged from hospital at a stable condition.

Figure IV: Peroperative photograph of patient

Patient was advised physiotherapy for limb weakness. After 1-month follow-up his muscle power improved. He could walk normally without limping.

**Discussion**

Congenital dermal sinus is an uncommon form of cranial or spinal dysraphism[17-18]. It occurs as a dermal indentation, found along the midline of the neuraxis and act as a conduit for the spread of infection. Occasional secretion might be noticed through this sinus tract[17]. Congenital dermal sinus form due to a focal failure or disjunction between the cutaneous ectoderm and neuroectoderm during the third to eight week of gestation and typically observed in the lumbar and lumbosacral region[17-20].

The stratified squamous epithelium of the congenital dermal sinus tract can extend to the spinal fascia of the dura mater or all the way to the spinal cord[19-20]. Thus, the congenital dermal sinus forms a point of entry for infection, this can allow for the formation of an abscess[18,20]. Infection can then travel up the spinal cord to result in meningitis, which can be fatal if left untreated[17,20]. Our patient had intraspinal abscess formation within lower part of spinal cord, that leads to rapidly progressive lower limb weakness and fever.

Congenital dermal sinus is often also associated with spinal fluid drainage, intradural cysts and spinal cord tethering; conveying neurological deficit[19]. Our patient also had history of recurrent episodes of watery discharge from lumbosacral region from the opening of sinus since birth. Neurological deficit can occur due to spinal cord compression from intradural dermoid cyst growth in the epidermis and dermis[18].

Tethered cord syndrome is the fixation (tethering) effect of inelastic tissue on the caudal spinal cord, limiting its movement. This abnormal attachment is associated with progressive stretching and increased tension of the spinal cord as a child ages, potentially resulting in a variety of neurological and other symptoms[21]. This syndrome is closely associated with spina bifida occulta, which is a common midline defect of vertebral bodies without protrusion of the spinal cord or meninges[22]. It can also be associated with dorsal dermal sinus. There are cutaneous manifestations that may be one of the high indexes of suspicion about the underlying malformation. They are hemangioma, discoloration of skin, pit, lump or hairy patch, that are best investigated with MRI[22]. In our patient, MRI revealed there was tethered cord along with congenital dermal sinus and abscess formation.

Tethered spinal cord can result in gait difficulties and sphincter dysfunction, as well as compressing the spine[20]. Our patient also had bowel and bladder incontinence. Neurological deficits are more likely to occur where diagnosis has not been timely, allowing cysts and or infection[18-19]. MRI allows the neural structures to be observed, visualizing the tract and its anomalies and lesions including inclusion tumors and spinal cord malformations. For diagnosis of TCS and dermal sinus in children, typical imaging features such as visualizing the dermal sinus, a low lying spinal cord and a thickened filum terminale is confirmed by MRI or computed tomography (CT) scan and ultrasound studies. This important feature is proved by the combination of endoscopy and surgical findings. In some cases, electromyography (EMG) and nerve conduction studies may be used to assess nerve function[21].

Treatment of congenital dermal sinus involves complete resection of the tract as well as intradural exploration[19]. Prophylactic surgical removal of the congenital dermal sinus tract is beneficial for the patient, allowing neurological and bladder function to be maintained[17]. Because of its functional (physiological) nature, tethered cord syndrome can be reversible if surgically treated in its early stage. In

children, surgery to release "untether" the spinal cord is recommended[21]. The type of surgery varies depending on the mechanical causes, such as an inelastic filum, myelomeningocele, lipomyelomeningocele, and dermal sinus. Accordingly, the surgical prognosis varies depending upon the presenting symptoms and tethering-producing anomalies. It is clear that in pediatric patients who have firm evidence of tethered cord syndrome, prompt surgical intervention results in reversal, or at least stabilization of symptoms in many cases.

## Conclusion

A dorsal dermal sinus tract with tethered cord syndrome is a rare association. These congenital anomalies can occur individually or together. Proper diagnosis by history taking, clinical examination, with imaging support can be done. Our patient had a discharging sinus at the lumbosacral region since birth, so if it was evaluated earlier properly, this neurological episode could be prevented. Early surgery can prevent cord infarction, paralysis of lower limb, bladder and bowel dysfunction, recurrent meningitis and thus can prevent future neurological sequelae.

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**Conflict of interest**
Other than technical and logistic support from the scientific partner the investigators did not have any conflict of interest in any means

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**Contribution to authors**
Sonia SF, Zaman M, Mahmood E conceived and designed the study, analyzed the data, interpreted the results, and wrote up the draft manuscript. Hassan MS, Banu SH involved in the manuscript review and editing. All authors read and approved the final manuscript.

**Data Availability**
Any inquiries regarding supporting data availability of this study should be directed to the corresponding author and are available from the corresponding author on reasonable request.

**Ethics Approval and Consent to Participate**
Ethical approval for the study was obtained from the Institutional Review Board. As this was a prospective study the written informed consent was obtained from all study participants. All methods were performed in accordance with the relevant guidelines and regulations.

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