

Spinal dorsal dermal sinus tract: An experience of 21 cases

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Abstract

Background: Spinal dorsal dermal sinus is a rare entity, which usually comes to clinical attention by cutaneous abnormalities, neurologic deficit, and/or infection. The present study was undertaken to know the clinical profile of these patients, to study associated anomalies and to assess the results of surgical intervention.

Methods: Medical records of 21 patients treated for spinal dorsal dermal sinus from September 2007 to December 2013 were reviewed.

Results: We had 21 patients with male: female ratio of 13:8. Only 2 patients were below 1-year of age, and most cases (15) were between 2 and 15 years (mean age = 8.2 years). Lumbar region (11 cases) was most frequently involved, followed by thoracic (4 cases), lumbosacral, and cervical region in 3 patients each. All of our patients presented with neurological deficits. Three patients were admitted with acute meningitis with acute onset paraplegia and had intraspinal abscess. The motor, sensory, and autonomic deficits were seen in 14, 6, and 8 patients, respectively. Scoliosis and congenital talipes equinovarus were the common associated anomalies. All patients underwent surgical exploration and repair of dysraphic state and excision of the sinus. Overall, 20 patients improved or neurological status stabilized and only 1 patient deteriorated. Postoperative wound infection was seen in 2 cases.

Conclusions: All patients with spinal dorsal dermal sinuses should be offered aggressive surgical treatment in the form of total excision of sinus tract and correction of spinal malformation, as soon as diagnosed.

Key Words: Complication, dermal sinus, dysraphism, presentation, spine

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INTRODUCTION

Spinal dorsal dermal sinus tract (DST) is a rare congenital dysraphism that occurs in approximately one in every 2500 live births.^[1,7,12,17,18] 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.^[2,3] They are seen more frequently at the extremes

of neuraxis with the majority of spinal DSTs occurring in the lumbosacral region.^[6,7,9,17] Spinal DSTs 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, DSTs are frequently associated with other anomalies of the central nervous system such as tethered cord, inclusion tumors, and

split cord malformations (SCMs).^[2,6] So despite its benign external appearance, it may harbor great risks to the patients' health if not timely addressed. The neurological examination is reported to be normal in the early childhood. However, as the age increases, there is more chance of neurological deficit, which tends to be more profound. There are few published series in literature which emphasize mainly the mode of presentation, radiological findings, associated anomalies and treatment; however, the symptom wise outcome is not studied in detail.^[1,3,6,12,17,18] The present study was undertaken to know the clinical profile, associated anomalies and detailed symptom wise outcome of the patients presenting with spinal DST.

PATIENTS AND METHODS

This is a retrospective study conducted in Pt. B.D. Sharma University of Health Sciences, Rohtak from September 2007 to December 2013. Medical records of all patients treated for spinal DST were reviewed. Information regarding patients' demographic variables, type of presentation, symptoms, physical examination, radiological and surgical findings, and histopathological evaluation were collected. Magnetic resonance imaging (MRI) was the investigation of choice and was performed in all cases. MRI revealed the relationship of the dermal sinus to the dural sac and also gave information regarding associated abnormalities in the cord like dysraphic state of spine or inclusion tumor.

Surgical intervention

The aim of surgery was to excise the sinus tract completely and to correct the dysraphic state in the same sitting. Surgery was performed in all cases through midline incision with encircling the sinus. DST was followed through the subcutaneous tissue and muscle layer sinus tract was traced until its end and excised completely. The course of DST was invariably rostral through the incompletely formed lamina or underneath the normal lamina. After doing the laminectomy, dura was opened in all cases irrespective of end of DST. In cases where DST was intradural, part of dura encircling the DST was excised. Intraspinous pathologies like SCM were dealt accordingly that is, dermoid and epidermoid were decompressed or excised; myelocoele and lipomeningomyelocoele were repaired; drainage of abscess in intramedullary abscess, removal of arachnoid adhesion in arachnoiditis and detethering of the cord was done in case of tethered cord. Those patients presenting with infectious complications were managed with appropriate antibiotics and then after recovery surgery for resection of DST, and correction of associated anomalies was performed. Postoperative follow-up ranged from 6 months to 5 years (mean - 2.8 years).

RESULTS

Records of total 21 patients were analyzed, of which 13 were male, and 8 were female. Patients' age on admission ranged from 9 months to 15 years (mean - 8.2 years). Every patient underwent a detailed neurological examination and a complete radiological workup to delineate any underlying/associated spinal abnormalities. DST was located most frequently in lumbar region (11 cases) [Figure 1], followed by thoracic (4 cases), [Figures 2 and 3] cervical [Figure 4] and lumbosacral region in 3 patients each [Table 1]. It was astonishing to note that all our patients presented with neurological deficits [Table 2]. Three patients presented with acute meningitis and acute onset paraplegia. History of recurrent meningitis was also positive in two of these cases. Gradually progressing motor deficit was seen in 14 cases. The deficit was in the form of limb weakness and atrophy, with or without gait disturbance. The sensory deficit was seen in 6 cases. Eight patients had bladder/bowel involvement at presentation out of which five were incontinent at the time of presentation. Associated skeletal anomalies were noticed in 5 cases. Scoliosis was the most common finding and was seen in 4 cases, followed by congenital talipes equinovarus in 2 cases. In the majority of the patients (15), sinus ostium was associated with another skin abnormality, the most common of which was abnormal pigmentation. Some patients had a combination of these findings. Dermal sinuses were seen in conjunction with lipomyelomeningocele in 2 patients. MRI was the investigation of choice and was performed in all cases. It revealed the relationship of the dermal sinus to the dural sac and also gave information regarding associated abnormalities in the cord [Table 3]. Epidermoid [Figure 2b] and dermoid tumor [Figure 3b] were seen in 2 and 8 cases respectively. SCM was seen in 6 cases and filum abnormality in 2 cases [Table 4]. Motor deficit (present in 14 cases) stabilized in 7 cases and improved in 6 cases [Table 5]. Two patients developed fresh deficits in the postoperative period, and one of them improved to preoperative status 3 months later. The sensory improvement was seen in 2 cases and sensory deficits stabilized in 4 cases. Neurologic function gradually returned to near normal state postoperatively in 2 of 3 patients who presented with acute paraplegia with incontinence of urine and stool, but there was no improvement in bladder and bowel function in all 3 patients. Of the other 5 incontinent patients, 1 improved, and 4 remained the same.

Overall, 11 patients showed neurological improvement, 9 patients stabilized neurologically while 1 patient deteriorated [Table 6]. Improvement in any of the neurological parameters viz-motor, sensory or bowel/bladder symptoms was considered to be an improvement.

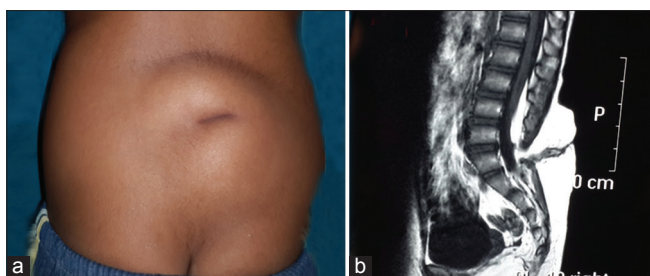


Figure 1: (a) Spinal dermal sinus tract of lumbar region with lipomyelomeningocele. (b) Magnetic resonance imaging (T1-weighted image) showing the tethered cord due to stalk extending from the sinus to the intradural space along with lipomyelomeningocele

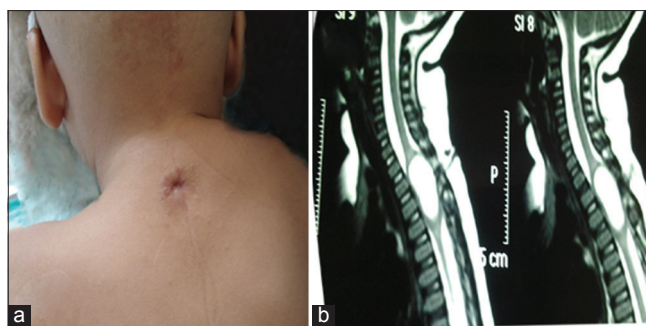


Figure 2: (a) Spinal dermal sinus tract of thoracic region. (b) Magnetic resonance imaging (T2-weighted image) showing intradural epidermoid tumor attached to the thin stalk extending from the sinus

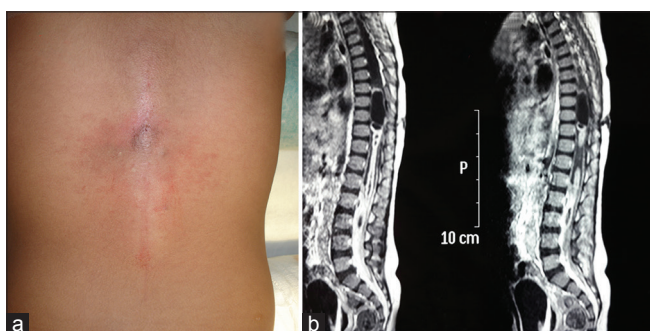


Figure 3: (a) Spinal dermal sinus tract of thoracic region. (b) Magnetic resonance imaging (gadolinium enhanced T1-weighted image) showing intradural dermoid tumor with intramedullary abscess

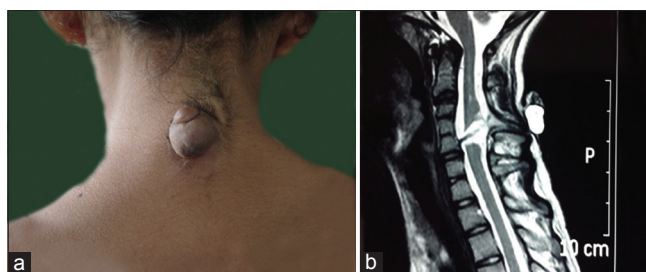


Figure 4: (a) Spinal dermal sinus tract of cervical region with rudimentary meningocele. (b) Magnetic resonance imaging (T2-weighted image) showing the tethered cord due to stalk extending from the sinus to the intradural space and attach to the bony spur

Table 1: Distribution of spinal DSTs (n=21)

Spinal level	Frequency (%)
Cervical	3 (14.2)
Thoracic	4 (19)
Lumbar	11 (52.3)
Lumbosacral	3 (14.2)

DST: Dermal sinus tracts

Table 2: Neurological signs and symptoms in patients with dermal sinus (n=21)

Sign and symptoms	Frequency (%)
Acute meningitis with acute paraplegia	3 (14.2)
Motor weakness	14 (66.6)
Sensory weakness	6 (28.5)
Club foot/scoliosis	5 (23.8)
Incontinence of urine/stool	5 (23.8)
Constipation	3 (14.2)

DISCUSSION

A spinal DST consists of a tract lined by stratified squamous epithelium found on or near the midline and is thought to result from the abnormal adhesions (or incomplete disjunction) between the neuroectoderm (destined to form the neural tube) and

the cutaneous ectoderm.^[4,10,11,15] The inward extent of the tract depends upon the extent of adhesions and may vary from deep fascia to the spinal cord. The tract elongates during the development, due to ascent of the cord and may traverse several levels within the epidural space before entering the subarachnoid space. Disorder of the notochord formation with sagittal splitting of the spinal cord and persistence of the dorsal cutaneo-endo-mesenchymal fistula has also been suggested as a cause of dermal sinus formation.^[11,16] The squamous lining of spinal DST may be encased in dermal and neurological tissue. Within the tract, one may find nerve or ganglion cells or fat, blood vessels, cartilage and meningeal remnants.^[14] Spinal DST may be associated with other abnormalities of the ectodermal, mesodermal or neural crest derivatives such as meningomyelocele or lipomeningomyelocele, reflecting a common ontogenic disorder. Nearly, 60% of the DSTs enter the subarachnoid space and 27% are attached to the neural elements of the conus, cauda equina or filum terminale.^[1,5,17,19] The tract may end blindly within the extradural space in 10–20% cases.^[6,17] Sinus tracts can occur anywhere from occiput to sacrum. Different studies showed that cervical area is least involved (<1% cases). Thoracic area is involved in 10% cases, lumbar and lumbosacral area in 40% and 12% patients respectively, sacrum in 23% and sacrococcygeal junction in 13% of cases.^[6] In the present study, the cervical region was involved in 14.2% cases,

the lumbar region in 52.3% cases, thoracic region in 19% cases and lumbosacral region in 14.2% cases. The higher incidence of DST in cervical area in the present study could be due to selection bias as cervical DST cases are always symptomatic while other authors might include asymptomatic lumbosacral cases.

Dermal sinuses should be distinguished from the more common coccygeal pits. Dermal sinuses are located above the intergluteal cleft and have a cephalically oriented course and are often associated with other pathologies. On the contrary, coccygeal dimples are usually simple blind sinuses with no associated cutaneous abnormalities that lie within intergluteal cleft a few millimeters cranial

to the tip of coccyx. They are oriented caudally or straight and are not associated with other intradural pathologies and thus do not warrant further evaluation.^[2,3,15] They may rarely have intraspinal extension, so it should be remembered that not all coccygeal pits can be dismissed. Another characteristic that differentiate coccygeal dimple from DST is location. Coccygeal pits are always in midline while DST is not strictly midline and should be investigated with high-quality MRI. If a sacral or coccygeal dimple is associated with other cutaneous abnormalities such as hypertrichosis or soft tissue mass, they should be investigated accordingly.

Dermal sinuses provide a portal of entry for bacterial agents into the intraspinal compartments that can cause meningitis or abscess formation that may be extradural, subdural, and intramedullary or infection of associated tumor. Also, aseptic meningitis can occur by spillage of inclusion tumor contents or other dermal elements into the cerebrospinal fluid.^[7,9,19] Therefore, one should have a high level of suspicion for DST and dermoids when encountering any young child presenting with aseptic meningitis. In the study conducted by Jindal and Mahapatra^[6] only 1 patient presented with infection out of 26 patients. Ackerman and Menezes^[2] also had a low rate (10%) of infectious complications. In the series of Radmanesh *et al.*,^[17] 37.1% had meningitis on admission or had experienced it before while 25.7% had abscess formation. The incidence of infection (meningitis) in our patients was 14.2%, all our infected patients had abscess of which two were intramedullary.

It has been said that nearly all children with spinal DSTs have intact neurological function at birth.^[6] However, due to the relatively high rates of associated pathologies such as tethered cord, infection, and inclusion tumors, neurological deterioration becomes more common with increasing age. It has been shown that the chances of developing neurologic deficit are higher in patients who present in older ages.^[2] Ackerman and Menezes^[2] studied the referral pattern among their patients and noted that patients who were younger than 1-year were more likely to be neurologically intact than older ones, concluding that delay in the diagnosis allows for development of neurologic sequelae. Probably this may be the reason that all of our patients presented with neurological deficits as 90% of our patients were more than 1-year of age which may due to lack of awareness at the primary health care level, which leading to delayed referral. Unfortunately,

Table 3: MRI finding in patients with dermal sinus (n=21)

MRI finding	Frequency (%)
Epidermoid	2 (9.5)
Dermoid	8 (38)
Thick filum	5 (23.8)
SCM	5 (23.8)
Lipomeningocele	2 (9.5)
Syrinx	6 (28.5)
Intraspinal abscess	3 (14.2)

MRI: Magnetic resonance imaging, SCM: Split cord malformation

Table 4: Intraoperative finding in patients with dermal sinus (n=21)

Intraoperative finding	Frequency (%)
Tethered cord	13 (61.9)
Epidermoid	2 (9.5)
Dermoid	8 (38)
Intramedullary abscess	2 (9.5)
Intradural granulation tissue	1 (4.76)
SCM	5 (23.8)
Arachnoiditis	4 (19)

SCM: Split cord malformation

Table 5: Symptom wise surgical outcomes in patients with dermal sinus (n=21)

Function	Number	Improved (%)	Same/stabilized (%)	Worsened (%)
Bladder and bowel	8	1 (12.5)	7 (87.5)	-
Sensory	6	4 (66.6)	2 (33.4)	-
Motor	14	6 (42.8)	7 (50)	1 (7.1)
Total	28	11 (39.2)	16 (57.1)	3.5

Table 6: Comparison of surgical outcome with different previous series

	Ackerman (%)	Jindal (%)	Ramnarayan (%)	Metz (%)	Presentseries (%)
Neurologically asymptomatic	11 (39)	3 (13)	1 (11)	12 (75)	-
Neurologically improved	12 (43)	8 (34.7)	4 (44.4)	-	11 (52.3)
Neurologically stabilized	2 (7)	14 (60.8)	4 (44.4)	4 (25)	9 (42.8)
Neurologically worsened	3 (11)	1 (4.3)	-	-	1 (5)

once a patient develops neurologic deficit, there is a relatively high chance of permanent defect.^[5,6]

Spinal dermal sinuses may be accompanied by other forms of spinal dysraphism such as lipomyelomeningocele and myelomeningocele, reflecting a possible common ontogenic pathway.^[17] Gupta *et al.* showed an association of 11.34% between dermal sinus and other forms of spinal dysraphism.^[5] The proposed mechanism for lipomyelomeningocele embryogenesis also includes disorders of disjunction that occurs prematurely in this entity. It is possible that there are some shared molecular pathways responsible for concurrence of these anomalies. Dermal sinuses are occasionally associated with tethered cord, although only 1% of patients with tethered cord have dorsal dermal sinus.^[2] In patients with DST, the tract or associated tumor may cause traction on spinal cord resulting in a low-lying conus and tethered cord syndrome.^[8] In our study, 13 patients (61.9%) had tethered cord. It is reported that up to 40% of patients with DST can have SCM.^[3,17] Conversely, DSTs are seen in 15–40% of SCM.^[19] Among our patients, five had SCM, three with Type 1, and two with Type 2. The incidence of filum terminale abnormalities was described by Jindal and Mahapatra^[6] and Radmanesh *et al.*^[17] Jindal and Mahapatra^[6] found filum abnormalities in 22% of his patients while Radmanesh *et al.*^[17] found filum terminale abnormality in 40% cases. In the present study, the filum terminale abnormalities were encountered in 5 cases (22%). The term tight filum terminale refers to a set of conditions in which a low-lying conus medullaris is associated with a short thickened filum without evidence of other tethering pathologies.^[7,8] This entity that arises from failed regression of caudal spinal cord during secondary neurulation causes typical signs and symptoms of tethered cord.

Approximately, half of all dermal sinuses are associated with dermoid or epidermoid tumor, usually at the termination of these tracts, but they may be located anywhere between the skin and the neural tube.^[2,6,13,17] Dermal sinuses and dermoid tumors seem to share a common origin.^[9] They are believed to result from focal expansion of these ectoderm-derived tracts. However, only approximately 30% of intraspinal dermoid tumors have an associated sinus tract.^[7] DSTs are associated more frequently with dermoid tumors (83%) than with epidermoid (13%).^[5] In the present study, two of our patients (9.5%) had epidermoid tumors while eight had dermoids (38%) proved by histology.

Postoperative complications were few and easy to manage. Our results indicate that once a patient developed bowel/bladder incontinence, there was about 12.5% chance of improvement in deficit while in patients with sensory or motor deficits; the chance of improvement was 66.6% and 42.8%, respectively [Table 5]. The risk of neurological

deterioration was only 3.5%. The patients presenting at later age had more chance of developing deficits. We have also compared the neurological outcomes in the different previous series with our study [Table 6]. In our study, the overall neurological improvement is better than the previous study that may be due to selection bias as all our patients were symptomatic. However, bowel/bladder improvement was seen in only 1 patient due to the delayed presentation.

Since none of imaging modalities can accurately show intraspinal details, all dermal sinuses above the sacrococcygeal region should be explored operatively regardless of neuroimaging findings.^[3,7,17] One should have a high index of suspicion for all the dimples above the intergluteal fold, despite a normal examination or neuroradiologic studies. Midline should be carefully examined whenever a child suffers from meningitis, especially when an unusual organism is cultured. Conservative treatment of spinal DST is not recommended. Surgery should be carried out prophylactically in advance of deficits, to maintain normal neurological function.

CONCLUSION

Spinal DST is an innocuous-appearing spinal dysraphism that may contribute to devastating morbidities if not timely addressed. Although there has been increased awareness about the impotence of dorsal midline cutaneous finding among primary health care physician, there still much more to be done especially in developing country. All patients with spinal DST should be offered aggressive surgical treatment in the form of total excision of sinus tract and correction of spinal malformation, as soon as diagnosed since chances of preserving and/or improving neural function are high (95%).

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