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ORIGINAL ARTICLE

Spectrum of Spinal Dermal Sinus: Analysis and Outcome Evaluation

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ABSTRACT

Objective: Congenital spinal dermal sinus is a rare form of spinal dysraphism. It often presents in childhood with varied symptomatology. We analyzed our patients to study the symptomatology, operative findings, and patient outcomes.

Materials and methods: We evaluated 10 patients with congenital dermal sinus (CDS) over spine who attended our outpatient clinic during the period 2006 to 2008.

Results: The male: female ratio was 1:1 in our series. Two patients were of below 2 years of age. We noticed 1 cervical, 3 dorsal, 2 lumbar, and 4 lumbosacral sinuses. Reasons for attending the clinic were neurological deficits in 4 patients and cutaneous findings in 6 patients. Pain, restricted neck movements, and infection were found in one patient each. Patients in pediatric age group (57%) were more likely to have neurological deficit than adults (33%). Bifid spinous processes were noticed in all except one patient. Tethered cord was a common finding in all patients. Two intramedullary dermoid with abscess formation in one of them was found. Terminal lipoma, terminal syrinx, and neurenteric cyst were found in each one. Hydrocephalus, split cord malformation, and epidermoids were not encountered by us. Postoperatively, six remained neurologically intact, three were unchanged, and one partially improved. Neurological worsening was not noticed.

Conclusion: Congenital spinal dermal sinus should be dealt surgically. Intradural exploration is the most important part of surgical excision. Early referral will prevent permanent sequel. Outcome is directly related to preoperative neurological status.

Keywords: Dermal sinus tract, Intramedullary dermoid, Spinal dysraphism.

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INTRODUCTION

Spinal dermal sinuses are an uncommon form of dysraphism. The incidence of CDS of spine is usually reported as 1 in 2,500 live births.[1] It does not represent the true incidence because in most of the series, coccygeal pits are included. More so, a large number of patients go unrecognized in our country. To establish the true incidence, a detailed study is essential. Most of CDS of spine is associated with midline cutaneous stigmata, which is obviously visible than a sinus opening. The presence of cutaneous stigmata over midline neural axis should prompt early referral by a primary care provider.[2-4]

MATERIALS AND METHODS

We retrospectively analyzed 10 patients with CDS of spine who attended our department during the period from 2006 to 2008. Thorough clinical examination was done on all to assess the neurological status and deficits. All of them were evaluated with X-rays and magnetic resonance imaging (MRI) of spine. Brain screening was done as and when necessary. All patients were treated surgically and intradural exploration was carried out in all.

RESULTS

We found that three were adults and seven were children. The male: female ratio was 1:1. Age ranged from 1½ to 27 years. The average age was 9.4 years. Two patients were below 2 years at presentation. Six patients attended the outpatient department (OPD) for their cutaneous stigmata only and during examination, one revealed pain in the sinus and the other one revealed restricted neck movement. Remaining four patients came to the OPD for their neurological deficits, one among them presented with flaccid paraplegia in an emergent situation. Bladder and bowel impairment was a common finding in those who presented with neurological deficits (Table 1 and Figs 1 to 3).

Regarding the location of sinus, we noticed CDS one in cervical, 3 in dorsal, 2 in lumbar, and 4 in lumbosacral level. Intraoperatively, we noticed the tethering of cord in all patients with sinus tract. Spina bifida was noticed in all patients except one in whom the tract was seen going through interlaminar space. Bifid spine corresponded to the dermal sinus tract (DST) into spinal cord. Adjacent-level bifid spines were also noticed in two patients.

Table 1: Summary of patients

Age/sex	*Location*	*Cutaneous finding*	*Termination*	*Clinical presentation*	*Associated anomaly*	*Operative findings*	*Possible embryology*	*Outcome*
Case 1 6 years/F	Cervical spine C5	Tuft of hair around sinus	Tract ended on the cord at the level C4	Restricted neck movement	Neurenteric cyst anterior to cord extending from C2–C3 to upper border of D2. Spina bifida of C4 and 5	Cystic lesion anterior to the cord was tapped and the content found to be turbid fluid. Cyst wall biopsied and excised	At the end of the third embryonic week, the development of the notochord is intimately related to endodermal cells. If notochord fails to detach itself from the endodermal layer, endodermal cells can be dragged forwards and upwards. This may lead to the formation of a cyst in front of the spinal cord	Neck movements improved
Case 2 7 years/M	Lumbar L5	Dermal sinus and hypertrichosis	Tethered to conus at L1 level	Bladder and bowel incontinence	Tethered cord and Terminal syrinx were present. Spina bifida of all lumbar vertebrae. Scoliosis at lumbar level	Dermal sinus extended to conus	Failure of disjunction of surface ectoderm from the neuroectoderm	Neurological status same as preoperative
Case 3 1½ years/M	Dorsal D10	Dermal sinus with surrounding inflammation	D8-10	Flaccid paraplegia with bladder and bowel involvement	Intramedullary dermoid with abscess formation extending three levels above. Associated spina bifida was also present	Sinus tract ending on intramedullary dermoid	Between the 3rd and 5th weeks of fetal development, cells fated for cutaneous ectoderm somehow become trapped within neural ectoderm during neural tube closure and eventually form tumors	No improvement postoperatively
Case 4 19 yrs/M	Lumbar spine L3	Tuft of hair around the sinus	Filum	Pain in the sinus area	Tethering of cord and low ending of spinal cord. Spina bifida	Simple tethering of cord. Dura and thickened filum terminale	Failure of disjunction of surface ectoderm from the neuroectoderm. Spina bifida is part of neural tube defect	Neurological status same as preoperative
Case 5 27 years/F	Dorsal spine D4	Dermal sinus	Ending on cord D3	Spastic paraparesis with bladder involvement	Intramedullary epidermoid with lipomatous elements extending to low cervical level C7. Spina bifida at D4. Scoliosis of dorsal level	Sinus tract ended on the spinal cord and it was associated with intramedullary dermoid	Between the 3rd and 5th weeks of fetal development, cells fated for cutaneous ectoderm somehow become trapped within neural ectoderm during neural tube closure and eventually form tumors	Spastic paraparesis and bladder involvement. Improved postoperatively
Case 6 5 years/M	Lumbosacral	Dermal sinus	Tethered to filum	Bladder and bowel incontinence Motor weakness	Tethered cord and terminal lipoma	Dermal sinus ended in the terminal part of blind end of dural sac with tethering of cord	Lipoma of the filum terminale is probably due to persistence of caudal cells that differentiate to fat cells. Tight filum is due to deranged canalization& retrogressive differentiation	Cerebrospinal fluid leak and wound gapping necessitated secondary suturing. Neurological status same as preoperative

(Cont'd…)

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Age/sex	Location	Cutaneous finding	Termination	Clinical presentation	Associated anomaly	Operative findings	Possible embryology	Outcome
Case 7 22 years/M	Dorsal D3 level	Dermal sinus	D3 Cord	No neurological deficit	Cord is tethered to the dermal sinus at D3 level. Spina bifida at D3	Dermal sinus extended into the dura and attached to the underlying spinal cord with dorsally tethered cord to the sinus	Failure of disjunction of surface ectoderm from the neuroectoderm	Neurological status same as preoperative
Case 8 1½ years/F	Lumbosacral	Lumbosacral dermal sinus	Filum	No neurological deficit	Tethered cord	Dermal sinus extended to filum and associated with tethered cord	Failure of disjunction of surface ectoderm from the neuroectoderm	Neurological status same as preoperative
Case 9 3 years/F	Lumbosacral	Dermal sinus at lumbosacral L5-S1 region with hypertrichosis	Conus	No neurological deficit	Tethered cord	Dermal sinus extended to conus associated with tethered cord	Failure of disjunction of surface ectoderm from the neuroectoderm	Neurological status same as preoperative
Case 10 1½ years/M	Lumbosacral	Lumbosacral dermal sinus	Filum	No neurological deficit	Tethered filum terminale	Sinus tract ending on filum terminale	Failure of disjunction of surface ectoderm from the neuroectoderm	Neurological status same as preoperative

Scoliosis was noticed in two patients. Intramedullary dermoid with abscess formation was seen in dorsal-level dermal sinus who presented with acute onset of flaccid paraplegia. Surgery was done as emergency in this patient; all others were operated on elective situation. Intramedullary dermoid, terminal syrinx, and terminal lipoma were encountered in one patient each. One patient with cervical-level sinus had neurenteric cyst with sinus tract ending on cord, but no communication was demonstrated between the sinus tract and the neurenteric cyst. Split cord malformations, epidermoids, and hydrocephalus were not encountered in any of our patients.

One patient developed cerebrospinal fluid (CSF) leak, which was successfully managed conservatively. No patient deteriorated during follow-up. In those who presented with neurological deficits, one showed partial improvement and the remaining three maintained their preoperative neurological status.

ILLUSTRATIVE CASES

Case 1

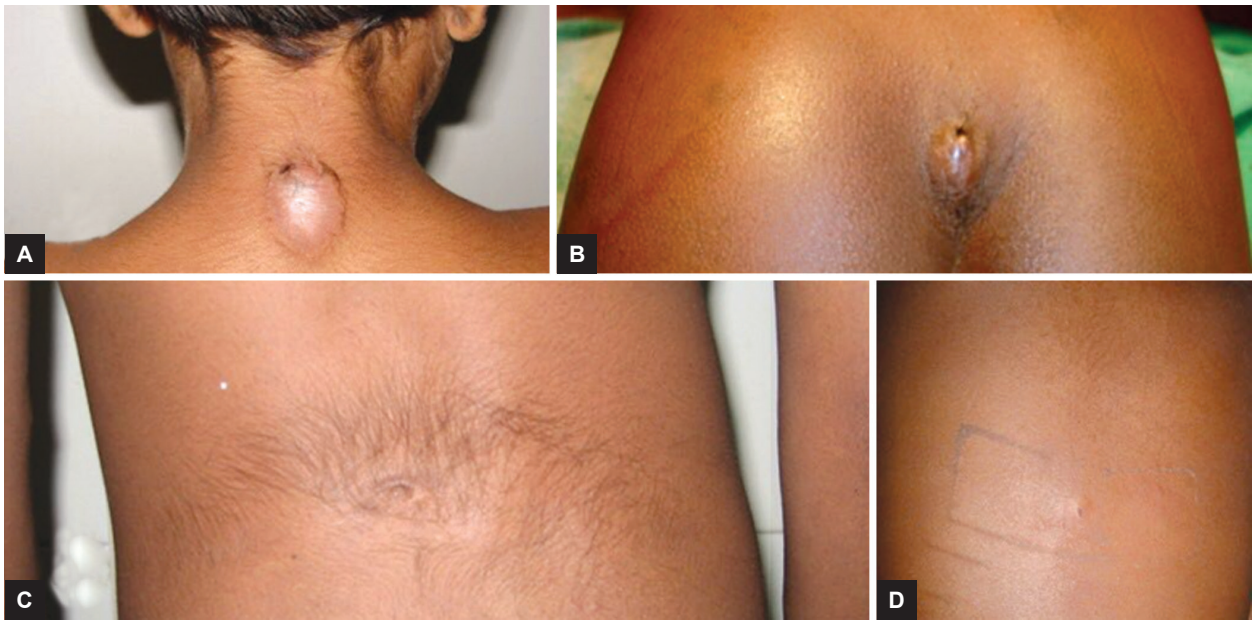
A 6-year-old female patient presented with restricted neck movements for a few months. Examination of the patient revealed a sinus over the C5 region and an area of altered pigmentation around the sinus. During evaluation, MRI showed the sinus tract ending on the cord at the C4 level with a cystic lesion anterior to the cord extending from C2 to D2 and spina bifida of C4 and C5. The tract was found to end on the cord and it was excised totally. The cyst was tapped which contained a turbid fluid. The cyst wall was biopsied and excised. Postoperatively, neck movements improved (Fig. 4).

Case 2

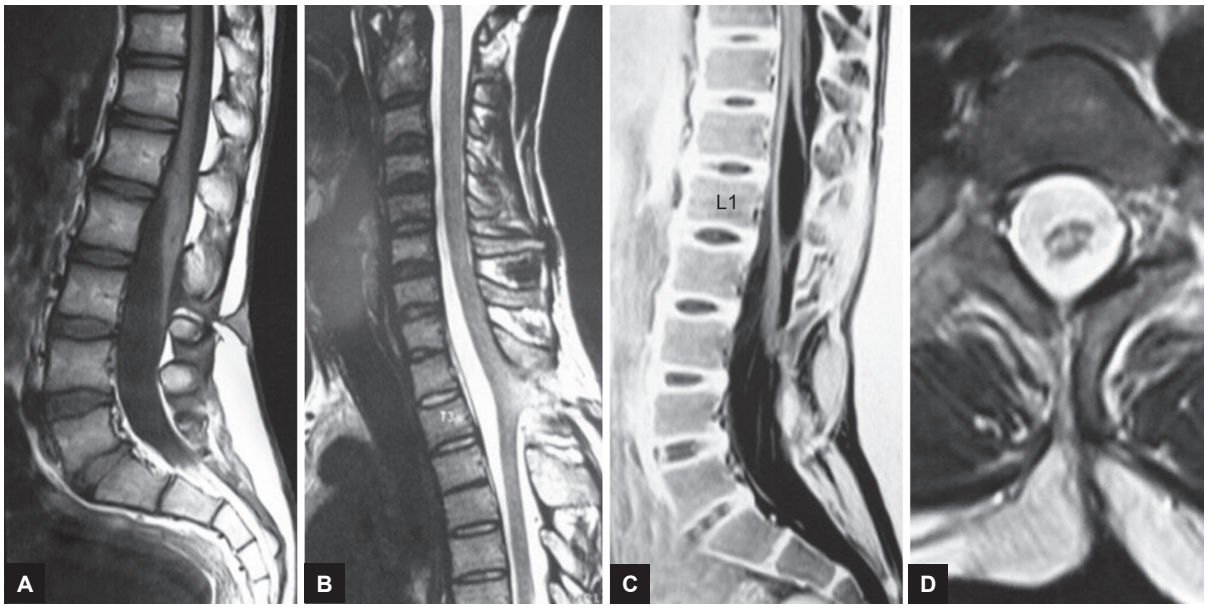
A 7-year-old male patient presented with bladder and bowel incontinence, and examination of him showed an area of hypertrichosis and a barely visible sinus in the L5 region with scoliosis of lumbar spine. The MRI examination demonstrated tethered cord with terminal syrinx and spina bifida of all lumbar vertebrae. Intradural exploration with detethering of cord was done and the tract was excised totally.

Case 3

A one-and-half-year male child was presented to the emergency department with acute onset paraplegia and bladder and bowel involvement. Barely visible sinus with surrounding inflammation over the D10 region was noted during examination. Spina bifida with DST and intramedullary abscess from D7 to D10 was found in the MRI evaluation. Emergency exploration was done and the dermoid was removed. Postoperatively, there was no improvement in the neurological status (Fig. 5).



Figs 1A to D: Various forms of cutaneous manifestations of spinal DSTs. (A) Sinus surrounded by hypopigmented patch and tuft of hair. (B) Small sinus with hair protruding through it. (C) Area of hypertrichosis and a sinus. (D) Barely visible sinus with surrounding minimal inflammation

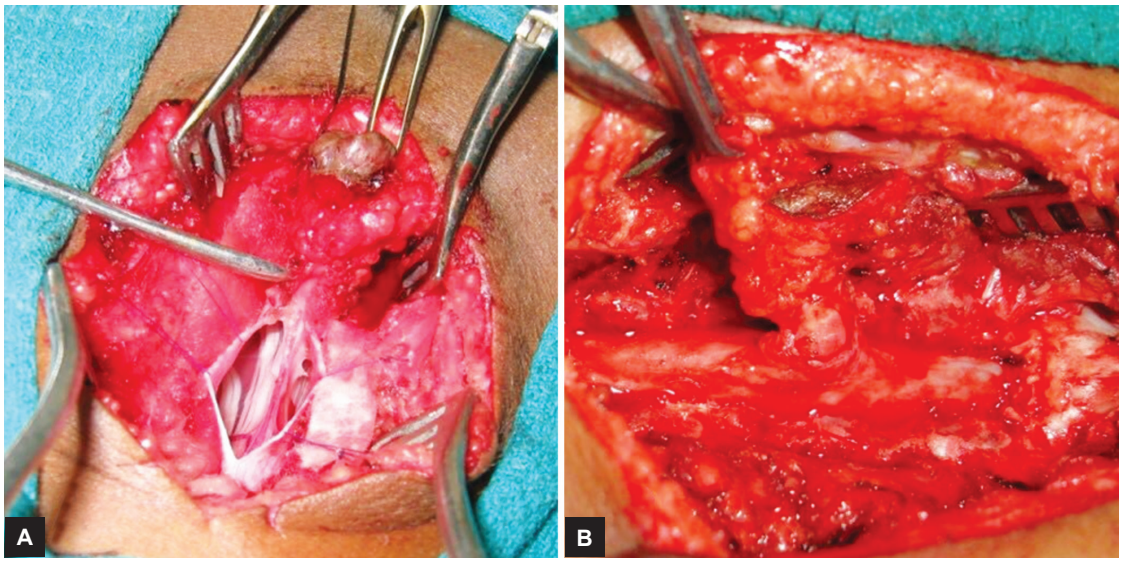


Figs 2A to D: MRI findings. (A) Low-lying cord tethered by DST. (B) Thoracic cord tethered by DST at D3 level. (C) Terminal syrinx in another patient, DST is not visible in the picture. (D) Axial MRI in another patient showing DST ending on the dura

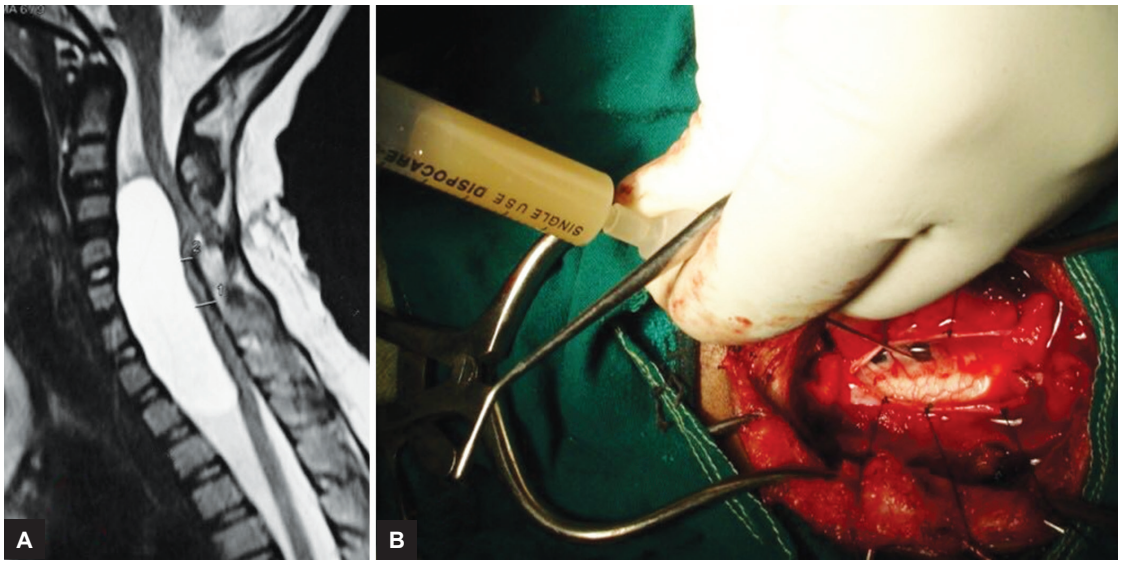
DISCUSSION

Congenital dermal sinus of spine consists of a tract lined by stratified squamous epithelium usually found on or near midline. It results from incomplete disjunction of surface ectoderm from neuroectoderm, a process likely to occur during 3 to 8 weeks of gestation.[5-7] The tract may extend inwardly up to the spinal cord. The tract develops with trapped surface ectoderm in dermal tissue and neuroectoderm. The dermatomal level of sinus may correlate with metameric level of spinal cord. The DST is usually associated with abnormalities of ectodermal, mesodermal, and neural crest derivatives which reflect its common ontogenic origin.[8]

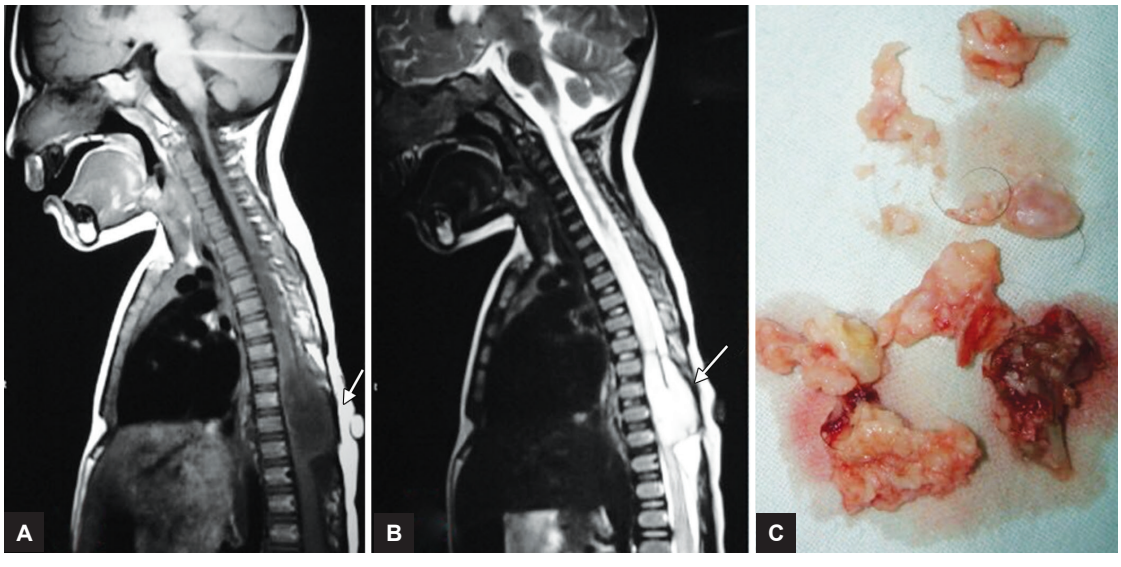
The common location of DST of spine is lumbar region (40%), though they were found anywhere between occiput to sacrum.[2-5,8-10] The cervical region is the least involved (<1%) than thoracic (10%). The cutaneous opening of DST has to be differentiated from coccygeal pits. The DSTs are found above the natal cleft and directed superiorly. Usually coccygeal pits are anatomically located below the cul de sac of subarachnoid space within the natal cleft and their tracts are either straight or directed inferiorly.[5,11]



Figs 3A and B: Intraoperative picture showing the DST entering intradurally (A) and the tract ending on the dura (B)



Figs 4A and B: (A) MRI of spine showing neurenteric cyst at cervical level compressing the cord posteriorly with DST. (B) Turbid fluid aspirated from cyst during surgery



Figs 5A to C: T1 (A) and T2 (B) images showing dermoid at thoracic level with DST. (C) Cheesy dermoid material with hair removed from it

Spinal dermal sinuses are commonly associated with cutaneous stigmatas like pit, hypertrichosis, capillary hemangiomas, and subcutaneous lipomas (Fig. 1).[12,13] Drainage of debris or fluid from sinus is noticed sometimes.[5,14] Local infection may spread to produce meningitis, which may be the presenting feature in some patients.[14-17] A spontaneous leakage of dermoid contents may cause chemical meningitis in some patients. The occurrence of meningitis may predispose patients to hydrocephalus.[18]

Presentation of DST of spine varies from asymptomatic cases to pain at the site of sinus and neurological deficits depending on the associated pathology. Diagnosis is usually made clinically. The MRI is the neurodiagnostic test of choice (Fig. 2).[5,19,20] But often, it fails to demonstrate the tract. Sometimes, ongoing infection may produce confusing picture in MRI. Ultrasonogram may be useful in selected cases to detect DST and its associated abnormalities.[21,22]

Management comprises excision of the sinus, its tract as well as intradural exploration and removal of intradural pathology (Fig. 3).[5,23] Tracts penetrating beneath the deep fascia have to be traced till its termination. Extensive scarring around the tract is likely to be present due to previous subclinical infections. Dermoids debulked from within capsule, which is usually adherent to the surrounding structures. Often, retention of epithelial surface will result in recurrence. Extensive scarring encountered during reexploration increases the likelihood of postoperative neurological deficit.

CONCLUSION

A high index of suspicion is required for diagnosing the DST of spine. The MRI is the investigation of choice. Intradural exploration is the most important part of surgical management. Associated pathologies are common and should be dealt appropriately. The outcome is directly related to the preoperative neurological status which further reiterates the importance of early diagnosis.

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