CASE REPORT

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Complete resection and untethering of the cervical and thoracic spinal dermal sinus tracts in adult patients

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ABSTRACT

Dermal sinus tracts (DSTs) of the cervical and thoracic spine are extremely rare, particularly in adult patients because diagnosis is typically made in the early stage after birth by pediatricians. These cases should be treated surgically as soon as possible to prevent neurological sequelae. This report describes two rare adult cases with cervical and thoracic spine DSTs. The first patient presented with back pain and headache, whose skin lesion had been long known, but disregarded since birth. The second patient had long suffered from residual cervical myelopathy from the prior incomplete surgical treatment. Both cases had these sinus tracts excised completely and had spinal cord untethered successfully without any neurological deterioration. There has been a trend toward earlier diagnosis of these entities, but still some cases that were diagnosed in a delayed fashion or underwent incomplete treatment are reported. Improper management during childhood could lead to irreversible neurological deficit caused by spinal cord tethering and/or direct compression due to DSTs-associated tumors. The early detection and prompt surgical intervention improve the chance of a good surgical outcome. Furthermore, complete excision of the sinus tracts and associated tumors could help prevent future bacterial contamination and recurrence.

Keywords: dermal sinus tract, cervical spine, thoracic spine, tethered cord, complete resection

INTRODUCTION

During normal embryogenesis, successful closure of the neural tube is followed by separation of the overlying ectoderm, a process termed disjunction.[1,2] Failure of disjunction results in a focal persistent connection between the cutaneous ectoderm and neuroectoderm,[3] which is termed a dermal sinus tract (DST). The most frequent sites of spinal DSTs are the lumbar region (41%), followed by the lumbosacral area (23%), the sacrococcygeal junction (13%). Appearance in the cervical or thoracic region is extremely rare.[4,5] Diagnosis is typically made in the first decade of life by pediatricians, however, it is often missed or dismissed as trivial by primary care professionals.[1]

This report describes two rare cases of adult patients with cervical or thoracic spine DST in

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a 34-year-old female and a 50-year-old male who presented with back pain and headache. The imaging showed cervical or thoracic spinal DST with dorsal tethering of spinal cord. The sinus tracts were excised surgically and untethering was conducted successfully without any neurological deterioration in both cases. We also review the English literature of adult patients with DSTs describing the presentation, early signs of symptoms, treatment options, surgical findings and outcomes. Early detection and prompt surgical intervention would significantly prevent future neurological deterioration.[1] In addition, complete surgical excision would minimize the chance of retethering and bacterial contamination.[3]

CASE REPORT

Case 1

History and Examination. A 34-year-old female presented to us with a 2-year-history of chronic back pain in the interscapular region, intermittent headache suspicious for meningitis. She also complained of occasional chest numbness in the T1-T2 dermatomal distribution and leg pain without weakness nor bowel and bladder discomfort. The results of neurological examination were otherwise completely normal. The visual inspection of her back showed a prominent dimple, approximately 5 cm in diameter, in the centre of hairy patch in the interscapular region, which ascended as it went deep (Fig. 1D). She had noticed some sort of flaky discharge from this tract on a regular basis. Her cervicothoracic spine computed tomography (CT) showed spina bifida of the T2 spinous process (Fig. 1A and C) and a T9 butterfly vertebral body (Fig. 1B). Her cervicothoracic spine Magnetic Resonance Imaging (MRI) (Fig. 2A-E) demonstrated a DST, which ran rostrally as it deepened, connecting skin with spinal cord through the space between T1 and T2 spinous processes. Spinal cord was tethered posteriorly by attaching to the DST. There was a syrinx cavity at the level of C7 and T1.

Operation. An elliptical excisional incision was placed around the hairy patch with the linear extension above and below that incision. We carefully dissected out the sinus tract and followed it all the way down to its attachment to the dura with great care not to pull it out and inadvertently injure the spinal cord. Total laminectomy of T2 and partial laminectomy of T1 and T3 were performed. The dura and the orifice of the tract into the dura were clearly identified (Fig. 3A). The dura was opened in the midline above the sinus tract, which was then followed down and around the tract (Fig. 3B). The intradural portion ascended for a few millimetres within the subarachnoid space and ended by attaching to the dorsal surface of the spinal cord with bands of fibrous tissue (Fig. 3C). The entire tract was excised after the fibrous bands tethering the spinal cord were severed to fully release the spinal cord (Fig. 3D).

Pathological findings. Histological examination showed that inner surface of the tract was lined with stratified squamous epithelium with hairs and adnexa of the skin. Inflamed granulation tissue was also found (Fig. 4A and B).

Postoperative course. Her postoperative course was uneventful without any complication. Her headache, back pain and occasional chest numbness completely disappeared. There was no evidence of communication between skin and spine on a postoperative MRI obtained at a 10-month follow-up visit. (Fig. 4C).

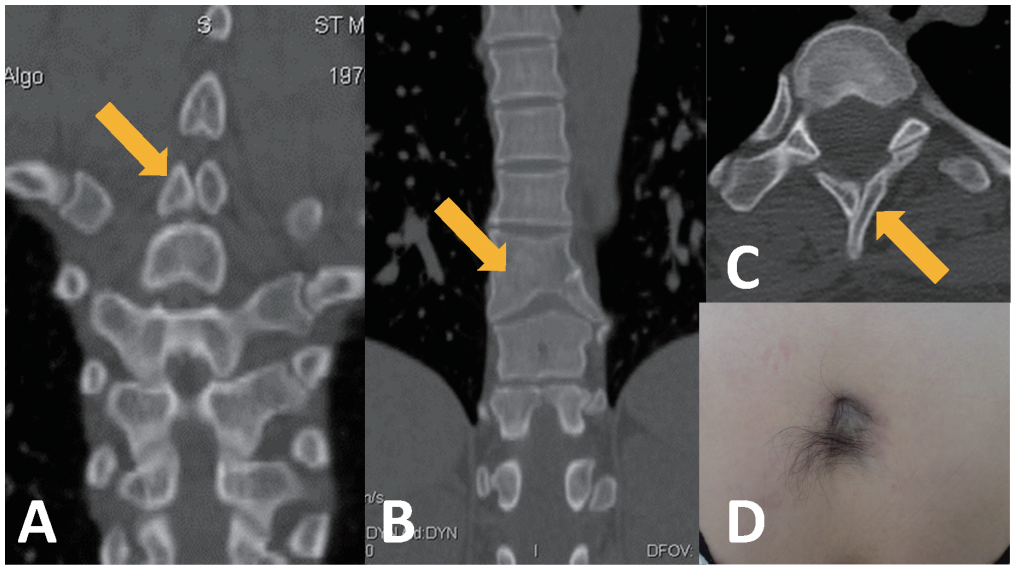

Fig. 1 Cervicothoracic spine computed tomography (CT) shows bone anomalies in association with dermal sinus tract

Spina bifida at T2 spinous process is detected on coronal image (A; arrow) and on axial image (C; arrow). T9 butterfly vertebral body is found out on coronal image (B; arrow). The visual inspection of her back shows a prominent dimple, about 5 cm in diameter, in the centre of hairy patch in the interscapular region (D). There is no infection sign, such as swelling or redness, and CSF leakage.

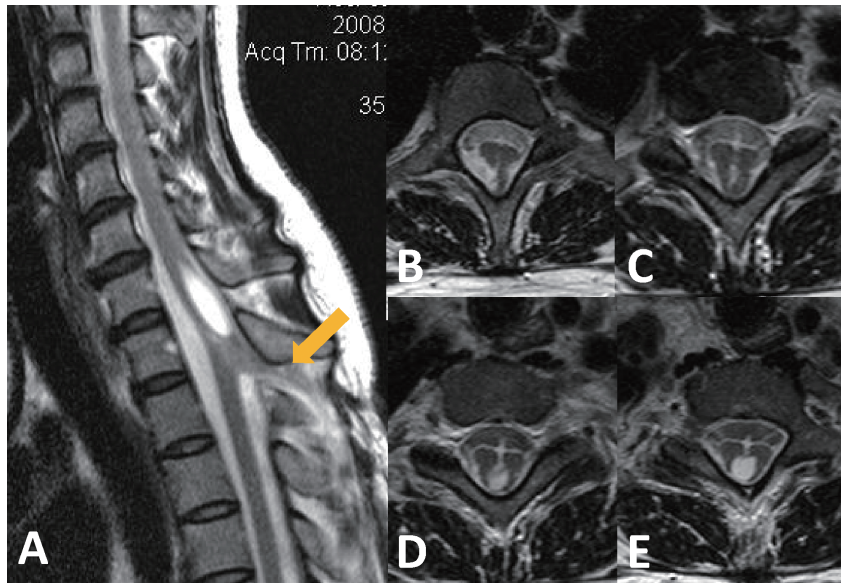

Fig. 2 Cervicothoracic spine Magnetic Resonance Imaging (MRI) T2 weighted image (T2WI) demonstrates a DST (sagittal image; A, axial image; B-E)

DST is running rostrally as it deepens, connecting skin with spinal cord through the space between T1-2 spinous processes (A; arrow). Spinal cord is tethered posteriorly by attaching to the DST. There is a large syrinx cavity expanding the dorsal aspect of spinal cord at the level of C7 and T1.

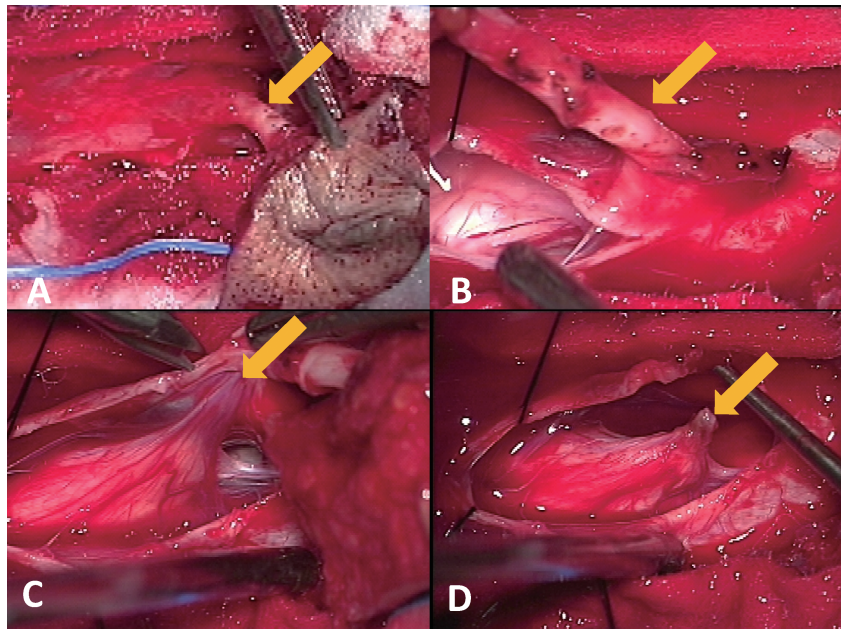

Fig. 3 Intraoperative photographs (A-D)
DST is successfully dissected off and isolated from the surrounding soft tissues (A). The dural attachment of the tract is clearly identified (A and B; arrow). The dura is opened in the midline above the sinus tract, which is followed down around the DST (B). The DST is connected to the dorsal surface of the spinal cord with bands of fibrous tissues (C; arrow), by which spinal cord is significantly displaced to dorsal direction (C). No associated tumor is observed in the subdural space (C). The fibrous band is severed to fully release the spinal cord and the entire tract is excised completely (D; arrow).

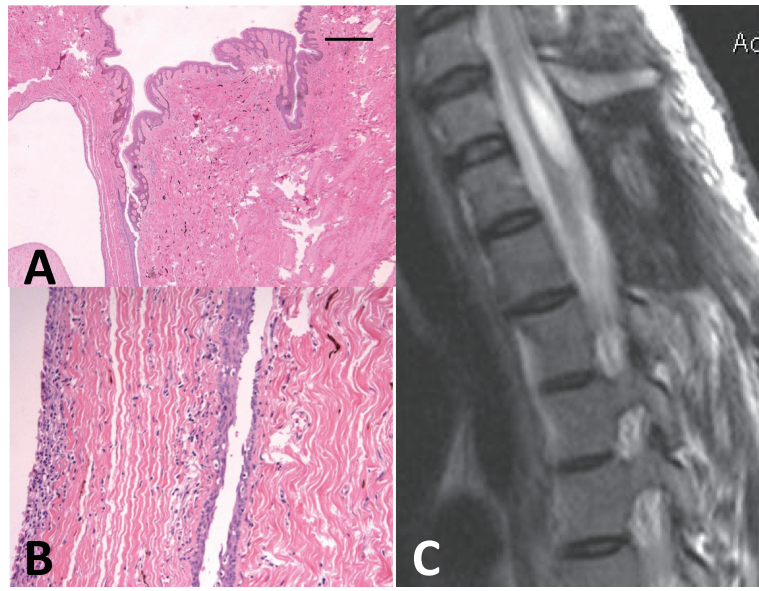

Fig. 4 Histological examination shows that inner surface of the tract is lined with stratified squamous epithelium. Inflamed granulation tissue is also found (A and B).
Postoperative MRI obtained at an 8-month follow-up visit shows there is no evidence of communication between skin and spine (C). Displaced spinal cord is restored to its normal position. Her headache, back pain and occasional chest numbness completely disappeared even as there remains a syrinx cavity.

Case 2

History and Examination. This 50-year-old male was admitted with overuse of pain medication for severe headache. He also complained of difficulty in fine finger movements and progressive gait instability. He had a remote history of previous cervical spine surgery without detailed documentation. Neurological examination demonstrated symmetrical bilateral arm weakness with grade 3 weakness in deltoid muscles, grade 4 in other muscles and an unsteady wide-based and spastic gait. On visual inspection, a prior surgical incision was found on his posterior neck and the dermal sinus tract was palpable on the skin. A cervical spine CT revealed spina bifida in the C1 and C2 laminae (Fig. 5A-C). A preoperative axial T2-weighted MRI (T2WI) demonstrated a small posterior fossa, downward displacement of the cerebellar tonsills and brain stem as well as a tethered cervical spinal cord with the presence of a DST (Fig. 6A-C). The DST was connected from subcutaneous fat tissue to cervical spinal cord through the muscle layer and spina bifida at C1-2 level. Based on these radiographical imaging findings, we speculated that the DST had been transected above muscle layer halfway to the bone in the previous surgery.

Operation. We were able to identify the sinus tract above the muscle layer and followed it all the way down to the dura mater. The DST was passing through the muscle layer and the C2 spina bifida. We performed laminectomy of C2 and partial laminectomy of C1 and C3. The DST was connected to the dura and we opened the dura around the DST, which was stuck to the spinal cord via fibrous bands. The entire tract was completely excised. We also worked laterally to the spinal cord to release it from the dural adhesions. After all these manipulations, the spinal cord became free.

Pathological findings. Histopathological examination of the surgical specimen revealed dense fibroadipose tissue with squamous epithelium lining the sinus tracts containing meningothelial elements, which was consistent with DST.

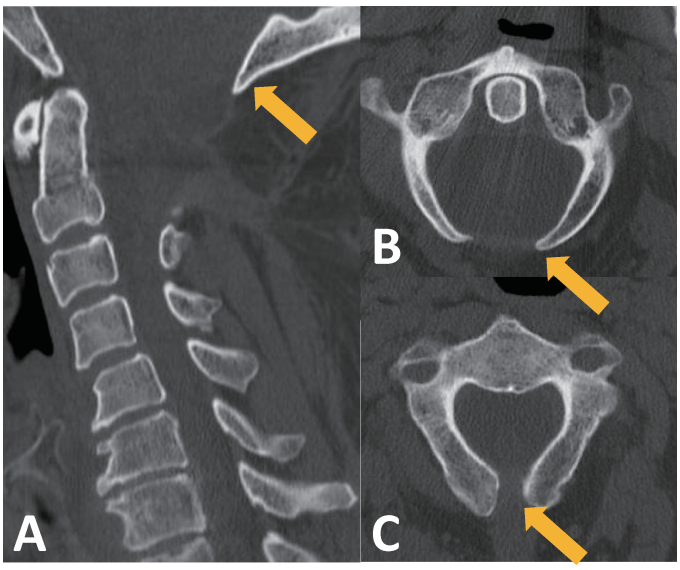

Fig. 5 A preoperative cervical spine CT reveals spina bifida in C1 (axial image; B; arrow) and C2 (axial image; C; arrow) laminae, which look like congenital anomaly given the configuration.
Occipital bone seems to be intact (A; arrow). From these findings, we assumed that there is no evidence of bony removal from the remote prior surgery.

Postoperative Course. He underwent postoperative MRI three days after the surgery, which showed that the dermal sinus had been completely resected and spinal cord had been untethered (Fig. 7A and B). His headache, ambulation difficulty, weakness and hand clumsiness had been

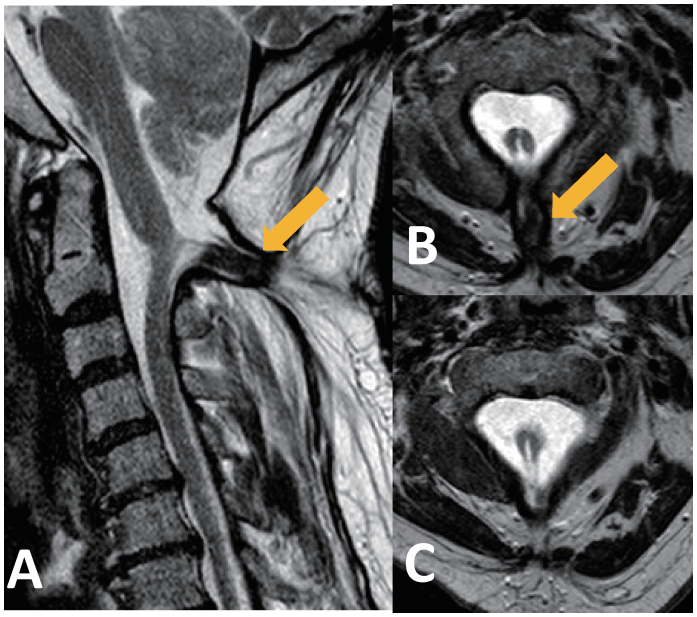

Fig. 6 A preoperative axial T2-weighted MRI (T2WI) (sagittal; A, axial; B and C) demonstrates small posterior fossa, tonsillar herniation and downward displacement of brain stem.
Cervical spinal cord is displaced to dorsal and posterior direction with the presence of a DST (A and B; arrow). DST is connected from subcutaneous fat tissue to cervical spinal cord through the muscle layer and spina bifida at C1-2 level (A; arrow). Axial image (B and C) reveals syrinx cavity.

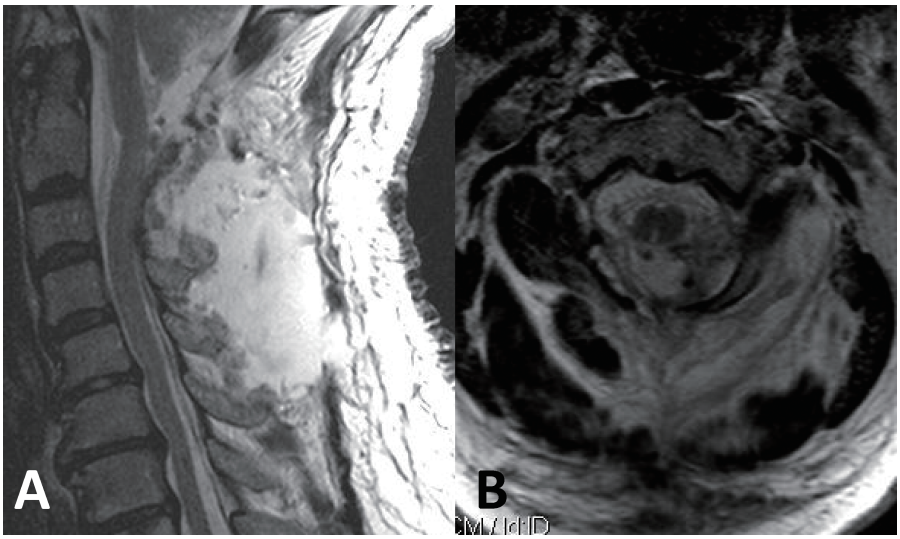

Fig. 7 Postoperative MRI (sagittal; A, axial; B) obtained three days after the surgery shows that the dermal sinus is completely resected, and displacement of spinal cord is corrected successfully (A).
Syrinx is also shrinking (B).

steadily improving at an eight-month follow-up visit.

DISCUSSION

DSTs of the cervical and thoracic spine are extremely rare, particularly in adult patients. The neuroectoderm is separated from the cutaneous ectoderm along the dorsum of the embryo.[5,6] Once the cutaneous ectoderm has fused at the surface and the neuroectoderm has closed at the depth forming the neural tube, both layers become totally separated.[2,6] The closure of the neural tube starts in the middle of the embryo and then proceeds both cranially and caudally,[6] therefore, a greater incidence of these cutaneous anomalies has been noted at the occipital and lumbosacral regions, which are the last parts of the closure.[2,6] Previous reports say that 1% of DSTs were located in cervical and 10% in thoracic spine.[5]

Diagnosis

With increasing awareness of the potential significance of cutaneous findings over the cranial-spinal axis, there has been a trend toward earlier diagnosis of these entities.[2,4] However, there are still some cases that have their pathology detected later in their lives in spite of obvious cutaneous manifestations during childhood even with some sort of discharge suspicious for recurrent meningitis like the present case 1. Examination of a child's back is particularly important if they develop recurrent meningitis. We reviewed and analyzed cervical and thoracic spine DSTs cases which were treated in their adulthood (adults are defined as more than 15 years old)[3-5,7-14] (Table 1 and 2). Even though almost all cases showed obvious skin dimples which were noticed in childhood, many of them tended to be disregarded as long as they were asymptomatic. This is most likely an issue related to physicians' awareness of a rare entity. We should associate symptoms, such as pain along the spinal axis, fever and neurological deficit, with skin lesions. Once a suspicious skin lesion is detected irrespective of symptoms, the neuroimaging modalities are promptly used to detect possible DSTs because there is a consensus for early surgical intervention. CT and plain X-ray are clearly able to demonstrate associated bone anomalies. MRI can demonstrate the extent and depth of the sinus, reveal additional intradural pathologies (other dysraphic anomalies, inclusion tumors, infectious complications, cord tethering) and assist with surgical planning.[2,5]

Tethered cord syndrome

Adult patients with DSTs tend to present with pain along the spinal axis with or without neurological manifestations. These symptoms are features of tethered cord syndrome.[2] The tethering forces pull and stretch the spinal cord posteriorly, which is called dorsal tetherin. Tethered cord syndrome is sometimes related to associated tumors such as dermoids, epidermoids or teratomas[2,11,15,16] or split cord malformations.[13,16] Direct compression of the spinal cord caused by these tumors is also an important factor for delayed neurological deterioration. According to previous reports,[3,17] DSTs patients with associated tumors tended to end up with a poor outcome because of the complexity of the surgery. In contrast, intradural fibrous bands, deemed as another cause of cord tethering, were found in seven cases in our analysis and required only simple transection. These cases were likely to make a good recovery.

Management

Surgical management for every case of DSTs should consist of intradural exploration and addressing any identified cause of cord tethering (fibrous bands, associated split-cord malforma-

Table 1 Review of published cases with cervical and thoracic spine DSTs; clinical presentation

Cervical cases	Authors and year	Age and sex	Location	Early sign in childhood	Treatment in childhood	Reason for neurosurgical referral	Skin lesion	Tethe red cord	Syrinx
1	Mixter 1932	23M	C3	NA	NA	NA	no	NA	NA
2	Sachs 1949	35M	C6–7	developmental delay, flaky material discharge from skin lesion	no specific treatment	arm and leg numb. and weak, gait disturbance	yes	yes	NA
3	Eller 1987	23F	C1	arm and leg weak., flaky material discharge from skin lesion	no specific treatment	H/A, arm and leg numb. and weak.	yes	yes	NA
4	Wu 1990	37M	C2–3	R. arm intemittent numb.	surgery (incomplete)	arm and leg weak. and difficulty in breathing	yes	yes	NA
5	Wu 1990	43M	C3–4	no	no specific treatment	arm weak. and numb., hand clumsiness	no	yes	yes
6	Ackerman 2002	16M	C5–6	NA	surgery	worsening of myelopathy	previous incision	yes	NA
7	Ackerman 2002	41F	C2	NA	surgery (incomplete)	neck pain, worsening of myelopathy	previous incision and CSF leak	yes	NA
8	Dagcinar 2008	24F	C3	no	no specific treatment	neck pain	yes	yes	yes
9	Coumans 2011	52F	C6	no	no specific treatment	arm numb.	yes	yes	yes
10	present case 2 2019	50M	C2	NA	surgery (incomplete)	H/A, arm weak, and numb., hands clumsiness, gait disturbance	previous incision	yes	yes
Thoracic cases	Authors and year	Age and sex	Location	Early sign in childhood	Treatment in childhood	Reason for neurosurgical referral	Skin lesion	Tethe red cord	Syrinx
11	Ottonello 1933	20F	T3	NA	NA	NA	yes	NA	NA
12	List 1941	19M	T2–3	NA	NA	NA	yes	NA	NA
13	Tytus 1956	40M	T1	NA	NA	NA	yes	yes	NA
14	Aydin 2001	24F	T1	scoliosis (asymptomatic)	no specific treatment	arm weak. and scoliosis	yes	yes	yes
15	Manfredi 2001	73F	T2–4	no	no specific treatment	arm and leg weak., gait distubance	yes	yes	yes
16	Ackerman 2002	55M	T11	no	no specific treatment	pain, arm and leg weak. and numbness	no	yes	no
17	Kara 2003	37M	T2	no	no specific treatment	H/A, arm numb. and spasm	yes	NA	no
18	present case 1 2019	34F	T1–2	flaky material discharge from skin lesion	no specific treatment	H/A, back pain, leg pain, chest numbness	yes	yes	yes

Abbreviations: NA= not available, numb. = numbness, weak. = weakness, H/A = headache

Table2 Review of published cases with cervical and thoracic spine DSTs; surgical findings and outcome

Cervical cases	Authors and year	Treatment	Associated lesion	Cause of tethering	Neurological status at final f/u	Final f/u
1	Mixter 1932	surgery	no	NA	NA	NA
2	Sachs 1949	surgery	dermoid	dermoid	arm and leg improved gait became stable	6 months
3	Eller 1987	surgery	no	fibrous band	leg improved arm unchanged	NA
4	Wu 1990	surgery	lipoma, neurenteric cyst	lipoma, neurenteric cyst	died of enlarging of cavitation and gliosis in spinal cord	18 months
5	Wu 1990	surgery	no	fibrous band	weak. and numb. resolved	6 months
6	Ackerman 2002	surgery	no	fibrous band	myelopathy improved	2 months
7	Ackerman 2002	surgery	no	NA	neck pain disappeared, myelopathy improved	7 months
8	Dagcinar 2008	surgery	no	fibrous band	neck pain disappeared	12 months
9	Coumans 2011	surgery	no	fibrous band	arm numb. improved	NA
10	present case 1 2019	surgery	no	fibrous band	H/A, hand clumsiness, arm weak. improved	2 months
Thoracic cases	Authors and year	Treatment	Associated lesion	Cause of tethering	Neurological status at final f/u	Final f/u
11	Ottonello 1933	surgery	dermoid	dermoid	NA	NA
12	List 1941	surgery	dermoid	dermoid	NA	NA
13	Tytus 1956	surgery	dermoid	dermoid	NA	NA
14	Aydin 2001	surgery	epidermoid tumor	epidermoid tumor	NA	NA
15	Manfredi 2001	refused surgery	lipomyeloschisis	lipomyeloschisis	NA	NA
16	Ackerman 2002	surgery	neurenteric cyst split cord malformation	split cord malformation	pain, arm and leg improved	10 months
17	Kara 2003	surgery	menigocele	not mentioned	H/A, arm numb, spasm improved	6 months
18	present case 2 2019	surgery	no	fibrous band	neck pain disappeared	8 months

Abbreviations: R = right, L = left, numb. = numbness, weak. = weakness, NA = not available, f/u = follow up

tion, lipomyelomeningocoeles, myelomeningocoeles, and tumors such as dermoid, epidermoid tumors).[4,9] All cases except for case 15, who declined the offer of surgical intervention, underwent surgical treatment.[3-5,7-14] It is imperative to follow the sinus tract all the way down to the dura and open it for intradural exploration. Four cases (case 4,6,7,10) had undergone surgery during childhood, three of which (case 4,7,10) including the present case 2 were incomplete due to poor understanding of anatomy. In these cases, tracts were misunderstood as being finished at

the layer of fascia. Total resection of intradural tumors and adequate untethering of spinal cord should be performed and confirmed with restoration of spinal cord pulsation. Lateral dissection of spinal cord for arachnoid adhesion is sometimes required to improve CSF flow like present case 2 since retethering of spinal cord could be caused by postoperative arachnoid adhesions like case 6. Real-time intraoperative ultrasound can also provide excellent surgical guidance for assessment of untethering by demonstrating spinal cord pulsations. A complete removal of the sinus tract facilitates a sterile excision by preventing any breach to this surface.[3] Additionally, developing an excisional plane around the tract leads to greater confidence in a complete excision of the DSTs.[3] Even if a patient is asymptomatic, the cord should be surgically released.[2-4] Further, if any tissue from a DST–associated tumor is left behind at surgery, this could lead to recurrence. Thus, complete removal of this lesion is mandatory.

Outcome

Early referral and treatment, especially at the stage of cutaneous markers alone or local pain without other neurological deficits, would likely result in better outcomes. Most reported cases achieved some improvement postoperatively, however, complete resolution was observed only in patients with pain along the spinal axis. Since neurological deficits indicate some sort of neural injury as a result of spinal cord stretching, it is very hard to expect a remarkable recovery.

In conclusion, patient outcome is adversely affected by the development of infection or a delay in treatment once symptoms develop, leading to irreversible neurologic damage. Thus, early surgical excision of the tract with intradural exploration is mandated. We should be made more aware of this condition and should associate a cutaneous lesion with pain along the spinal axis, recurrent meningitis, and neurological deficit.

CONFLICT OF INTEREST DISCLOSURE

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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