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## Case Report

# Misdiagnosed adult presentation of diastematomyelia and tethered cord

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## ABSTRACT

Diastematomyelia with tethered cord is an uncommon congenital anomaly that is generally diagnosed in childhood but may rarely present in adulthood, we present the case of a 48-year-old man with diastematomyelia and tethered cord whose diagnosis was initially missed, leading to unnecessary spine surgery. The correct diagnosis was made from follow-up imaging. Because common clinical complaints such as back pain may be caused by unusual conditions, the authors suggest that radiologists and treating physicians should remain vigilant for unusual presentations of rare diseases.

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## Background

Diastematomyelia also known as split cord malformation is a rare entity at which the spinal cord is split into 2 halves at sagittal plane [1]. Presenting with specific symptoms and signs, the patients are usually diagnosed in childhood. Adult presentation is very rare with few case reports published in the literature. Authors present a case of late presentation of diastematomyelia with tethered cord in a middle aged adult who was misdiagnosed and mistreated for disc herniation.

## Case presentation

A 48-year-old man complaining from gradual weakness of lower limbs and paresthesia for 1 year—increased for last 2 months—that limited his routine daily movements and was prescribed a lumbar spine magnetic resonance imaging (MRI) by an orthopedic surgeon.

The MRI exam performed by 0.4 T scanner reported multilevel disc bulges and herniation. The patient subsequently underwent laminectomy based on the given MRI report. In the following weeks after surgery, the patient developed

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging.

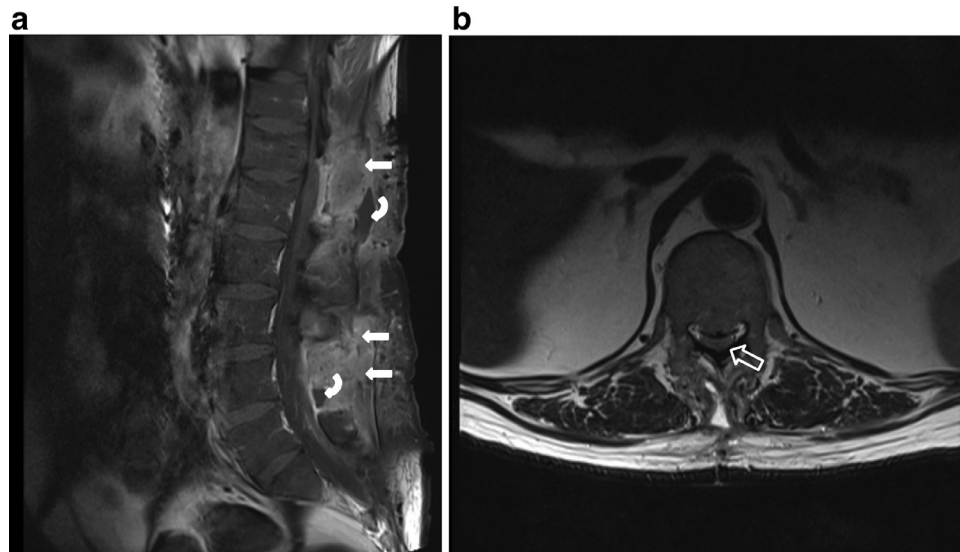
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**Fig. 1 – 48-year-old man with diastematomyelia and postoperative complications from laminectomy. (A) Postcontrast T1WI sagittal view: marked soft tissue enhancement in the paraspinal soft tissues (white arrows) with multiple abscess formation (curved white arrows) representing postsurgical infectious process. (B) Axial T2WI: absolute stenosis of the spinal canal at T12 level.**

symptoms for surgical complications and was prescribed a second MRI. The patient came to the authors department for the second MRI examination. MR imaging was performed with 1.5 T Siemens scanner 3 weeks after the surgery. The images revealed postlaminectomy status at L1 level and postsurgical infectious process and abscess formation in the para spinal muscles (Fig. 1). More to that, it was surprisingly seen in T2-weighted sagittal cuts that the discs did not have as much bulges or herniation to be accepted as the cause of patient's clinical symptoms (Fig. 1). Further look of the vertebral bodies alerted the radiologist that there is absolute stenosis of the spinal canal at T12 level and surprisingly it is caused by posterior indentation of the vertebral body rather than the disc (Fig. 1B). No suspected mass lesion was seen in the vertebral body to think of any neoplastic lesion as the cause of posterior indentation nor was there any history of traumatic injury present to think of traumatic cause. The conus medullaris was not seen to terminate at normal L1/2 level, but the cord still descended to L4 level (tethered cord) (Fig. 2). While reviewing the axial images it was found that 2 separate thecal sacs with 2 hemicords were present (Fig. 2B). Complementary computed tomography cuts were taken from the region which revealed bony spur arising from the superior-posterior aspect of the L1 vertebral body and divided the spinal canal to 2 halves incompletely (Fig. 3). All these findings confirmed the diagnosis of diastematomyelia (type I) and tethered cord. Retrospective view of available sagittal images of previous MR exam, also showed that there was no significant disc bulge/herniation while the posterior bony indentation was clearly visible at L1 level (the bony spur) (Fig. 4).

Further questions about clinical history showed the patient had no clinical signs and symptoms prior to 1 year. Normal urinary bladder and anorectal function was present. As the

patient was under treatment in other facility, hence he was lost of follow-up to evaluate the long-term management plan and outcomes.

## Discussion

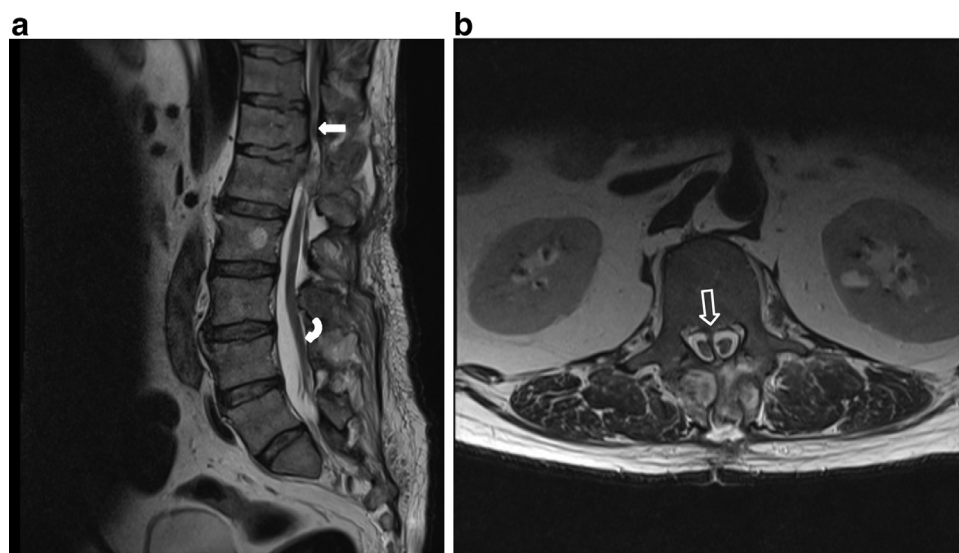
Diastematomyelia is a rare congenital disorder in which spinal cord is longitudinally split into 2 halves [2]. The disease was first reported by Olivier in 1837 [3]. It constitutes about 4% of all congenital spinal defects and is 3 times more common in females compared to males [1].

The condition could arise either due to an abnormal development of notochord between the 15th and 18th days of pregnancy or abnormal adhesion of ectoderm and endoderm. This entity can be associated with other vertebral anomalies (ie, spina bifida, scoliosis, lordosis, butterfly vertebrae, and hemivertebrae) tethered cord syndrome, Chiari malformations, dermoid, lipoma, and syringohydromyelia [1,2].

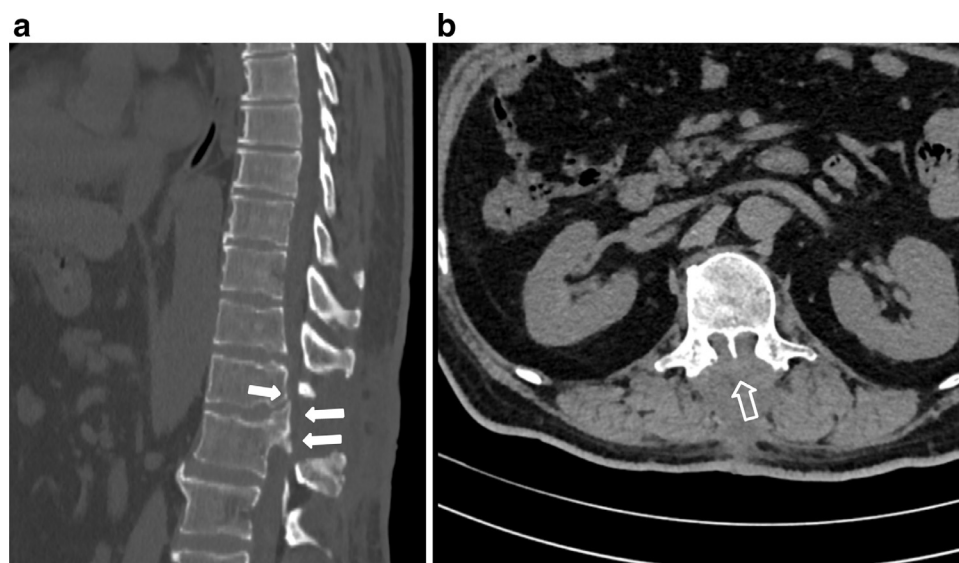
The disease usually involves T9 to S1 vertebral levels: 50% being between L1 and L3 and 25% between T7 and T12. Cervical involvement is very rare.

Diastematomyelia has 2 types. Type I makes 40%-50% of cases which has 2 separate dural sacs each containing a hemicord. The 2 sacs are separated—symmetrically or asymmetrically—by a bony spur. Type II makes 50%-60% of cases which has single dural sac containing both hemicords. The cord is divided—symmetrically—into 2 halves by a fibrous intradural spur [4].

The patients are diagnosed mainly in the childhood—majority being under 7 years—but they rarely may have adult presentation [5]. Cases with very late diagnosis are reported in the literatures being as 72 and 87 years of age [4].



**Fig. 2 – (A) Sagittal T1WI;** no significant disc bulge or herniation is seen at any level along the lumbar spine to be accepted as the cause of patient's clinical presentation. However, there is absolute stenosis of the spinal canal at T12 level caused by bony posterior indentation of the vertebral body rather than the disc herniation (white arrow). No suspected mass lesion is seen in the vertebral body to think of any neoplastic lesion as the cause the posterior indentation of the vertebral body. The cord descends to L4 level representing the tethered cord (curved white arrow). **(B) Axial T2WI show split cord with 2 separate thecal sacs (open arrow).**

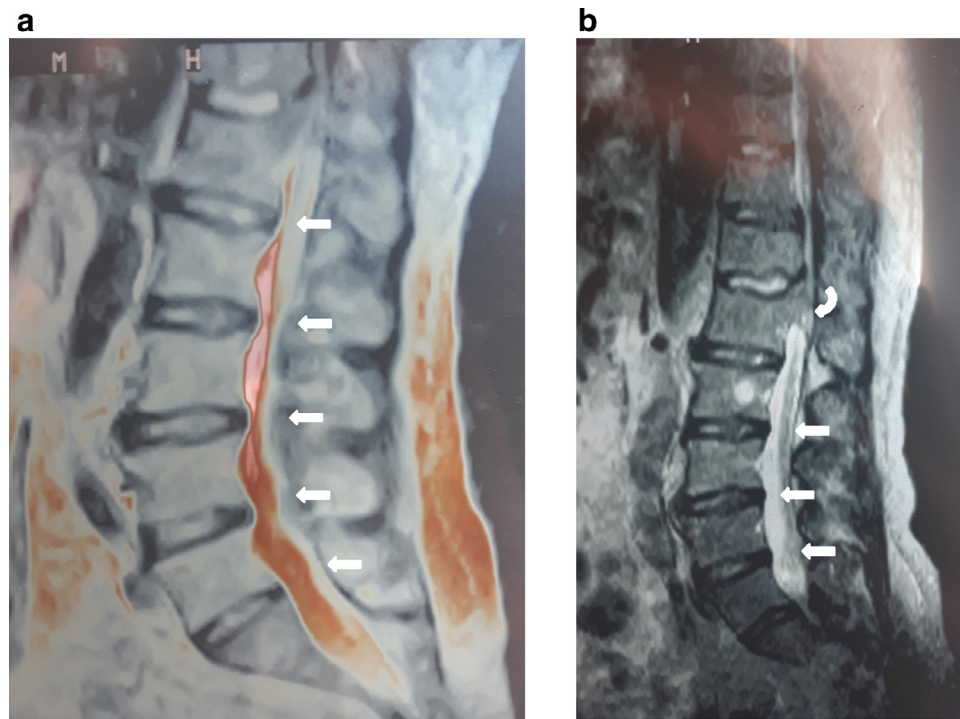


**Fig. 3 – (A) Sagittal CT section bone window showing posterior bulging of T12 vertebral body and upper half of L1 vertebral body is seen. (B) Axial CT section, soft tissue window showing bony spur arising from the superior-posterior aspect of the L1 vertebral body and divided the spinal canal to 2 halves.**

The clinical signs and symptoms could arise due to stretching and compression of spinal cord by the septum. Usually, diastematomyelia causes lumbago, lumbosciatica, or perianal dysesthesia in adults. Some patients may have asymmetry in the buttocks, leg muscles, or gluteal folds [4].

Antenatal ultrasonography in the third trimester can detect the disease by visualization of an echogenic focus (the spur) between the fetal spinal lamina spreading out the poste-

rior elements [1,2]. In children and adults, MRI is the modality of choice for depiction of the split cord, the dural sacs, tethered cord, and other accompanying vertebral or cord anomalies. Computed tomography scan depict will the bony spurs and abnormalities of the vertebral bodies and posterior arches. Functional evaluation of spinal cord and cauda equina can be done via somatosensory evoked potential or urodynamic studies, which are considered important tools besides radiologic examinations [4].



**Fig. 4 – Retrospective photograph of sagittal hard copy of preoperative low field MRI. (A) There is no significant disc bulge at lumbar levels (white arrow). (B) The bony posterior indentation (curved white arrow) at superior posterior aspect of L1 vertebral body is seen at midsagittal level (the bony spur). \*(The images are of low quality as they were taken from relatively damaged hard copies of old films.)**

The treatment options are controversial. Type I anomaly usually requires surgical treatment due to the deterioration of neurologic symptoms [4]. Prophylactic removal of spur is not recommended in asymptomatic adults except if the patient has scoliosis. Lewandrowsky and Boossaadani suggested surgery for patients who have spinal canal stenosis or tethered cord syndrome. Cheng et al suggested surgical treatment for patients with following criteria: prophylactic surgery for children with type I disease, deteriorative neurologic symptoms, preventing injury to spinal cord before correction of spinal deformity, and finally patients with backache radiating to lower limb [4].

Asymptomatic patients do not require any treatment while those with tethered cord syndrome can get benefit from surgical release of spinal cord and resection of bony spur [1].

We conclude the discussion with specific suggestions and messages from the case:

- Congenital anomalies like diastematomyelia may still present in adulthood, albeit rarely, therefore radiologist, spine surgeon, neurosurgeon, and orthopedist should remain aware of unusual diseases while dealing with patients.
- Sometimes, delicate and subtle clues may guide the radiologist and clinician in detecting erroneous imaging findings; therefore, physicians should be open to reviewing the work of other team members when curious features are found.

### Conflict of interest

None.

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