

## Case Report

# Acute Brown-Sequard syndrome caused by thoracic disc herniation in atypical Scheuermann's disease: a case report

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**Abstract:** *Objective:* The purpose of this study was to report a rare case with acute progressing Brown-Sequard syndrome due to a soft thoracic disc prolapse in atypical Scheuermann's disease (SD), and discuss its clinical characteristics, current diagnosis and surgical treatment with literature reviews. *Methods:* A 46-year-old Chinese male patient in good general health was admitted for brutal back pain associated with progressive motor impairments of left extremity and walking disorders, accompanying with fecal and urinary incontinence for 9 hours after the long bus ride. The case was verified by personal history, physical examination, laboratory tests, X-ray, CT and MRI findings. *Results:* CT scan revealed evident irregularities and Schmorl nodules in thoracic endplate surfaces, anterior wedging in vertebra corpuses (T11-12). MRI images showed severe spinal cord compression due to an anterior lesion of disc protrusion at the T9-10 disc level. The patient underwent discectomy followed by interbody fusion (T8-11), and satisfactory therapeutic outcomes were obtained after three months of surgery. Frankel grades improved from C to D in the patient. *Conclusion:* Symptomatic thoracic disc herniation (TDH) could be seen as a neurological complication of SD. Early decompression surgery should be carried out as early as possible for patients in atypical Scheuermann's disease with acute spinal myelopathy or paraplegia caused by a protruded disc.

**Keywords:** Brown-Sequard syndrome, thoracic disc herniation, Scheuermann's disease, neurological complication, spinal surgery

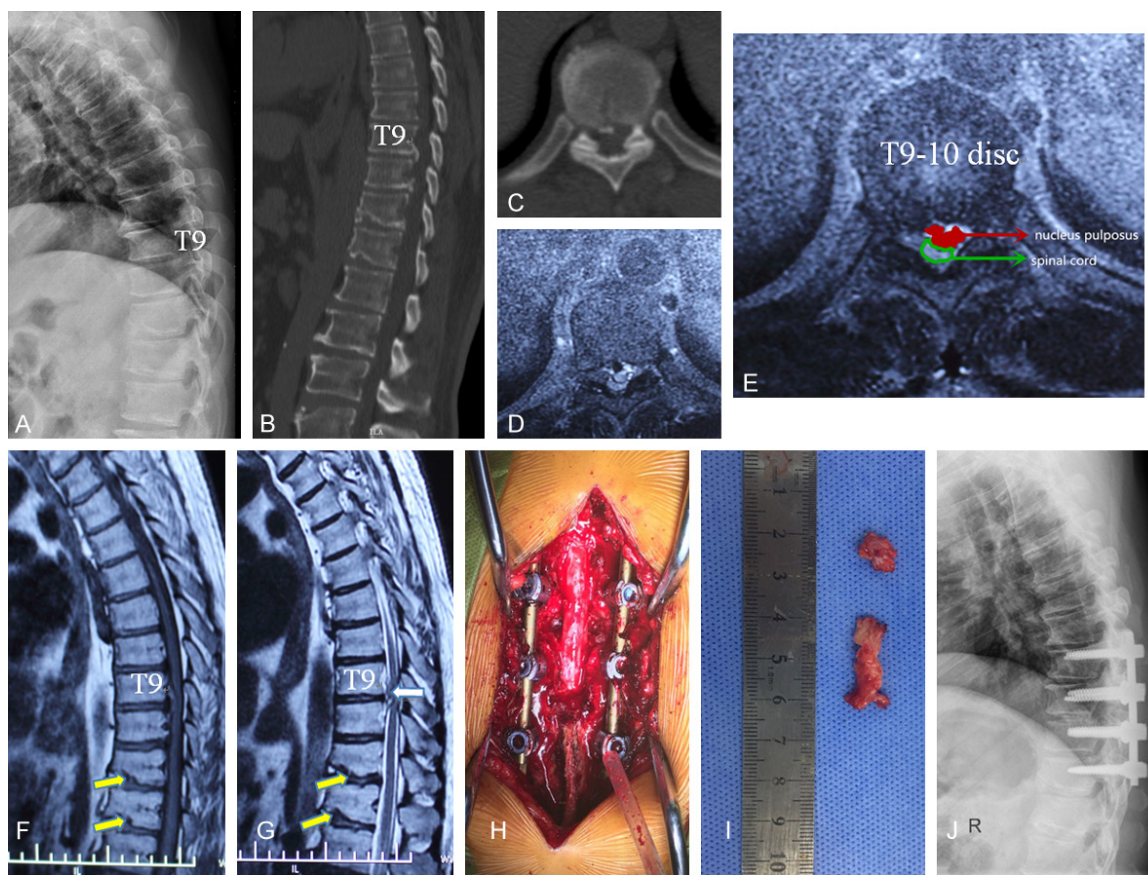
## Introduction

Thoracic disc herniation (TDH) is relatively unusual and accounts for only about 0.25%-0.75% of all symptomatic herniated discs in adults [1]. Majority of posterior thoracic disc herniations are quite small and clinically insignificant [2]. Approximately 4% of TDHs patients are with an acute myelopathy [3]. What is more, acute spinal cord hemisection syndrome due to intervertebral disc protrusion is even less common and more serious.

Interestingly, we particularly noticed that the patient visiting our emergency department (ED) who presented with acute Brown-Sequard syndrome due to TDH also had four special radiologic features, which are Schmorl's nodes (SN), wedge-shaped vertebrae (WV), irregular vertebral end plates (IE) and posterior bony avulsions of the vertebrae (PBA) with calcifications.

By coincidence, these changes were all related to another spinal disorder: Scheuermann's disease (SD) [4-6].

Scheuermann's disease, also known as juvenile kyphosis dorsalis traditionally, a painful structural kyphosis of the thoracic spine [7-9], may result from excessive mechanical stress during the growth of the spine on a weakened vertebral endplate from a genetic background [10]. The typical radiographic aspects [11] are related to the vertebral endplate lesions and include vertebral wedging of more than 5° of at least three adjacent vertebrae at the apex of the kyphosis, irregularity of the vertebral endplate and Schmorl's node. For patients with only one or two WV, and no notable kyphosis, but characteristic endplate/disc lesions, the term "atypical Scheuermann's disease" was proposed [12-14]. Unlike the classical form that was usually asymptomatic, the atypical form was most



**Figure 1.** There was no notable thoracic kyphosis on an X-ray (A). Computed tomography scan showed evident irregularities and Schmorl nodules in thoracic endplate surfaces, anterior wedging in vertebra corpora (T11-12) (white arrows) (B, C). MRI images showed severe spinal cord compression due to an anterior lesion of disc protrusion at the T9-10 disc level (D-G). Illustration of an axial view of the spine between T9 and T10 (D, E) showed the compressive effects of the disc herniation on the sensory and motor pathways of the spinal cord on the left side. MRI scans (T2-weighted) of the spine in axial (E) and sagittal (G) view, showing a large, herniated intervertebral disc between T9 and T10 (red arrow) with spinal cord compression. Although only two wedged vertebrae (T11 and T12) are observed, Schmorl's nodes, irregular end plates, wedge-shaped vertebrae and disc-space narrowing coexist. Thus, he was diagnosed with SD by the modified Heithoff's criterion. Intraoperative photographs (H, I): The disc-like material was removed in 2 pieces, and the sizes of the lesions were 2.2\*0.6 cm and 0.9\*0.7 cm. Postoperative X-ray indicated the stabilization of thoracic vertebrae (J).

often painful [15]. In most patients, the radiographic study is sufficient to establish the diagnosis and differentiate both forms [16].

In this study, we describe one case of acute spinal cord hemisection syndrome in atypical Scheuermann's disease adult due to spinal cord compression caused by herniated thoracic discs, and discuss its clinical presentation, neuroimaging findings, diagnostic considerations and surgical treatment with literature reviews.

### Case presentation

An appropriate written informed consent was obtained from the patient reported in this case.

A 46-year-old Chinese male patient in good general health was admitted for brutal back pain associated with progressive motor deficit of left extremity and walking disorders, accompanying with fecal and urinary incontinence for 9 hours after the long-distance bus ride. No other obvious triggers were identified. Physical examinations revealed sensation to sharp touch and temperature was diminished in the dermatomes on the right side caudal to T9. Moreover, the following signs affected the patient's left side: an almost complete deficit in all myotomes of the lower extremity (muscle force for zero level), abnormal proprioception of the toes, brisk reflexes and an upgoing plantar reflex, and positive Babinski sign. Laboratory

tests: White blood cell (WBC):  $8.4 \times 10^9/L$  (3.5-9.5), Erythrocyte sedimentation rate (ESR): 5.1 mm/h (<16), C-reactive protein (CRP): 1.72 mg/L (0.00-7.44), Calcium: 2.34 mmol/L (2.05-2.85), and Blood urea nitrogen: 6.45 mmol/L (1.79-7.14) were within normal limits. No previous spine injection was performed previously. Complementary exams have eliminated neurological, cardiologic and infectious diseases. There was no notable thoracic kyphosis on an X-ray (**Figure 1A**). Computed tomography (CT) confirmed posterior central herniation within the disc space at the T9-10 level and posterior bony avulsions of the vertebrae with calcifications (**Figure 1B, 1C**). Signs of Scheuermann's disease consisted on multi-levels endplates irregularities and Schmorl's nodes more pronounced in thoraco-lumbar junction (T11-12). Magnetic resonance imaging (MRI) revealed severe hemi-spinal cord compression on the left side between the T9-10 disc level from an anterior lesion, in which there was a hyperintense core and surrounding hypointense area (**Figure 1D-G**). Acute idiopathic epidural hematoma, abscess or thoracic disc herniation could be suspected from the primary clinical and imaging diagnosis.

The patient, without predisposing conditions for disc herniation, underwent prompt posterior surgical decompression and resection of the protruded disc, which required division of the T10 nerve root on the left side, followed by interbody fusion with instrumentation (**Figure 1H-J**). Using a small curette and a pituitary rongeur, the soft disc was easily removed in a piecemeal fashion. The disc-like material was removed in 2 pieces, and the sizes of the lesions were 2.2\*0.6 cm and 0.9\*0.7 cm (**Figure 1I**). The remaining little calcified substance was removed by continuous irrigation and suction. Soon after the emergency surgery, the patient had complete relief of his backache, but he did not reveal any substantial functional improvement after two months of surgery. However, there was a significant incremental recovery of neurological deficit in about three months. Frankel grades improved from C to D. The patient regained ambulatory function and only experiences the expected left-sided sensory deficit at the T10 level. Of course, he still had slight neurological damage, like hypoesthesia.

### Discussion

Scheuermann's disease (SD) is the most common cause of degenerative structural thoracic or thoracolumbar hyperkyphosis associated with back pain in adolescents and could be observed in two patterns, a typical (thoracic) more common pattern (1-8% of the population) and an atypical (thoracolumbar) less common one (0.4-4% of the population) [17, 18]. In terms of radiological examinations, some authors consider the presence of wedging in at least three successive vertebra at an angle of over 5° as diagnosis criterion for typical SD [17], while others think wedging in a vertebra along with irregular endplates enough as diagnosis criterion of atypical SD [19, 20]. The presentation of atypical SD in all patients was more endplates irregularities with Schmorl's nodes (SN) and trail sign than vertebral bodies wedging. Adequate evidence has shown that SD and its radiographic changes including SN, irregular vertebral end plates (IE), wedge-shaped vertebrae (WV) and posterior bony avulsions of the vertebrae (PBA) with calcifications were all found to enhance thoracic disc degeneration (TDH) [21-24].

Viewing this entity in this way could help in explaining the etiology of acute spinal cord hemisection syndrome due to TDH in atypical SD in our case. The thoracic spinal cord would account for a large proportion of volumes in the spinal canal. Once anterior or posterior compression occurs, cushion space of the thoracic spinal cord would be very small. PBA was considered an avulsion of the posterior ring apophysis due to the lack of fusion between the separated bony mass and the vertebral body [21]. It was frequently found to appear together with other signs of SD [21]. Previous studies already demonstrated PBA would occur at the lower lumbar spine at a low incidence and tend to promote symptomatic disc herniation [21, 25, 26]. The high incidence of PBA in symptomatic TLDH patients may be partly attributed to increased thoracolumbar kyphosis, as observed in this series, which could produce increased stretching stress on the posterior annulus of the discs. Morphologically, PBA corresponds to the "anterior Schmorl's node" [23], which itself may be anterior bony avulsion, found in "lumbar SD" because of the lordosis of the lumbar spine. It is possible that increased

thoracolumbar kyphosis in symptomatic TLDH patients accelerates posterior disc herniation by means of PBA, whereas the lordosis of the lumbar spine, in contrast, slows disc herniation by an opposing mechanism. This finding may explain why numerous symptomatic TLDH patients do not have disc herniation in the lower lumbar spine that is naturally more mobile than the thoracolumbar spine, with T11 to T12 most commonly described [27].

Neurological complications in Scheuermann's disease are rare but serious [28, 29], thus limiting the number of publications on that topic. Disc impairment is more frequently observed in Scheuermann's disease than in normal spines [30], and thoracic disc herniation was considered as one of main types of direct compression of spinal cord in Scheuermann's disease cases further. Disc ruptures tend to occur at the apex of the thoracic kyphosis [31-33], mostly at T7-T8 or at T8-T9 levels. Even if there is no obvious kyphosis, disc ruptures used to occur at the relative apex. Any thoracic disc herniation can produce a major neurological deficit. Moreover, disc herniation can lead, whatever their size, to an ischemia of spinal cord by artery compressions. Expedited MRI of the spine should be performed in emergency for any patient presenting with Brown-Sequard syndrome or any compressive sign. In addition, many degenerative diseases accompany SD, especially atypical pattern. When multiple endplate irregularities and anterior vertebral wedging are detected in MRI of patients thought to have thoracic disc pathology, SD should be considered. However, these hallmark findings are associated with penetrating injuries to the spinal cord, compressive extramedullary spinal tumours, hematomas, herniated intervertebral discs and numerous other causes [34, 35].

Generally, characteristic neurological patterns for symptomatic thoracic disc herniation are lacking and the localization of pain induced by thoracic disc herniation is sometimes ambiguous. For these reasons, accurate diagnosis of symptomatic thoracic disc herniation has been reported to be considerably difficult [36]. These facts can lead to delay in diagnosis, which may result in progressive neurological impairments. Previous reports have shown, however, that postoperative results of acutely developing thoracic disc herniation are generally satisfactory

[37]. Therefore, appropriate diagnosis and earlier treatment based on accurate neurological examination and diagnostic imaging, such as MR imaging, can lead to excellent recovery of neurological function. Once diagnosed, prompt surgical intervention is often required.

With new generation posterior constructs, satisfactory radiographic and cosmetic results have been achieved with posterior fusion surgery [38]. The decision of performing a posterior approach was based on the following preoperative imaging findings: those changes in intervertebral disc space height in calcified herniated discs are not obvious; preoperative images of calcified herniated discs display a protruded intervertebral disc with a clear edge, whereas ossified intervertebral disc herniation would show an unclear edge. In addition, imaging studies and determining the real hardness of the calcified protruded disc by intraoperative palpation were diverting, and provided good suggestions for the surgical management of future similar cases [39]. Intraoperative findings in our case confirmed that the protruded disc was soft, and was easily removed via posterolateral approach surgery.

As for its clinical significance, currently, we should pay attention to the thoracolumbar discs in patients where radiological signs of SD are present on their spine images. Moreover, further studies should concentrate on the natural progression and risk of deterioration in patients with Scheuermann's disease. Prospective studies with longer follow-up duration and control groups, using valid outcome measures are needed to provide the surgeons with more accurate tools for treating patients with Scheuermann's disease [40]. Rigorous methodology clinical trials are essential to evaluate the efficacy of conservative interventions, especially different exercises and manual therapies and their combinations with braces. Particularly, there is no doubt that early decompression surgery should be carried out as early as possible for patients in atypical Scheuermann's disease with acute spinal myelopathy or paraplegia caused by a protruded disc. In the future, if a definite association between symptomatic TLDH and SD is established, it will be possible to offer prevention and treatment strategies against SD during the course of caring for TDH patients.

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## Disclosure of conflict of interest

None.

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