George A. Kapetanos Paraskevas T. Hantzidis Kleovoulos S. Anagnostidis John M. Kirkos

Thoracic cord compression caused by disk herniation in Scheuermann's disease

A case report and review of the literature

Received: 3 May 2005 Accepted: 30 August 2005 Published online: 19 January 2006 © Springer-Verlag 2006

G. A. Kapetanos (⋈) · P. T. Hantzidis K. S. Anagnostidis · J. M. Kirkos "Papageorgiou" General Hospital, 3rd Ortopaedic Department, Aristotle University of Thessaloniki, Thessaloniki, Greece

E-mail: kanagn@auth.gr Tel.: +30-2310-344271 Fax: +30-2310-344271 Abstract We present the case of a 14-year-old male with Scheuer-mann's disease and significant neurological deficit due to thoracic disk herniation at the apex of kyphosis. He was treated with an anterior decompression, anterior and posterior fusion in the same setting using plate, cage and a segmental instrumentation system. The patient had an excellent outcome with complete neurological recovery.

Keywords Scheuermann's disease · Thoracic disc herniation · Spinal cord compression · Acute myelopathy · Spastic paraparesis

Introduction

Scheuermann's disease (juvenile kyphosis dorsalis) is a structural kyphosis of the thoracic spine initially described by Scheuermann in 1921 [19]. It occurs commonly in adolescents (0.4–8.3% of the general population) and in most cases is characterized by minimal deformity and few clinical symptoms. Scheuermann's disease is rarely associated with neurological complications [5, 20].

The purpose of the present paper is to present the case of a patient with Scheuermann's disease and severe neurological deficit due to a thoracic disc hernia at the level of kyphos, to review the literature and compare with previously published similar cases.

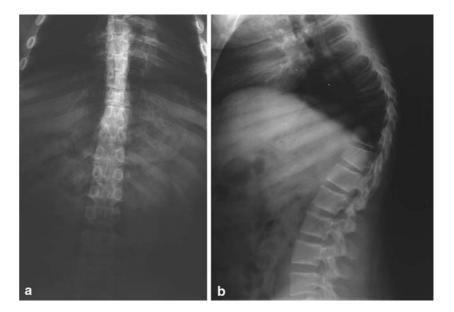
Case report

A 14-year-old male was admitted with a severe spastic paraparesis. He had gradually developed a gait disorder and bilateral numbness of lower limbs in the past 5 months. During that period his symptoms deteriorated rapidly. He was unable to walk and a week before his admission he developed spastic paraplegia. There was no history of trauma.

Clinical examination revealed spastic paraparesis below T9. The superficial sensation was diminished below the T9 dermatome. He had a muscle strength of grade II according to the Medical Research Council (MRC) scale. A barely detectable contraction could be seen below the knees by palpation over the muscles (M1 on the MRC scale). Plantar responses (Babinski sign) were present on both sides. Knee and ankle reflexes were brisk bilaterally, clonus was present in both ankles as well. Patient had hypalgesia and decreased temperature sensation. Light touch and pin prick sensation were diminished over both lower limbs and absent from the knees downward. Rectal tone and sensation while urinating were decreased.

All laboratory findings were normal including an investigation for systemic diseases. Plain radiographs of the spine in standing position revealed anterior wedging of more than 5° of several adjacent vertebrae (T7:12°, T8:14°, T9:17°, T10:15°) at the apex of the kyphosis and vertebral endplate irregularities. The thoracic kyphosis from T5 to T12 measured 66° (Fig. 1). These findings are in agreement with the criteria for diagnosis of

Fig. 1 a, b Anteroposterior and lateral radiographs of the thoracic spine in standing position, at the time of admission. The thoracic kyphosis from T5 to T12 is 66°



Scheuermann's disease. A magnetic resonance imaging scan revealed thoracic disc heniation with spinal cord compression at T8–T9 level (apex of the kyphosis). The intervertebral spaces from T6 to T10 were also very narrow and the vertebral bodies had an anterior wedging (Figs. 2, 3).

Surgical treatment was decided based on the severity of the neurological deficit. A right seventh rib transthoracic approach to the spinal column was performed followed by decompression at T8–T9 level. A disc fragment was found and removed behind the thoracic body of T9. We performed an anterior fusion using plate, screws, an

Fig. 2 a, b Magnetic resonance imaging scan revealed thoracic disc herniation with spinal cord compression at T8–T9 level (apex of the kyphosis)

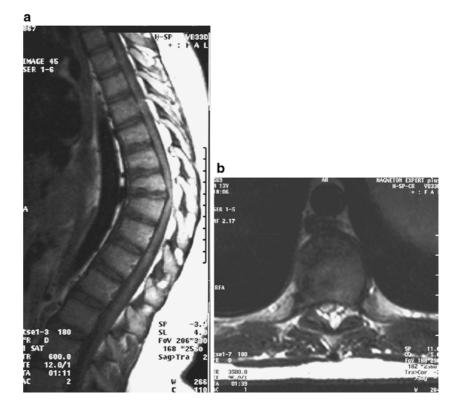




Fig. 3 Three-dimensional reconstruction computed tomography of the thoracic spine preoperatively

interbody titanium cage and also a posterior fusion from T6 to T12 using a double-rod multihook and transpedicular screws segmental instrumentation system. Bone

Fig. 4 a, b Postoperative anteroposterior and lateral radiographs of the thoracic spine. Note the anterior and posterior spinal fusion

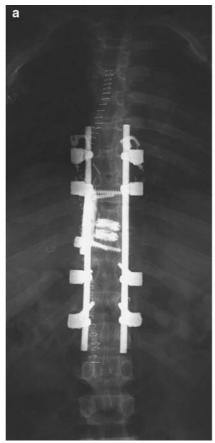
grafts were applied between T8 and T9 and intertransversaly from T7 to T10 (Fig. 4).

Postoperatively the patient had a surprisingly rapid improvement of the neurological deficit. Four weeks after the operation he could walk steadily and 2 months later his neurological examination was entirely normal.

At 2 years follow-up the patient had a normal gait, he was symptoms free and there was no increase of the spinal deformity.

Discussion

Scheuermann in 1921 [19] described a condition, which he called juvenile dorsal kyphosis, distinguishing it from the more common postural kyphosis. The etiology of Scheuermann's disease is unknown. Scheuermann proposed that the kyphosis resulted from avascular necrosis of the ring apophysis of the vertebral body [19]. Schmorl suggested that the vertebral wedging was caused by herniation of disc material into the vertebral body (Schmorl's nodes). According to Ippolito and Ponseti [10] a biochemical abnormality of the collagen and matrix of the vertebral endplate cartilage may be another





contributing factor. Bradford et al. [6] claimed that osteoporosis may be responsible for the development of Scheuermann's disease. Mechanical factors and repetitive trauma have been also considered to play a significant role in the appearance of the disease [20]. Case reports in monozygotic twins support the theory that there is also a genetic contribution [9]. In conclusion, the etiology of Scheuermann's disease remains unclear and probably is multifactorial.

The criteria for the diagnosis of Scheuermann's disease are: (1) more than 5° of wedging of at least three adjacent vertebrae at the apex of the kyphosis, (2) endplates irregularities and (3) a thoracic kyphosis of more than 45° [5]. Our patient had typical roentgenografic features of Scheuermann's disease and his kyphosis was 66° from T5 to T12.

Neurological complications in Scheuermann's disease are rare [2–4, 7, 11, 15, 18, 21, 24]. Three different types of neural compression have been reported: (1) extradural spinal cyst, (2) compression of the cord at the apex of the kyphos and (3) disk hernia at the apex of the kyphos [4]. To the best of our knowledge only 20 cases fall in the last category [2–4, 8, 12, 14, 17, 21–24]. Data of all these cases are presented in Table 1.

Despite the fact that this condition has a uniform sex distribution [15, 20], only 6 of all 20 cases were female. Males with Scheuermann's disease are at greater risk for the development of spinal cord compression in the second decade of life [1, 15]. This is in agreement with our case (male, 14 years old). This difference between the sexes is probably due to the fact that males have more longitudinal spinal growth than females and their maximum growth in trunk length, as well as the maximum deformity of the kyphosis, occurs about 2 years later.

In all cases in which the degree of deformity was reported, the average angle was only 56.3°. The severity of the neurological complications had no obvious correlation with the magnitude of the deformity [11, 18]. In our case the deformity was 66°. Lonstein et al. [13] found that in 43 cases with neurological deficits secondary to spinal deformity (what ever the etiology) the average angle of kyphosis was 95°. He suggested that the greater the angle of kyphosis, the greater the risk of neurological impairment. Neurological complications are less likely to occur when the deformity is present over a large number of segments, instead of sharply angular deformities.

Disc ruptures tend to occur at the apex of the thoracic kyphosis [13]. The majority of them have occurred at T7–T8 or at T8–T9. This is also confirmed by our case (at T8–T9).

It is interesting that a relatively small disc herniation such as in our case produces such a major neurological deficit. Factors that may influence the onset of cord compromise are the tenuous vascularization and the relatively small canal diameter of the thoracic cord [16]. Additionally, in young patients the disc material is not degenerated and can act as a solid mass. Bradford and Garcia [4] suggested that the high incidence of Schmorl's nodes in Scheuermann's disease can prevent disc herniation by decompressing the disc space. In our case there were no Schmorl's nodes either at this level or at any other level.

Surgical treatment is indicated in the cases of Scheuermann's disease with acute cord compression and generally combines anterior release with posterior instrumentation and fusion. A posterior decompression is ineffective because the compressive force is acting from the front [11, 13, 15, 18]. Although some authors have advocated posterior laminectomy and decompression, Patterson and Arbit [16] showed that 45% of patients undergoing laminectomies for thoracic disc herniations either had no relief or deteriorated.

For the 20 reported cases with thoracic disc herniation and Scheuermann's disease six patients were treated with anterior spinal fusion, four patients with laminectomy, four with posterior spinal fusion, three patients with interbody fusion and pedicular screw plate fixation and one patient was treated with discectomy without spinal fusion. Only one patient was treated conservatively with bed rest and Minerva jacket without any decompressive surgery [22]. In one case [14] the treatment was not reported. If a significant or progressive kyphosis, i.e. 55° or more exists, a combined posterior spinal fusion and instrumentation should be considered [18, 21]. Due to the age of the described patient and to the degree of kyphosis we performed discectomy, decompression, anterior and also posterior fusion using a double-rod multihook and transpedicular screws segmental instrumentation system.

The majority of the reported cases had good results. In 10 out of 20 cases the patients had complete recovery while only in one case the final outcome was paraplegia [17]. Our patient had an excellent result with complete recovery in 3 months. At the final follow-up 2 years postoperatively, he is neurologically entirely normal and the spinal fusion is solid.

This case is interesting because of the severity of the symptoms. The patient had a progressive bilateral spastic paraparesis and finally a spastic paraplegia. Spastic paraparesis was the most common symptom in all cases but neurological symptoms were not so severe to result in paraplegia. Only one case had paraplegia, as the described patient [14]. It is impressive that despite the significant neurological deficit there was a complete recovery. It seems that when symptomatic compression of the spinal cord occurs, surgery is the best option.

Table 1 Cases of Scheuermann's disease causing spinal cord compression as the result of thoracic disc herniation

References	Level	Age/se	x Findings	Appro a	ch Procedure	Results (follow up)
Muller [14] Van Landingham [23]	T10 T7–T8–T9	40 M 17 M	Paraplegia Spastic paraparesis, no sphincter dysfunction	P	Not reported T7–T9 laminectomy	Not complete recovery Complete recovery (4 months)
Roth et al. [17] Turinese and sRayen [22]	T9-T10 T5-T7-T8	61 M 16 M	Spastic paraparesis Spastic paraparesis, sphincter dysfunctio	P	T9–T10 laminectomy Bed rest, Minerva jacke	Paraplegia t Complete recovery
Bradford and Garcia [4]	T7-T8	16 M	Spastic paraparesis	P	T7-T9 laminectomy	Residual hypereflexia (120 months)
Ryan and Taylor [18]	T8-T9	18 M	Spastic paraparesis	T	T8–T10 partial vertebrectomy, anterior spinal fusion	Spastic paraparesis (36 months)
Yablon et al. [24]	T7-T8	29 M	Spastic paraparesis, sphincter dysfunctio	CT/T	T7–T8 discectomy and fusion	Complete recovery (14 months)
Lesoin et al. [12] (six cases)	T9-T10	49 F	Monoparesis (left)	P	T8-T10 laminectomy	Not complete recovery (12 months)
	T8-T9	55 F	Spastic paraparesis	T	T8-T9 discectomy	Small motor deficit (8 months)
	T11-T12	61 M	Monoparesis (right)	P	Posterior spinal fusion	Not complete recovery (10 months)
	T8-T9	27 M	Lower back pain, Babinski's sign (right)	PL	T8–T9 discectomy, T7–T10 posterior spinal fusion	Complete recovery (3 months)
	T8-T9	30 M	Lower back pain, slight motor deficit (left)	PL	T8–T9 discectomy, T8–T9 posterior spinal fusion	Complete recovery (3 months)
	T8-T9	35 M	Spastic paraparesis	PL	T8–T9 discectomy, T8–T9 posterior spinal fusion	Small motor deficit (3 months)
Bohlman and Zdeblick [3] (two cases)	T11-T12T12-L	138 F	Monoparesis. Back and leg pain (for 16 years)	CT	T11–L1 discectomy and fusion	Complete recovery (36 months)
	T8-T9	25 F	Back and leg pain (for 7 years)	T	T8–T9 discectomy and fusion	Significant pain relief (26 months)
Stambough et al. [21]	T6-T7-T8	21 M	Spastic paraparesis	T	T6–T8 discectomy, anterior spinal fusion	Complete recovery (24 months)
Bhojraj and Dandawate [2 (three cases)	2] T11–T12	16 M	Spastic paraparesis, urinary hesitancy	P	T10–T12 laminectomy, posterior interbody fusion	Complete recovery (36 months)
	T12–L1	24 M	Spastic paraparesis, urinary hesitancy	P	T12–L1 laminectomy, posterior interbody fusion	Complete recovery (24 months)
	T11–T12	16 F	Spastic paraparesis	P	T11–T12 laminectomy, posterior interbody fusion	Transient worsening postoperatively, subsequently complete recovery (15 months)
Chiu and Luk [8]	T11-L2	35 F	Spastic paraparesis	T	T11–L2 discectomy, spinal fusion	Small motor deficit (24 months)
This report (2005)	T8-T9	14 M	Rapidly progressive spastic paraparesis, finally spastic paraplegia	T	T8–T9 discertomy, anterior fusion, T6–T12 posterior fusion	Complete recovery (25 months)

M Male, F female, P posterior, T transthoracic, CT costotransversectomy, PL posterolateral

Conclusion

Thoracic disc herniation is an extremely rare complication of Scheuermann's disease, resulting in cord compression

and marked neurological deficit, most commonly in young patients. This rare form of spinal cord compression once demonstrated, should be treated as soon as possible with surgical decompression and stabilization of the spine anteroposteriorly.

References

- 1. Anderson M, Hwang SC, Green WT (1965) Growth of the normal trunk in boys and girls during the second decade of life. J Bone Joint Surg (Am) 47(8):1554–1564
- Bhojraj SY, Dandawate (1994) Progressive cord compression secondary to disc lesions in Scheuermann's kyphosis managed by posterolateral decompression, interbody fusion and pedicular fixation. Eur Spine J 3:66–69
- Bohlman HH, Zdeblick TA (1988)
 Anterior excision of herniated thoracic discs. J Bone Joint Surg (Am) 70A:1030–1047
- Bradford DS, Garsia A (1969) Neurological complications in Scheuermann's disease. A case report and review of the literature. J Bone Joint Surg (Am) 51:567–572
- Bradford DS (1974) Scheuermann's kyphosis and round back deformity. J Bone Joint Surg (Am) 56:740–758
- Bradford DS, Brown DM, Moe JH (1976) Scheuermann's kyphosis: a form of juvenile osteoporosis? Clin Orthop 118:10–15
- Bruns J, Heise U (1994) Spastische paraparese bei morbus Scheuermann. Z Orthop 132:390–393
- Chiu KY, Luk KDK (1995) Cord compression caused by multiple disc herniations and intraspinal cyst in Scheuermann's disease. Spine 20(9):1075–1079

- 9. Graat HC, van Rhijn LW, Schrander-Stumpel CT, van Ooij A (2002) Classical Scheuermann disease in male monozygotic twins: further support for the genetic etiology hypothesis. Spine 27(22):E485–E487
- Ippolito E, Ponseti I (1981) Juvenile kyphosis: histological and histochemical studies. J Bone Joint Surg (Am) 63-A:175
- 11. Klein DM, Weiss RL, Allen JE (1986) Scheuermann's dorsal kyphosis and spinal cord compression: case report. Neurosurgery 18:628–631
- Lesoin F, Leys D, Rousseaux M, Dubois F, Villete L, Pruvo JP, Petit H, Jomin M (1987) Thoracic disk herniation and Scheuermann's disease. Eur Neurol 26:145–152
- Lonstein JE, Winter RB, Moe JH, Bradford DS, Chou SN, PintoWC (1980) Neurologic deficits secondary to spinal deformity. A review of the literature and report of 43 cases. Spine 5(4):331–355
- Muller R (1951) Protrusion of thoracic intervertebral discs with compression of the spinal cord. Acta Med Scand 139:99–104
- Normelli H, Svensson O, Aaro S (1991) Cord compression in Scheuermann's kyphosis-a case report. Acta Orthop Scand 62:70–72
- Patterson RH Jr, Arbit E (1978) A surgical approach through the pedicle to protruded thoracic disc. J Neurosurg 48:760–764

- Roth G, Lambert H, Chu CK (1965)
 Hernie discale thoracique double et
 maladie Scheuermann. Rev Med Suisse
 Romande 85:296
- Ryan MD, Taylor TKF (1982) Acute spinal cord compression in Scheuermann's disease. J Bone Joint Surg (Br) 64(4):409–412
- Scheuermann H (1921) Kyphosis dorsalis juvenilis. Z Orthop Chir 41:305–317
- Sorensen KH (1964) Scheuermann's juvenile kyphosis clinical appearances, radiography, etiology and prognosis. Munksgaard, Copenhagen, pp 11–222
- Stambough JL, VanLoveren HR, Cheeks ML (1992) Spinal cord compression in Scheuermann's kyphosis: case report. Neurosurgery 30(1):127–130
- Turinese A, Raven C (1966) Complicance neurologicere nella osteochondrite vertebral jiuvanile. Psichiat E Neuropatia Giom 23:215–238
- Van Landingam JH (1954) Herniation of thoracic intervertebral discs with spinal cord compression in kyphosis dorsalis juvenilis (Scheuermann's disease), case report. J Neurosurg 11:327– 329
- 24. Yablon JS, Kasdon DL, Levine H (1988) Thoracic cord compression in Scheuermann's disease. Spine 13(8):896–898