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Thoracic scoliosis and restricted neck motion: a new syndrome? A report of six cases*

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Abstract The cases of six adolescent males with a mild thoracic scoliosis of unknown etiology and a marked limitation of neck flexion are reported. All six complained of mild thoracic spine pain and were 13–17 years old at the onset of their symptoms. In addition to the thoracic scoliosis, all were found to have a marked limitation of neck flexion. They were unable to touch their chest wall with their chin. None had a positive family history of scoliosis. Radiological examination revealed a thoracic scoliosis of 10°–40°, no abnormalities were detected on cervical

spine plain radiographs. Extensive investigations, including MRI of the entire spine, failed to disclose the cause of the spinal deformity or the mechanism causing the limitation of neck motion. Apart from the limited cervical flexion and mild pain, this condition was found to be benign and the spine deformity to be mild or moderate in nature and with limited progression. Review of the literature failed to identify similar cases.

Key words Scoliosis · Restricted neck motion

Introduction

Limitation of neck motion, usually a significant physical finding, may arise from a variety of traumatic, inflammatory, congenital, or neoplastic processes that may involve ligaments, discs, muscles, bone, or the nervous system. When limited neck motion is associated with scoliosis the most likely diagnosis is Klippel-Feil syndrome [5]. In this syndrome a short neck with marked motion limitation results from congenital cervical interbody fusion(s). The cervical abnormalities are associated with dorsal scoliosis or kyphosis.

Recently, six adolescent males presented with a mild to moderate thoracic scoliosis of unknown etiology associated with a marked limitation of neck flexion. Extensive investigations, including MRI examination of the entire spine, failed to disclose the cause of the spinal deformity or the mechanism causing the limited neck motion. Review of the literature failed to identify similar cases.

Case reports

The six reported patients presented over a 3-year period at a scoliosis clinic with an annual patient population of about 800, of which about 20 undergo corrective surgery. All six patients were adolescent males aged 13–17 years at the onset of symptoms (Table 1). All six complained of mild thoracic spine pain and presented with mild to moderate thoracic scoliosis. A negative family history of similar manifestations was obtained in all six patients. Five adolescents had a right thoracic curve and one a left thoracic curve. All had mild to moderate rib humps. Despite the small or moderate curve magnitude, these curves exhibited limited correction on side bending maneuvers. In addition, a marked restriction of cervical spine flexion was detected in all six patients. While neck extension was found to be normal, neck flexion, measured with a goniometer, was found to be markedly restricted (35°–45°). The marked flexion restriction in these patients was manifested by an inability to touch their chest wall with their chin (Fig. 1).

In some, the attempt at neck flexion was associated with dorsal and chest wall pain. A complete neurologic examination was unremarkable and no other significant physical findings were detected.

Plain radiographs revealed thoracic scoliosis of 10°–40° measured by the Cobb method (Table 1). All had a grade 1 Nash-Moe

Table 1 Clinical data of the six patients

Case no.	Age at onset (years)	Length of follow-up (years)	Thoracic curve	Limited neck flexion	Other symptoms
1	15	6	T1–T6 15°	35°	Neck pain
2	15	5	T4–T10 10°	45°	Thoracic pain
3	14	6	T4–T10 14°	40°	Thoracic pain
4	13	4	T5–T10 18°	35°	Chest wall and back pain
5	14	3	T4–T10 32°	40°	Thoracic pain
6	17	4	T6–T11 40°	35°	Thoracic pain

Fig. 1 A–C Case 1: a 17-year-old male complaining of neck pain. **A** Clinical photograph (taken at age 17). Note the marked limited neck flexion of 35°. **B** Standing anteroposterior plain radiograph of the thoracic spine. Note the mild right thoracic curve of 10° (T1–T6). **C** Lateral plain radiograph of the cervical spine with no detectable abnormalities

Fig. 2 Case 2: a 17-year-old male with thoracic spine pain and limited neck flexion. Standing anteroposterior plain radiograph of the thoracic spine. Note the mild thoracic scoliosis of 15° (T4–T10). A grade 1 Nash-Moe rotation is present

vertebral rotation. In four patients the sagittal contour was normal while two had a “lordoscoliotic” deformity. There were no detectable bony anomalies (Figs. 1, 2). Specifically, congenital scoliosis or neurofibromatosis were excluded. Two patients also had a compensatory lumbar curve (judged to be compensatory by supine side-bending views as well as the lack of vertebral rotation). Cervical spine plain radiographs were found to be normal (Fig. 1). CT scans of the cervical spine appeared normal, including the shape and volume of the posterior neck muscles. Technetium bone scan was normal as well. MRI examination of the entire spinal column did not disclose any abnormalities except for mild thoracic scoliosis. There was no evidence for Chiari malformation or cord tethering. In three patients electromyographic studies of the neck and upper thoracic musculature were within normal limits. In two patients blood creatine phosphokinase and aldolase were checked and found to be normal.

The cause of the scoliosis or the limited neck flexion could therefore not be determined. There was no change in the patients’ symptoms during subsequent follow-up visits (3–6 years). In five out of the six individuals the scoliosis was found to be stable. In one patient (case 5), mild curve progression was halted by bracing. All six patients have currently finished their skeletal growth.

Discussion

The present report describes the association between mild thoracic scoliosis and marked limitation of neck flexion in six adolescent males. Despite the fact that the etiology of the spinal deformity or the associated neck flexion limitation were not elucidated, the clinical presentation in these adolescents was by no means typical for idiopathic scoliosis. The spinal deformity was mild and stable in five of the six cases. One patient was braced with subsequent stabilization of the curve. The most prominent feature in the six reported patients was the thoracic or neck pain associated with the limited neck flexion. While the normal neck flexion of adolescent males is around 65° [13], all six reported patients had a limited neck flexion of between 35° and 40° only. They were unable to touch their chest wall with their chin.

A variety of pathological processes could give rise to restricted cervical spine motion. The first condition on the differential diagnosis list reminiscent of the six cases described is Klippel-Feil syndrome. The most obvious physical findings of patients with Klippel-Feil syndrome are a short neck and limited cervical range of motion, accompanied by a scoliotic deformity [4]. A Sprengel’s deformity can also be found in Klippel-Feil syndrome [4]. Occipitocervical synostosis may also mimic a clinical presentation similar to our patients’, including a spinal deformity.

mity accompanied by limited neck mobility [1]. No evidence for congenital bony anomalies in the cervical, thoracic, or lumbar spine was found in any of the six patients.

Scoliosis and limited neck motion can also occur in patients with neurofibromatosis. Yong-Hing and associates [12] found cervical spine abnormalities in 17 of 56 patients with neurofibromatosis. Of the 34 patients with scoliosis secondary to neurofibromatosis, 44% had a variety of cervical spine abnormalities, including limited and painful neck motion [12]. These cervical spine abnormalities were most commonly found in individuals with thoracic scoliosis [12]. None of the six patients in the present report had evidence of neurofibromatosis. Likewise, none of them had evidence of spinal canal or spinal cord anomalies, such as syringomyelia. Indeed, syringomyelia can give rise to neck pain and thoracic scoliosis [11]. Space-occupying lesions of the central nervous system or the spinal cord can also lead to restricted cervical range of motion and scoliosis [10]. Such abnormalities were not found in any of the six reported individuals.

There was also no evidence in these six patients of a rheumatic disease. Juvenile rheumatoid arthritis is known frequently to involve the cervical spine, giving rise to a stiff neck in its initial stages [5].

A myopathy or muscular dystrophy of some kind [8, 9] could also explain the association of scoliosis and limited cervical range of motion found in the six reported patients. For example, Emery-Dreifuss muscular dystrophy can give rise to a variety of orthopedic deformities, including scoliosis and restricted neck flexion [8]. Except for the spine deformity and limited neck flexion, none of the patients had clinical or laboratory evidence of a muscular disease. DeMuth and colleagues [3] reported on two patients with

extension contracture of the cervical spine, limiting neck flexion. This was found to be caused by a tight ligamentum nuchae and was not accompanied by a spinal deformity.

A perturbation in the spinal cord biomechanics could give rise to scoliosis and limited neck flexion. Breig [2] found that the cervical and upper thoracic portions of the spinal cord elongate during spinal flexion. In addition, the cervical and upper thoracic cord moves longitudinally relative to the corresponding vertebrae, by up to 18 mm [7]. It is therefore possible that the six reported patients may have suffered local or distant cord tethering. This could explain the occurrence of scoliosis as well as the limited and painful cervical spine flexion. This was not corroborated, however, in the patient population, as the MRI examination of the entire spine revealed no abnormalities. Thus, gross soft tissue, bony, or cord abnormalities were ruled out in the six reported patients.

Conclusions

It is concluded that the six patients described in this paper represent an unreported genuine clinical entity of unknown etiology. All six were adolescent males with mild thoracic scoliosis and a striking limitation of neck flexion accompanied by mild thoracic spine or neck pain. No other anomalies were detected. Apart from the limited neck flexion and mild neck or upper thoracic spine pain, this condition seems to be benign, and the spinal deformity is small and usually non-progressive.

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