The case:

A 17-year-old adolescent boy presented with progressive lower back pain and fatigue.

Figure: Upright lateral (A) and anteroposterior (B) radiographs of the thoracic spine.

Your diagnosis?

For answer see page 66
A 17-year-old adolescent boy presented with progressive thoracolumbar back pain, fatigue, and a noticeable thoracic kyphosis. He had no history of antecedent trauma. On physical examination, a rigid thoracic angulation with level shoulders and pelvis and point tenderness over the T8 to T12 vertebrae were detected. There was evidence of tight hamstring musculature, but no sign of scoliosis or neurological deficits. The initial upright lateral radiograph showed loss of anterior vertebral body height from T7 to T11 resulting in 88° of kyphosis (Figure 1A). There was associated intervertebral disk narrowing and minor end-plate irregularities and flattening, without scoliosis, on the anteroposterior radiograph (Figure 1B). No fractures or bone destruction were present, and the paraspinal soft tissues were unremarkable. This constellation of findings is consistent with Scheuermann’s disease.

Scheuermann’s disease was first described in 1921 as a unique cause of juvenile “roundback” resulting from pathological changes in the thoracic vertebral endplates. This novel description explained the rigid kyphosis seen clinically, dismissing previously held views suspecting weak paraspinal musculature and posture as the causative factor. Epidemiological studies have shown Scheuermann’s disease to be the most common structural cause of kyphosis in the adolescent population, with prevalence rates ranging from 0.4% to 10% depending on the stringency of the inclusion criteria used.¹³ There are conflicting reports of gender disparity, with no consistent predilection found.¹⁴ Although the underlying etiology of Scheuermann’s disease continues to elude physicians nearly a century later, a prompt diagnosis can be confidently made based on thoracic radiographs alone.
potentially avoiding unnecessary studies or procedures.

**Scheuermann’s Disease Terminology**

The term “Scheuermann’s disease” is often used indiscriminately to convey the radiological finding of excessive spinal kyphosis, regardless of the level of the deformity. True Scheuermann’s disease should be distinguished from the variant known as lumbar Scheuermann’s disease (also known as juvenile disk disorder or atypical Scheuermann’s disease), which includes lumbar spine involvement. Such precision is necessary for prognostic and treatment implications.

**Normal Anatomy and Development of the Thoracic Vertebrae**

The vertebral column transmits weight from the cranium and thorax onto the pelvis, absorbs ground forces, and allows for rotational and translational movement while protecting the neural elements. The basic structure of the vertebral column is established in utero as the paraxial mesoderm adjacent to the neural tube undergoes segmentation and forms the 2 primary kyphotic curves in the thoracic and sacral spine. The thoracic curve continues to undergo rapid growth after birth until 3 to 4 years of age and again during the prepubertal growth spurt between ages 13 and 16. Although the degree of normal thoracic kyphosis varies considerably throughout the population, on average, the angulation increases from 20° in toddlers to 40° by adulthood. With large growth contribution in the adolescent, the risk of spinal deformity progression in Scheuermann’s disease is primarily dependent on the starting degree of deformity and the estimated growth potential remaining.

The thoracic vertebrae are formed through endochondral ossification at 3 primary ossification sites forming the vertebral body and each neural arch. These ossification centers are united by 3 cartilaginous unions, each fusing by 6 to 8 years of age. The body itself is composed of a central core of trabecular bone surrounded by thick compact bone. This outer shell interfaces with hyaline cartilage at the inferior and superior surface of the vertebral body, together forming the vertebral end plates. Functionally, the vertebral end plate attaches to the adjacent intervertebral disk via lamellae of the inner annulus fibrosis, preventing herniation of the nucleus pulposus into the vertebral body and accommodating mechanical loading of the spine. In the developing spine, small blood vessels penetrate the vertebral end plate, providing nutrients for the growing intervertebral disks, which are otherwise nearly devoid of blood supply. Some have theorized that as these blood vessels close throughout development, they leave behind vascular grooves that persist as focal areas of potential weakness and predispose the end plate to mechanical failure. Indeed, recent evidence suggests that vertebral end-plate irregularities can be found in nearly 25% of the general population as a normal variant.

It appears that changes in the vertebral end-plate integrity accompany the development of Scheuermann’s disease, but the definitive pathogenesis remains unclear. Most theories have suggested underlying mechanical and genetic influences to account for the anterior vertebral wedging and end-plate irregularities. In favor of a mechanical mechanism, Scheuermann’s disease is associated with increased load on the spine secondary to increased general body height and weight, and decreased sternal length. Mechanical failure of the end plate was originally proposed because herniations of the nucleus pulposus into the vertebral body, known as Schmorl’s nodes, are frequently present in Scheuermann’s disease. However, this theory fails to explain the occurrence of Schmorl’s nodes at nonwedged levels of the spine in addition to the occurrence of Schmorl’s nodes in individuals who do not have Scheuermann’s disease. Vertebral wedging due to osteoporosis resulting in compression fractures has also been suggested. However, dual-energy x-ray absorptiometry scans have demonstrated normal bone mineralization in patients with Scheuermann’s disease. Biochemical changes in the end-plate cartilage, including alterations in proteoglycan proportions, collagen, and cellular and matrix integrity likely play a role in vertebral wedging by promoting vertebral incompetence and possibly asymmetrical growth. There have been numerous investigations into a genetic cause of Scheuermann’s disease, with familial clustering indicating an autosomal dominant inheritance. However, recently Damborg et al. found a multifactorial inheritance pattern using twin studies.

**Clinical Presentation**

Scheuermann’s disease presents during the prepubertal growth spurt with a rigid thoracic kyphosis. Patients seek care for either cosmetic reasons or back pain (which is commonly exacerbated by prolonged sitting, standing, or strenuous exercise). On examination, the apex of the deformity is usually localized to the T7-T9 vertebrae or the thoracolumbar junction and classically persists with hyperextension of the spine. There is often a compensatory lumbar and cervical hyperlordosis to maintain sagittal balance with secondarily, increased muscular strain (ham-
string, iliopsoas, and anterior shoulder girdle). There may or may not be reproducible pain with palpation. Approximately one-third of patients have coexisting scoliosis, which tends to be mild and usually self-limiting. Patients rarely present with severe secondary manifestations, including spastic paraparesis from myelopathy (secondary to spinal cord compression from severe deformity or disk herniation) or restrictive cardiopulmonary disease. Unfortunately, failure to differentiate Scheuermann’s disease from poor posture often delays referral and appropriate management.

**IMAGING**

**Radiography**

The diagnosis of Scheuermann’s disease must be confirmed with appropriate imaging studies. Erect anteroposterior and lateral thoracic radiographs are sufficient to make the diagnosis and should be obtained with the arms flexed to 90° (Figure 1). A lateral radiograph obtained in hyperextension with a wedge at the apex of the deformity is helpful to rule out postural kyphosis, which allows for correction with extension. Lateral radiographs are used to quantitate the sagittal plane deformity by calculating a Cobb angle, which is measured at the intersection of lines drawn parallel to the vertebral bodies at the most superior and inferior extent of the curve. Although there is little agreement on the exact radiologic criteria necessary to diagnose Scheuermann’s disease, the core features of the criteria proposed by Sorensen in 1964 are commonly used. Anterior wedging greater than 5° in 3 or more adjacent thoracic vertebral bodies in an adolescent spine must be demonstrated, often accompanied by vertebral end-plate flattening and irregularity, narrowing of the intervertebral disk spaces, and thickening of the anterior longitudinal ligament. Thoracic kyphosis with a Cobb angle greater than 40° and the presence of Schmorl’s nodes (Figure 2) are also suggestive of, but not specific to, Scheuermann’s disease. Likewise, compensatory hyperlordosis of the cervical and of the lumbar spine are common but nonobligate features. Recommendations to decrease the stringency of the criteria used for diagnosis result in earlier detection of Scheuermann’s disease, but should be applied with caution, given recent findings of vertebral end-plate irregularities, Schmorl’s nodes, and anterior vertebral wedging in the normal adolescent population.

**Computed Tomography**

Computed tomography (CT) of the spine can clearly demonstrate the vertebral abnormalities of Scheuermann’s disease, and is indicated when the diagnosis is unclear on thoracic radiographs. Loss of anterior vertebral height, vertebral end-plate irregularities, and Schmorl’s nodes are easily visualized on CT (Figure 3). Computed tomography is also particularly useful to differentiate between Scheuermann’s disease and other causes of thoracic kyphosis, such as vertebral fractures, congenital kyphosis, and scoliosis. Computed tomography may also be used for surgical planning. However, careful consideration should be given to the value of additional information gained by a CT scan because of the significantly increased radiation exposure, especially for younger patients.

**Magnetic Resonance Imaging**

Magnetic resonance imaging (MRI) is unnecessary for the initial diagnosis of Scheuermann’s disease, but can be performed if there are clinical signs of thoracic disk herniation with spinal cord compression, or for preoperative assessment and surgical planning. Compared with radiographs, MRI is exquisitely sensitive for defining the extent of spinal cord impingement as well as detailing extradural meningeal cysts, which appear to be more common in the region of the kyphotic deformity and may precede the development of Scheuermann’s disease in certain patients. Loss of intervertebral disk height can be easily visualized (Figure 4). Magnetic resonance imaging can also evaluate for tumors, vertebral infections, and disk abnormalities masquerading as Scheuermann’s disease when the diagnosis is unclear.

**DIFFERENTIAL DIAGNOSIS**

The proper identification of Scheuermann’s disease is critical because the prognostic and treatment implications can vary significantly when compared with other causes of thoracic hyperkyphosis. The differential diagnosis of Scheuermann’s disease is broad and includes postural kyphosis, congenital scoliosis, congenital kyphosis, skeletal dysplasia, and kyphosis secondary to fractures, endocrinopathies, tumors, and vertebral infections. Postural kyphosis is a common cause of thoracic kyphosis in the adolescent population, classically presenting with a mild deformity that resolves on hyperextension in contrast to the rigid angulation seen in Scheuermann’s disease. Further, in postural kyphosis, vertebral apophyseal lines are smooth and no vertebral wedging is seen (Figure 5). In congenital scoliosis, thoracic hyperkyphosis may coexist with the coronal plane convexity (Figure 6). The findings of Scheuermann’s disease with mild scoliosis may overlap...
with congenital scoliosis, but Scheuermann’s disease can be easily differentiated on radiographs, having characteristic anterior vertebral body collapse and end-plate changes. Congenital kyphosis type II, also known as failure of segmentation, tends to worsen with vertebral column growth, similar to Scheuermann’s disease. However, congenital kyphosis presents at an earlier age than Scheuermann’s disease (usually before age 10), and radiographs show fusion of the anterior vertebral processes and loss of disk space. Skeletal dysplasias, including spondyloepiphyseal dysplasia congenita, and Morquio syndrome are rare causes of thoracic kyphosis that present with a myriad of clinical features absent in Scheuermann’s disease. Acute compression fractures secondary to trauma or Cushing’s disease-induced osteoporosis can cause kyphotic deformities as well and should be ruled out by thoracic radiography or CT, showing vertebral collapse in the absence of anterior wedging of consecutive vertebral bodies in these scenarios (Figure 7). Spinal tumors and vertebral infections should also be included in the differential diagnosis, although they can often be excluded based on clinical presentation and MRI findings if necessary. Finally, aggressive laminectomy or radiation treatment to the spine prior to skeletal maturity are important iatrogenic causes of thoracic kyphosis that must be considered. Clinical history and imaging findings in these cases would be discrepant to the information obtained from the patient.

**TREATMENT**

Due to a paucity of data on natural history and long-term outcomes with treatment, the management of Scheuermann’s disease remains controversial. It is generally assumed that the majority of cases are benign, with resolution of symptoms on skeletal maturity. However, it appears that patients with a kyphosis greater than 75° are more likely to show progression of the deformity, degenerative spondylosis, disk herniation, and back pain as adults.²,¹⁰ Murray et al²⁰ followed 67 patients with Scheuermann’s disease for an average of 32 years, noting more severe back pain compared with their age-matched controls but no associated disability, interference with daily living, or cosmetic dissatisfaction. Conversely, Ristolainen et al²¹ found higher rates of back pain and decreased ability to perform
activities of daily living in 49 patients with Scheuermann’s disease at 37-year follow-up.

In general, the treatment of Scheuermann’s disease depends on the patient’s age, degree of angulation, and estimated remaining growth. Management should focus on decreasing symptoms and preventing progressive deformity and the associated morbidity. For mild kyphosis less than 50 to 55°, treatment is mainly supportive. Analgesics and physical therapy are generally sufficient to manage pain and limit fatigue. A combination approach to strengthen the paraspinal musculature and increase hamstring and iliopsoas flexibility provides symptomatic relief, although physical therapy has no role in correcting the underlying deformity. Serial radiographs should also be obtained to monitor disease progression until skeletal maturity. Once the kyphosis surpasses 55° or patients have failed conservative treatment, bracing with a thoracolumbosacral orthosis or a Milwaukee brace has been recommended for patients with residual growth remaining. Bracing should continue until passive correction is achieved and reversal of anterior wedging is observed. Although correction is often partially lost on cessation of brace use, an initial correction of at least 15° correlates with a favorable prognosis.

Surgical treatment is rarely necessary and only indicated for a severe kyphosis greater than 70° and debilitating pain or for patients with neurological deficits. Thoracotomy with anterior release and fusion of the spine followed by posterior fusion and stabilization has previously been the preferred surgical technique. However, the best method to achieve and maintain correction while minimizing complications is highly debated. Given improved 3-column fixation with pedicle screws, posterior-only fixation has been shown to provide good correction with stable construct until healing while limiting significant morbidity associated with anterior and posterior surgery combined for deformity up to 104°.

CONCLUSION

Scheuermann’s disease is a common cause of adolescent thoracic kyphosis remarkable for a generally self-limiting course. Standing anteroposterior and lateral radiographs showing anterior wedging of consecutive thoracic vertebrae are the gold standard for diagnosis. They should be the first step to differentiate other causes of excessive kyphosis that require more aggressive treatment. For lesser degrees of kyphosis, conservative treatment with physical therapy and bracing is effective in limiting pain and fatigue until skeletal maturity is reached. In more severe cases, surgery with spinal fusion is indicated to improve the deformity and symptoms.

REFERENCES

14. Summers BN, Singh JP, Manns


2015 Continuing Medical Education Courses

Ways to Register

Online: foreonline.org
Phone: 813-910-3652

#CSSETampa #CSFA Tampa #PushingEnTrauma #ATLTrauma #CSOTFlorida