

Thoracic disc herniation in spondyloepiphyseal dysplasia

A report on two cases

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We present two cases of spondyloepiphyseal dysplasia, complicated by thoracic disc herniation.

Two Japanese patients were diagnosed as having a tardy form of spondyloepiphyseal dysplasia, because they were short-trunk dwarfs with normal facies, manifested during childhood, and had generalized platyspondylitis and dysplasia of the proximal epiphyses (Langer 1964). Laboratory examinations were normal, including urinary mucopolysaccharides and serologic markers for collagen disease.

Case 1

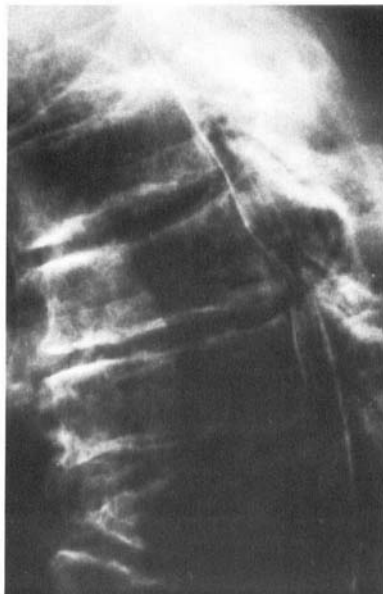
A 50-year-old woman was referred with paraparesis

and urinary incontinence. The patient had one month's history of back pain until, one month before admission, she suddenly became unable to walk and pass water. Her family history was unremarkable. There was no history of trauma. She had been using crutches since her 20s because of pain and limitation of movement in both hips.

She had a mild thoracic kyphosis with decreased movement of the cervical and lumbar spine and tenderness between the T4–5 spinous processes. Motion was limited in the large joints and the metacarpophalangeal joints. There was a spastic paraparesis and loss of sensation below the T7 level, with left sacral sparing. Radiologic examinations, including myelography and MRI, showed a thoracic disc herniation at the T4–5 level.

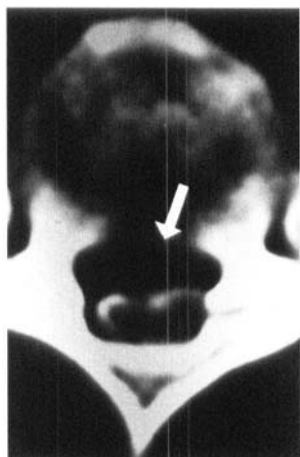
Case 1.

Myelography (left) and MRI (right) showing generalized platyspondylitis and herniation of nucleus pulposus at the T4–5 level (black arrows).



Case 2.

Myelogram CT showing herniation of nucleus pulposus (white arrow) at the T2–3 level.



A T3–5 laminectomy was performed 4 months after the onset of the neurologic deficits, but the patient did not improve. 10 weeks later, an anterior decompression and fusion of the T4–5 and T5–6 disc spaces were performed. 10 months postoperatively, the patient's strength had improved; she was able to walk with the assistance of a walker, but there was no resolution of her bowel and bladder incontinence.

Case 2

A 19-year-old man was admitted with gait disturbance and urinary incontinence of 2 weeks' duration. He had an episode of sudden back pain 1 week before the onset of the symptoms. There was no history of trauma, but he had a history of bilateral coxalgia since the age of 13. His father had the same type of skeletal dysplasia.

He had a mild thoracic kyphosis with tenderness over the spinous processes of the upper thoracic spine. Movement of hip joints was mildly reduced. There was a spastic paraparesis with a positive Babinski's sign. He had decreased sensation below the T4 level, and loss of sensation below the T12 level bilaterally. Radiographic examination showed a herniated intervertebral disc at the T2–3 level.

Using the extradural approach through a right T2–3 wide hemilaminectomy, the nucleus pulposus was removed 3 weeks after the onset of symptoms. Immediately after the operation, the neurologic deficit showed marked resolution, followed by full recovery within 5 weeks.

Discussion

Spondyloepiphyseal dysplasia may cause predisposition to thoracic disc herniation: severe degenerative changes of the spine are a feature of this condition, especially of the thoracic vertebrae (Langer 1964, Carter and Sutcliffe 1970). Recent reports have demonstrated a genetic abnormality in type II collagen, which is the main constituent of the cartilage and the nucleus pulposus (Lee et al. 1989, Tiller et al. 1990).

The optimal method of treating thoracic disc herniation remains controversial. Disappointing results of laminectomy have been reported (Arseni and Nash 1960, Perot and Munro 1969), but removal of the protruded disc using an anterior (Crafoord et al. 1958) or a posterolateral (Carson et al. 1971) approach has been recommended with good results.

Thoracic disc herniation is difficult to detect early, in spite of the need for early decompression of the spinal cord. Preexistent deformities and contractures due to skeletal dysplasia preclude careful monitoring of the neurologic examination, making the diagnosis even more difficult and reducing the possibility of neurologic recovery. The orthopedic surgeon should be aware of this possible complication in patients with spondyloepiphyseal dysplasia.

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