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Instrumentation for correction and fixation of scoliosis in fibrous dysplasia of the thoracolumbar spine

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Abstract A case report is presented in which correction of a thoracolumbar scoliosis, due to extensive fibrous dysplasia, is achieved using segmental spinal instrumentation with a 6-year follow-up. Pseudoarthrosis developed due to insufficient hook fixation at the sacrum.

Key words Fibrous dysplasia · Scoliosis

Introduction

Fibrous dysplasia is a benign condition in which bone is replaced by fibro-osseous tissue [5]. The presence of multiple lesions of fibrous dysplasia in the thoracolumbar region of the spine is uncommon [1]. A case is presented in which stabilisation and fixation of a scoliosis, due to extensive fibrous dysplastic involvement of the thoracolumbar spine, is achieved.

Case report

A 12.6-year-old boy was referred to our hospital suffering daily from a pain in the spinal region after a blow in a karate contest some weeks earlier. Before this incident he had been without complaints. At examination there was a severe scoliosis and the patient had a patch of hyperpigmented skin on his back. There were no neurologic deficits. No haematological, biochemical or endocrinologic abnormalities were present.

Roentgenograms revealed a recent fracture of two vertebrae (T12 and L1). Multiple lucent expansile lesions and deformed vertebrae of the thoracolumbar spine resulting in a scoliosis were vis-

ible (Fig. 1). A radionuclide scan (technetium 99m) demonstrated focal lesions of C2, C5 and C6 and extensive lesions of the thoracic and lumbar spine, left sacroiliac joint and ribs. CT with 3-D reconstruction revealed massive destruction of parts of vertebrae at multiple levels of the thoracolumbar spine and sacrum (Fig. 2).

An open biopsy was performed and confirmed histologically the suspected diagnosis of fibrous dysplasia.

At first the patient was treated in an orthosis. After a short period his pain became aggravated and a progression of the scoliosis developed. Therefore, at the age of 14.6 years a posterior spinal arthrodesis was performed using Cotrel-Dubousset instrumentation from the fifth thoracic vertebra to the sacrum for correction and stabilisation. Hooks were fixed only on laminae or pedicles that appeared normal (Fig. 3). Unfortunately, no fixation at L5 was possible because of involvement and partial collapse of the laminae. This left insufficient bone quality and no space for hook fixation at L5.

Corticocancellous autografts from the ilium were used. The ilium wings were not themselves involved in the process of fibrous dysplasia. Multiple biopsies at multiple levels were taken during operation. All confirmed histologically the diagnosis of fibrous dysplasia. Postoperatively, a plaster cast was worn for 3 months. Thereafter, a thoracolumbar orthosis was worn for 6 months.

At follow-up the distal part of both rods detached 1 month after surgery; however, he had no complaints. A pseudoarthrosis developed in the low lumbar region, for which a second bone graft

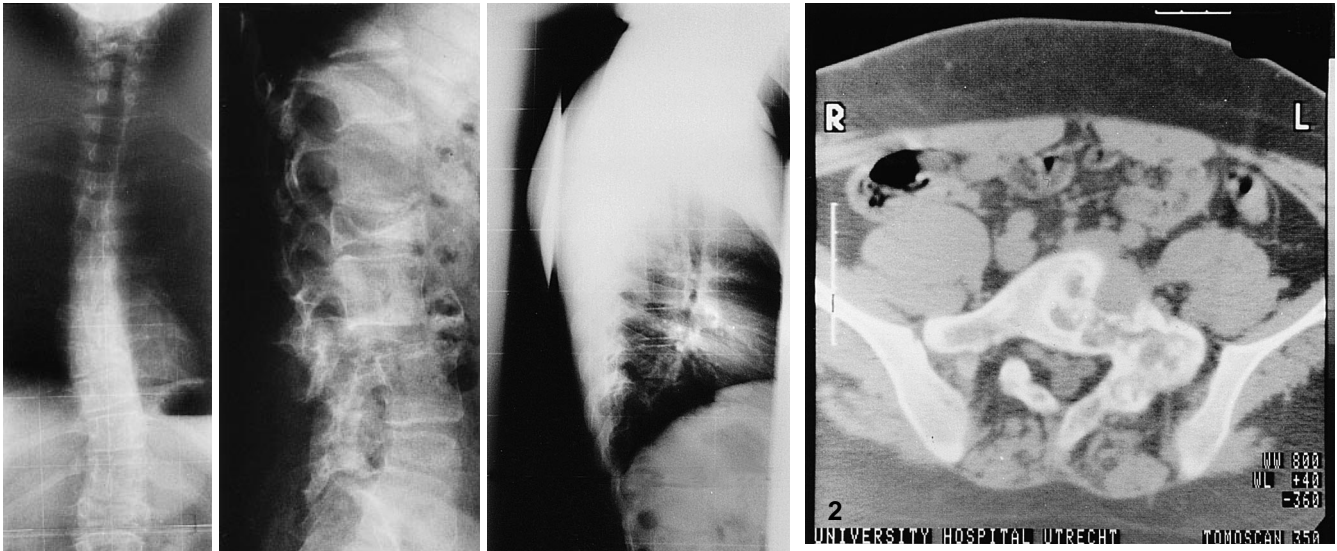


Fig. 1 (May 1990.) Preoperative roentgenograms (A antero-posterior, B, C lateral) demonstrate fractures at multiple levels. The Cobb angle from T4 to T10 measured 26° and from T10 to L1, 15°

Fig. 2 (May 1990.) The CT scan of S1 shows lucent lesions predominant at the left side

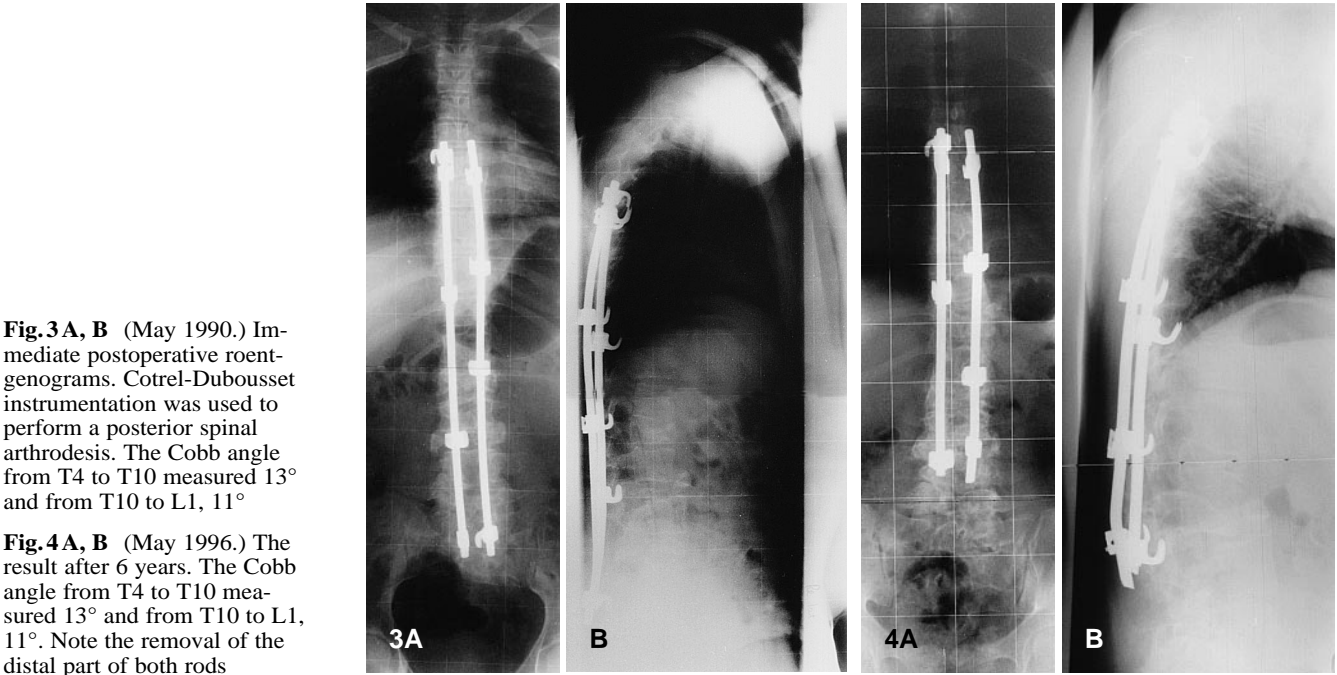


Fig. 3 A, B (May 1990.) Immediate postoperative roentgenograms. Cotrel-Dubousset instrumentation was used to perform a posterior spinal arthrodesis. The Cobb angle from T4 to T10 measured 13° and from T10 to L1, 11°

Fig. 4 A, B (May 1996.) The result after 6 years. The Cobb angle from T4 to T10 measured 13° and from T10 to L1, 11°. Note the removal of the distal part of both rods

was performed 2 years later and the distal part of both rods were removed at the same time. A supplementary hook was placed at the lamina of L2, left, followed by a second cast period for 3 months.

At follow-up 4 years after his last operation, roentgenograms showed a solid fusion mass (Fig. 4). The patient is doing well and is now working in a sheltered workshop.

Discussion

Incidence of involvement of the spine in polyostotic fibrous dysplasia ranges from 7% in the cervical spine to 14% in the lumbar spine [3]. The focal lesions of the cervical spine of this patient are small and not painful, and for this reason not operated upon. These lesions are regularly screened for expansion. The thoracolumbar scoliosis developed after multiple pathological fractures and resulted in a painful and cosmetically deforming spine. Several reports exist describing spinal fusion with instrumentation in the monostotic form of fibrous dysplasia of the spine [6]. Resection of the pathologic mass and solid fixation at adjacent vertebrae is possible in these cases. A literature search revealed one report of correction and fixation of kyphoscoliosis arising from polyostotic fibrous dysplasia of the spine [7]. Shikata et al. performed in a similar case an anterior release and a posterior fixation with Luque segmental spinal instrumentation [7]. Bone grafts were placed anterior and posterior. However, they stated that the laminae "were not weakened and posed no problem for laminar wiring". At 1-year follow-up bony union was established. Guille [2] reported on three patients with scoliosis from polyostotic fibrous dysplasia. One was treated non-operatively, and the other two underwent a posterior spinal fusion without correction of the kyphoscoliosis. No instrumentation was used because of the fear of cutting through

the hooks or wires, despite the preoperative plan to use Harrington rod instrumentation in one.

In our patient, due to the extensive involvement of the spine, fixation at multiple levels was indicated, for which CD-instrumentation was chosen. By selecting accurately laminae and pedicles that appeared normal a secure correction of the scoliosis was achieved. The planned fixation of the distal part of the rods seemed to be insufficient at L5. Due to our bad experience with the Galveston method, and the poor quality of the left side of S1, the decision was made during the operation to fix the distal part with hooks on S1 [4]. In conjunction with the prolonged conservative after-treatment it was thought to be sufficient. Sacroiliac screws or sacral pedicle screws were not yet available at our hospital at that time.

Despite the second intervention we think we managed this case correctly.

This case report shows that, despite serious involvement of the whole spine including posterior elements, surgical correction is possible with segmental spinal instrumentation by careful preoperative planning, with a lasting result. Fixation of the sacral hooks at an involved part of the bone did not hold, despite prolonged conservative treatment.

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