

# Grade 4 spondylolisthesis of the L5 vertebra associated with dural ectasia in neurofibromatosis

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## ABSTRACT

Spondylolisthesis associated with neurofibromatosis is rare, and only 12 cases have been reported so far. However, only one report of grade 4 spondylolisthesis with neurofibromatosis has been reported in the literature. A 15-year-old boy with neurofibromatosis was admitted for back pain and neurological claudication. Radiograph showed grade 4 spondylolisthesis of the L5 vertebra with scalloping of the L4–L5 vertebrae. L4–L5 laminectomy, reduction, L3–S1 posterior instrumentation and fusion were performed. The reduction of the spondylolisthesis was done entirely from the posterior approach using pedicle screws. Radiography at four months showed a broken S1 screw with a loss of reduction. The patient was re-operated on, to provide additional stability with pelvic fixation. He was pain-free with a good fusion at the two-year follow-up. Adequate posterior stabilisation with fusion gives good results in grade 4 spondylolisthesis associated with neurofibromatosis and dural ectasia.

**Keywords:** dural ectasia, grade 4 spondylolisthesis, neurofibromatosis, spondylolisthesis, vertebral scalloping

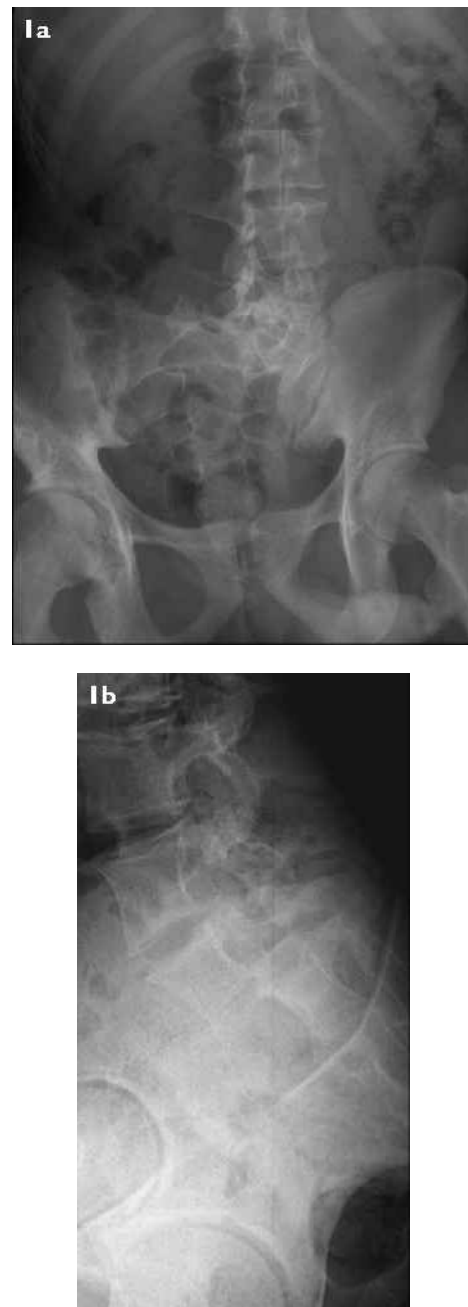
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## INTRODUCTION

Neurofibromatosis is a phacomatosis with mendelian-inherited dominance. It affects the spine and spinal cord (10%–60%) in addition to the skin and soft tissues.<sup>(1)</sup> Neurofibromatosis may be associated with dural ectasia, which is a ballooning or dilatation of the dural sac. Its mechanism is not well understood.<sup>(2)</sup> Lumbosacral spondylolisthesis with neurofibromatosis is rare, and only 12 cases have been reported so far.<sup>(1-8)</sup> We report a rare case of grade 4 spondylolisthesis of the L5 vertebra associated with dural ectasia in neurofibromatosis, and discuss the treatment strategy.

## CASE REPORT

A 15-year-old boy presented with severe back pain



**Fig. 1** Preoperative (a) anterior-posterior and (b) lateral radiographs show spondyloptosis at the lumbosacral junction with L4–L5 vertebral scalloping.

associated with neurological claudication, and a past history suggestive of Type I neurofibromatosis and scoliosis. The patient was on regular follow-up at our

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**Fig. 2** Sagittal CT image shows L4-L5 vertebral scalloping and L5 spondyloptosis with a rounded sacral endplate and widened spinal canal.



**Fig. 3** Sagittal (a) T1-W and (b) T2-W MR images show dural ectasia. (c) Axial T1-W MR image of the L4 vertebra shows vertebral scalloping with a narrow pedicle on the right side.



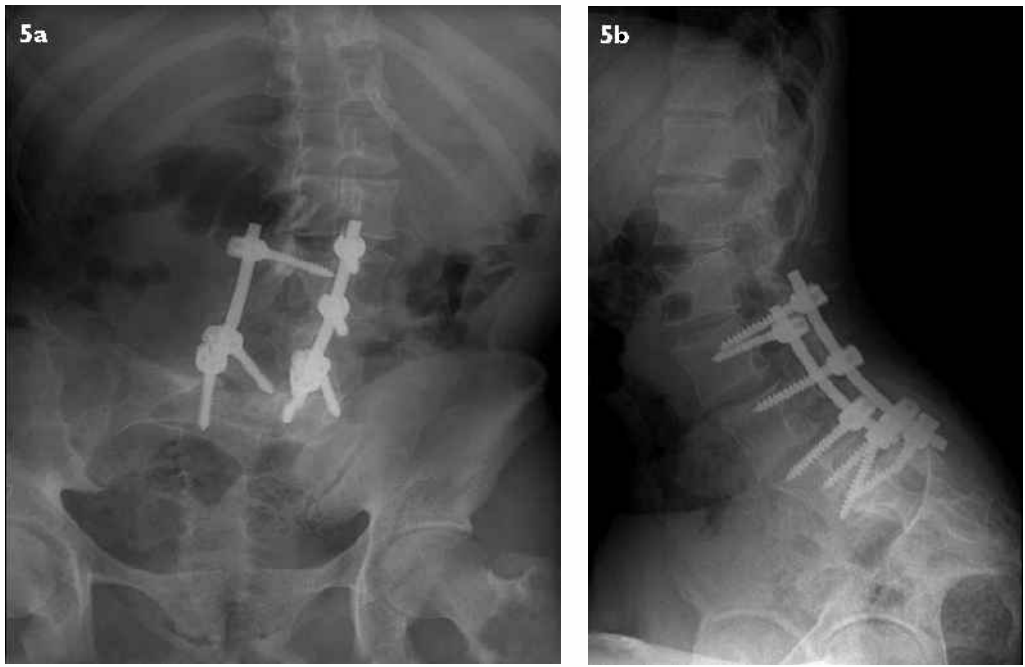
clinic, for spinal instability (grade 1 spondylolisthesis) with low back pain, and was investigated with radiographs and magnetic resonance (MR) imaging. He was advised to undergo surgical fixation and fusion procedure. However, his parents were reluctant for surgery until he

presented with a sudden onset of severe back pain and neurological claudication. He had no specific history of trauma or accident prior to presentation of the symptoms. On examination, he had multiple café au lait spots, mild left lumbar scoliosis and increased lumbar lordosis with a palpable step. Straight leg raising was 60° bilaterally and associated with hamstring tightness. He did not have any motor or sensory deficit in both lower limbs, and deep tendon reflexes were also bilaterally normal.

Radiographs showed left lumbar scoliosis, grade 4 spondylolisthesis of the L5 vertebra, scalloping of the L4-L5 vertebrae and rounding of the sacral endplate



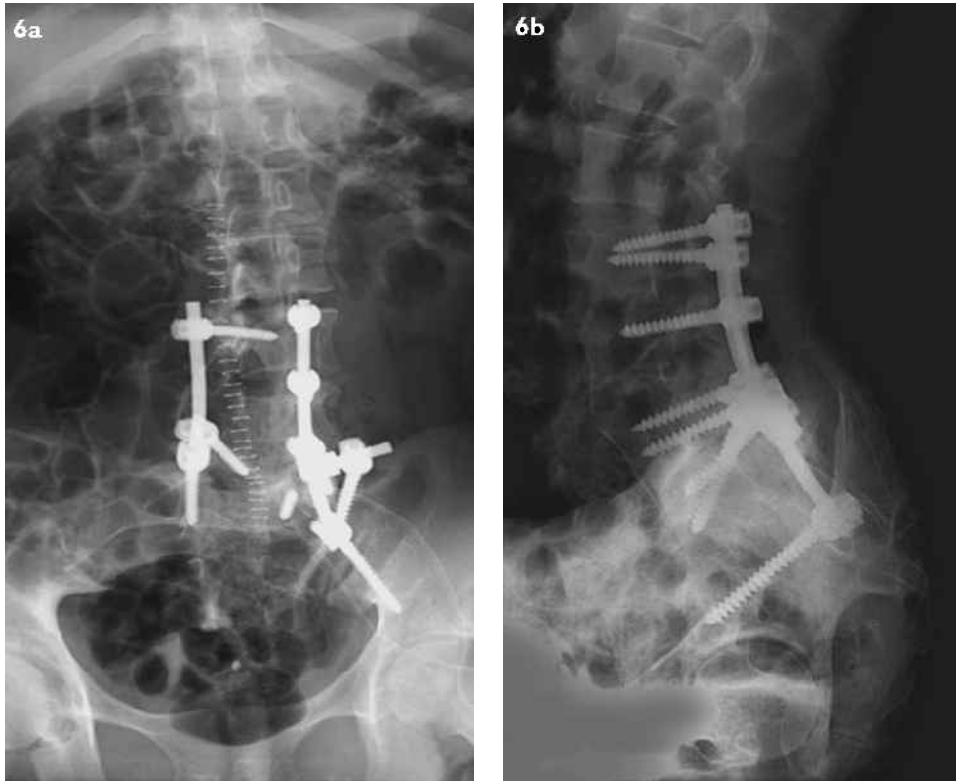
**Fig. 4** Postoperative (a) anterior-posterior and (b) lateral radiographs show spondylolisthesis reduced completely with posterior instrumentation using the pedicle screw extending from the L3 to S1 levels (pedicle screw on the L4 right side was not passed as it was narrow).



**Fig. 5** Follow-up (a) anterior-posterior and (b) lateral radiographs taken at four months show the broken S1 pedicular screw with a 25% loss of reduction.

(Figs. 1a & b). The slip angle was 44° and the Cobb's angle was 15°. Computed tomography (CT) showed a L4-L5 vertebral scalloping with a widened spinal canal (Fig. 2). The L4 pedicle was thinned out on the right side, and all other pedicles were normal. Dural ectasia was

seen on MR imaging that was taken earlier (Figs. 3a-c). Under general anaesthesia, the patient was placed in a prone position, and exposure was performed at the L3-S2 levels. L4-L5 laminectomy was done with pedicular screw fixation done at the L3-S1 levels (excluding the L4



**Fig. 6** Postoperative (a) anterior-posterior and (b) lateral radiographs taken after the second surgery shows extension of the fixation on the left side to the pelvic bone with the help of a pedicular screw through the iliac bone and extension rod. The broken screw was not removed.

pedicle on the right side due to pedicle wall breakage), and posterolateral fusion was done using autograft and allograft after reduction of the grade 4 spondylolisthesis by the posterior levered method. Complete reduction was achieved (Figs. 4a & b). During the operative procedure, continuous motor-evoked potential was used to monitor the spinal cord function. There was no abnormal finding throughout the procedure.

Postoperatively, the patient developed a transient tingling sensation in both lower limbs without any motor or sensory deficit, which subsided within four weeks. He was mobilised two weeks after surgery with the brace, and was advised to continue bracing until further advice. The patient was followed-up every two months. During the second follow-up (four months after surgery), the patient complained of backache but no radiculopathy. His backache was increased compared to the previous postoperative follow-up. Radiography showed a broken S1 screw on the left side with a loss of reduction by 25% (Figs. 5a & b). Therefore, the patient was re-operated on, and the fixation was extended caudally by a pedicular screw, through the left iliac bone without disturbing the broken screw, and bone grafting was done (Figs. 6a & b). During the second surgery, complete reduction was not attempted; instead, the goal was stabilisation and

fixation. Immediately after the surgery, his back pain was relieved. The patient was mobilised and followed up for the first six months with the bracing; thereafter, the brace was discontinued. Two years after the second operation, he was pain-free and returned to leading a normal life. Radiography showed good bony fusion, no increase in vertebral scalloping and no further loss of correction.

## DISCUSSION

Lumbosacral spondylolisthesis with neurofibromatosis is a rare disorder, and only 12 cases have been previously reported. McCarroll in 1950 reported four cases of spondylolisthesis with neurofibromatosis in a series of 46 patients;<sup>(6)</sup> Hunt and Pugh also reported two cases of spondylolisthesis with neurofibromatosis in a series of 192 patients, but did not mention the grade of slip, pedicle agenesis, dural actasia and management.<sup>(4)</sup> Mandell reported one case of bilateral hypoplastic pedicle producing spondylolisthesis with neurofibromatosis,<sup>(5)</sup> while Crawford reported one case of spondylolisthesis in a series of 116 cases of neurofibromatosis.<sup>(3)</sup> There were only three reports, the first by Winter and Edwards,<sup>(7)</sup> the second by Wong-Chung and Gillespie,<sup>(8)</sup> and the third by Toyoda et al,<sup>(2)</sup> of surgically-treated cases of spondylolisthesis associated with neurofibromatosis.

Wong-Chung and Gillespie treated their case surgically by posterolateral fusion and body cast.<sup>(8)</sup> Toyoda et al treated a case of grade 4 spondylolisthesis in a 15-year-old girl surgically by decompression, posterior lumbar interbody fusion (PLIF) and posterior stabilisation.<sup>(2)</sup>

Our patient is the third reported case of spondylolisthesis with neurofibromatosis and dural ectasia, after Bensaid et al<sup>(1)</sup> and Toyoda et al.<sup>(2)</sup> He presented with severe backache with neurological claudication but no motor or sensory deficit, and widening of the spinal canal. Because the dural ectasia with neurofibromatosis provided a wider space for the spinal cord, this allowed the cord to escape injury for cases of grade 4 spondylolisthesis.<sup>(9)</sup> This might have explained why our patient did not have any deficit even with such a severe degree of slip. To our knowledge, this is the second report suggestive of a grade 4 spondylolisthesis with neurofibromatosis.

Treatment of grade 4 spondylolisthesis in patients with neurofibromatosis varies according to the region of the spine affected, the amount of spinal instability and the symptoms.<sup>(9)</sup> In our case, because of severe instability and neurological claudication, decompression by laminectomy at the L4–L5 level, and posterior stabilisation with pedicle screws and fusion was done. The reasons for laminectomy were to avoid kinking of the dura due to the reduction procedure, to decompress the nerve roots and to visualise the dura while attempting the reduction. Yue et al published the largest series of 27 patients with high-grade spondylolisthesis in which an anterior approach for vertebrectomy and a posterior approach for laminectomy and fixation were used.<sup>(10)</sup> Their aim for laminectomy was to achieve decompression and reduction of the slipped vertebra. However, none of their patients had neurofibromatosis.

Posterior stabilisation in our case was done as advised by Winter and Edwards,<sup>(7)</sup> and Wong-Chung and Gillespie;<sup>(8)</sup> we also felt that anterior fusion would be hazardous and difficult using a pedicle screw because the vertebral bodies were small due to the scalloping by dural ectasia. Toyoda et al had suggested not to use a pedicle screw,<sup>(2)</sup> as pedicles are narrow in patients with dural ectasia. Instead, they used a long fixation from the T9 level to the pelvis using pedicle hooks and Galveston pelvic rods. In our case, we treated the patient with pedicle screw fixation, with the screws in the iliac wings. Additionally, our level of fixation was also relatively short after the second fixation. McCarroll also reported that spondylolisthesis may occur in conjunction with a congenital defect of the pedicles.<sup>(6)</sup> However, in our patient, as the L4 pedicle was narrow on the right side, the pedicle screw was avoided on that side during

stabilisation. We chose a short segment of fusion caudally (up to the S1 level) during the first surgery, which was probably the cause for the implant failure, and hence the second surgery was required. We think that the short distal fixation, by not including the pelvis during the first surgery, resulted in the development of stress concentration on the S1 pedicle screws followed by screw breakage, in spite of the good reduction initially. Based on our experience, we recommend choosing a long posterior fusion with the inclusion of pelvic fixation, like the Galveston method and PLIF in L5–S1 (as advised by Toyoda et al<sup>(2)</sup>), or pedicle screw fixation along with the inclusion of iliac wing fixation with screws, as was done in our case.

Another possible reason for the implant failure was the complete reduction performed during the first operation, which might have resulted in excessive stress on the S1 screw, that might have in turn resulted in the screw breakage. However, although the reduction was initially a little difficult with the posterior lever technique, due to soft tissue contractures, it was attempted gradually, and once it was achieved, it was stable enough to maintain the reduction status after the reduction force was released. Therefore, we opine that the short distal fixation, by not including the pelvic fixation, was the cause of the stress concentration on the S1 screw during the rehabilitation phase.

In summary, grade 4 spondylolisthesis is a difficult and controversial surgical problem. Yue et al suggested that high-grade spondylolisthesis may be associated with problems such as low fusion rate, loosening of implants, and pseudarthrosis, and therefore proposed an anterior approach in addition to posterior fixation.<sup>(10)</sup> Treatment becomes more difficult when the condition is associated with neurofibromatosis and when dural ectasia arises.<sup>(2,11)</sup> Adequate posterior stabilisation, along with posterolateral fusion, gives good results as shown by limited reports, if proper evaluation is done preoperatively to assess the presence of any pedicle or vertebral body abnormality, and if surgery is planned accordingly, thus avoiding complications associated with surgery in dural ectasia. However, in our case, we could get good fusion with no further sequel after the second operation. To conclude, patients with grade 4 spondylolisthesis with dural ectasia in neurofibromatosis requires a careful preoperative assessment and long-term follow-up, to avoid damage to the vertebral body and pedicle by dural ectasia.

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