Correction of Severe Thoracolumbar Spondylolisthesis (Grade 4) Secondary to Neurofibromatosis with Posterior Spinal Instrumented Fusion Alone. A Case Report

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SUMMARY

A 15-year-old teenager with Type 1 Neurofibromatosis presented with grade 4 spondylolisthesis over T12/L1 junction resulting paraparesis (Frankel D). Radiograph showed a Cobb angle of 88 degrees. Computed tomography scan showed dysplastic vertebral bodies, pedicles and facet joints of T11, T12 and L1 vertebra with complete T12/L1 facets dislocation. Magnetic resonance imaging confirmed presence of spinal cord compression. He underwent posterior instrumentation and posterolateral fusion (T8 to L4) using hybrid instrumentation. Extensive corticotomy of the posterior elements was followed by the use of large amount of bone graft. Post operatively, his neurology improved markedly back to normal. Radiographs showed a good correction of the deformity. He was immobilized in a thoracolumbar orthosis for six months. A solid posterior fusion was achieved at six months follow up. At 36-month follow up, he remained asymptomatic. This case report illustrates a successful treatment of a grade 4 thoracolumbar spondylolisthesis secondary to neurofibromatosis with posterior spinal fusion alone.

KEY WORDS:
Thoracolumbar, spondylolisthesis, neurofibromatosis

INTRODUCTION

Neurofibromatosis Type I, also known as Von Recklinghausen disease occurs in one in four thousand population and has 50% mutation rate [1]. This disease is one of the most common single gene disorders affecting the spine [2]. Spondylolisthesis of the thoracolumbar spine in neurofibromatosis is rare. Published English literatures in the past have been of isolated case reports in various spinal regions [3,4]. Majority have advocated the use of circumferential fusion as the mode of treatment of this condition [5,6]. This case report illustrates the treatment of a high grade thoracolumbar spondylolisthesis (grade 4) secondary to neurofibromatosis causing severe kyphotic deformity with neurological deficit corrected with posterior instrumented fusion alone.

CASE REPORT

A 15-year-old teenager with underlying Type 1 Neurofibromatosis presented with progressive deformity of the thoracolumbar spine for the past three years. Six weeks prior to presentation, he experienced gradual onset of severe back pain associated with radiating pain over his bilateral lower limbs. There was no history of trauma, fever, night sweats, loss of weight and loss of appetite.

On examination, the patient had a thoracolumbar gibbus with prominent facet joints and limited range of motions. (Figure 1) Multiple café-au–lait spots were noted over his body. Neurological examination revealed grade 4 motor power with hyperreflexia of both lower limbs. However, no sensory loss was noted. Anal tone was intact.

Results of the laboratory investigations were unremarkable. Plain radiographs of the thoracolumbar spine showed grade 4 spondylolisthesis over T12/L1 junction with a Cobb angle of 88 degrees. Computed tomography scan showed dysplastic vertebral bodies, pedicles and facet joints with complete T12/L1 facets dislocation. Magnetic resonance imaging showed spinal cord compression. (Figure 2)

The patient underwent posterolateral instrumented fusion extending from T8 to L4. Somatosensory evoked potential monitoring was used during the operation. Paravertebral muscles were dissected using a standard subperiosteal dissection. Intraoperatively, we noted that the posterior elements of T11 and T12 and L1 were dysplastic and this had given rise to a jumped facet joint of T12/L1. Hybrid instrumentation using combination of pedicle screws and hooks were used due to the dysplastic pedicles. Bilateral T12 inferior articular facet joints were removed to facilitate the reduction. The total operation time was 4 hours with total blood loss of approximately 2.1 liters.

Bone graft was harvested from the right posterior iliac crest. Extensive corticotomy of the posterior elements was performed. Large amount of autologous bone graft combined with 5 mls of demineralised bone matrix (Grafton DBM Putty, Osteotech, USA) was used. These grafts were laid in layers; the deepest layer being autologous cancellous graft, followed by demineralised bone matrix and covered with autologous corticocancellous graft.

Post operatively, his neurology had markedly improved. He was immobilized in a thoracolumbar orthosis (TLSO) for six

This article was accepted: 13 March 2012
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months. Figure 1 and Figure 2 show post operative clinical and radiographic findings which show a good correction of the gibbus with solid union noted in six months. He remained to be pain free at three years follow-up with no recurrence of spinal deformity or neurological deficit.

DISCUSSION

Spondylolisthesis in neurofibromatosis is rare, particularly in the thoracolumbar junction. In English literature, there were only two cases which have been reported on the thoracic spine to date. The first case involved a 7-year-old girl with irreducible thoracic spondyloptosis occurring at T6/T7 [3]. The other case report involved a 21-year-old female patient with spondyloptosis of T9/T10 [4]. Involvement of other levels of the spine has also been reported sporadically [3,4]. The causes for the spondylolisthesis or spondyloptosis deformities are usually due to intraspinal pathology such as tumors, meningoceles or dural ectasia. On the other hand, in some cases, no intraspinal lesion was noted [3,4]. In this patient, dysplasia of the bodies, pedicles and posterior elements, secondary to dural ectasia, may account for the deformity.

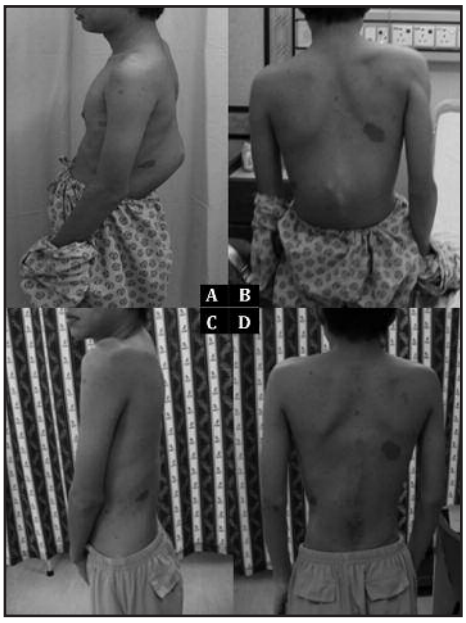

Fig. 1: (A) shows Gibbus deformity at the thoracolumbar junction; (B) shows visible facet joints noted posteriorly; (C) and (D) show correction of deformity post operatively.

In most previous reports, the surgical treatment instituted was a combined anterior and posterior approach to achieve a circumferential fusion [3,4]. Various reports have shown that posterior spinal instrumented fusion alone had a higher failure rate compared to a combined approach [5,6]. This could be due to the absence of anterior support of the spine which is often compromised by the disease process. Eventually, this would lead to failure of the implants before bony union could be achieved.

In this case, we managed to achieve a good sagittal correction using posterior approach alone. With extensive posterior corticotomy and generous application of both the autologous bone graft and demineralised bone matrix, a solid posterior bony fusion was achieved before failure of the posterior instrumentation set in. Besides that, a good sagittal alignment is also important to provide a conducive mechanical environment for bony union to take place. A postoperative external support is advisable to further enhance the stability achieved by the surgery. Six months of TLSO usage was adequate in this case.

Surgical problems encountered in this case are dysplastic small pedicles. Therefore, a preoperative computed tomography to assess the pedicle diameter is essential. A hook system should be made available if the preoperative computer tomography showed small pedicles as exhibited in

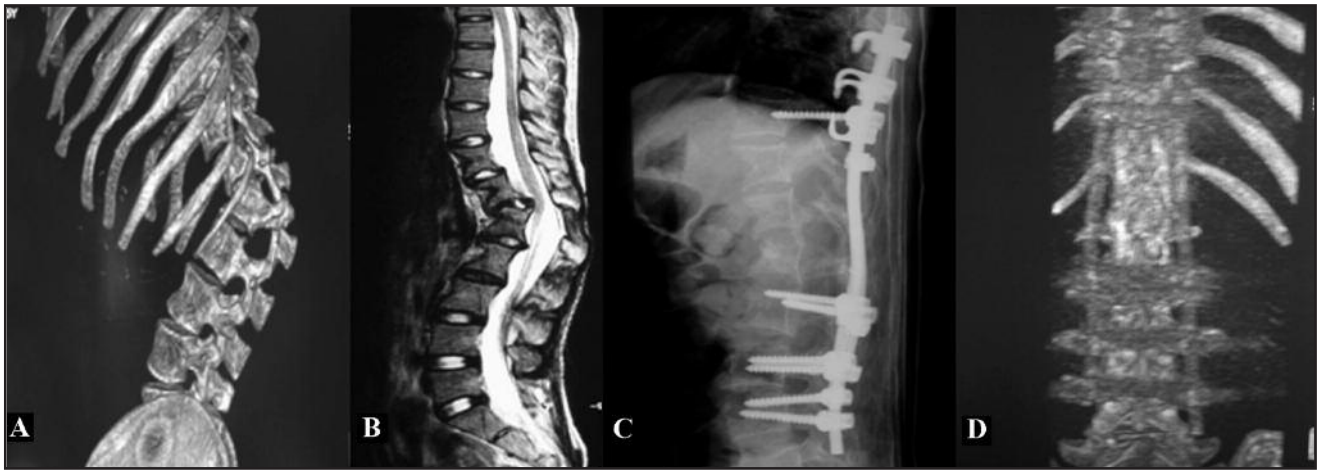

Fig. 2: (A) Reconstructed computed tomographic shows a dysplastic spine over the thoracolumbar region with bilateral facet dislocation; (B) Magnetic Resonance Image shows compression of the spinal cord; (C) Post operative radiograph showed a well corrected deformity and (D) Computer Tomography scan showed a solid fusion at 6 months.

this case. In view of the presence of preoperative neurological deficit and the amount of correction incurred, an intra-operative neurological monitoring is advisable due to the potential neurological complication.

CONCLUSION

This is a rare case of severe thoracolumbar spondylolisthesis due to neurofibromatosis which was successfully treated with posterior surgery alone.

ACKNOWLEDGEMENT

The authors wish to thank Hew Yook Ping and Shirley Wong Kueng Chii for their invaluable help in this study, as well as the case write up preparation. There is no competing of interests or supports by grant from any sources in this manuscript write up.

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