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Case Reports & Case Series

Posterior spinal fusion for scoliosis in mucopolysaccharidosis I (Hurler syndrome)

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ABSTRACT

Spinal deformity is a characteristic feature of mucopolysaccharidosis (MPS), for which surgical correction is indicated when the deformity is progressive to avoid neurological deficits and respiratory impairment. However, there exist few reports on the surgical treatment of spinal deformity in MPS and no treatment standard has been established. We herein describe the clinical and radiological outcomes of a patient with MPS-I receiving surgery for severe thoracolumbar (TL) kyphoscoliosis. The patient was a 12-year-old girl with MPS-I who underwent posterior spinal fusion with an all pedicle screw construct from T9 to L4 for a preoperative TL curve Cobb angle of 50° and TL kyphotic angle of 71°. Postoperative Cobb angle of the TL curve and TL kyphotic angle improved to 5 and 12°, respectively. Although her lumbar lordosis decreased slightly due to mild back-out of the pedicle screws in L4, bony union was achieved 3 years after surgery without perioperative complications. Based on the present case, posterior spinal fusion represents a strong treatment option for severe spinal deformity in MPS.

1. Introduction

Mucopolysaccharidosis (MPS) disorders are inherited lysosomal storage conditions caused by a deficiency in the enzymes required for glycosaminoglycan degradation. They are characterized by skeletal involvement that is often an early and prominent feature of the disease and usually results in growth deficits requiring multiple orthopaedic surgeries [1]. MPS is frequently diagnosed by the appearance of the gibbus deformity during infancy [2]. In mucopolysaccharidosis-I (MPS-I), or Hurler syndrome, the enzyme alpha-L-iduronidase necessary for the degradation of dermatan and heparan sulphate is deficient [3]. MPS-I occurs in roughly 1 in 100,000 live births [4].

Surgical correction for the spinal deformity typical of MPS patients is indicated in progressive cases to avoid neurological deficits and respiratory impairment [5,6]. To date, however, there exist few reports on surgery for spinal deformity in MPS and no treatment standard has been established. We herein describe the clinical and radiological outcomes of a patient with MPS-I who was surgically treated for severe scoliosis and thoracolumbar (TL) kyphosis.

2. Statement of informed consent

The patients and/or their families were informed that data from the case would be submitted for publication, and gave their consent.

3. Case report

This study was approved by the ethics committee of our hospital (No. 3765). The patients and/or their families were informed that data from the case would be submitted for publication, and gave their consent. A 12-year-old girl had been diagnosed as having MPS-I immediately after birth. Scoliosis was first noted at 2 years of age, and she exhibited severe TL kyphoscoliosis at age 12. Her height was 118 cm, weight was 38.1 kg, and body mass index was 27.4 kg/m^2 at consultation. We performed posterior spinal fusion by means of an all pedicle screw construct from T9 to L4 for her preoperative TL curve (T11-L5) Cobb angle of 50° and TL kyphotic angle of 71° (Figs. 1a and 2a). No obvious vertebral abnormalities were detected in the affected segment. Ponte osteotomy was performed from T11/12 to L2/3. Surgical time was 273 min and blood loss volume was 50 g. Postoperative Cobb angle of the TL curve improved to 5° and TL kyphotic angle was ameliorated at 12° (Figs. 1b, 2b). Her lumbar lordosis decreased slightly

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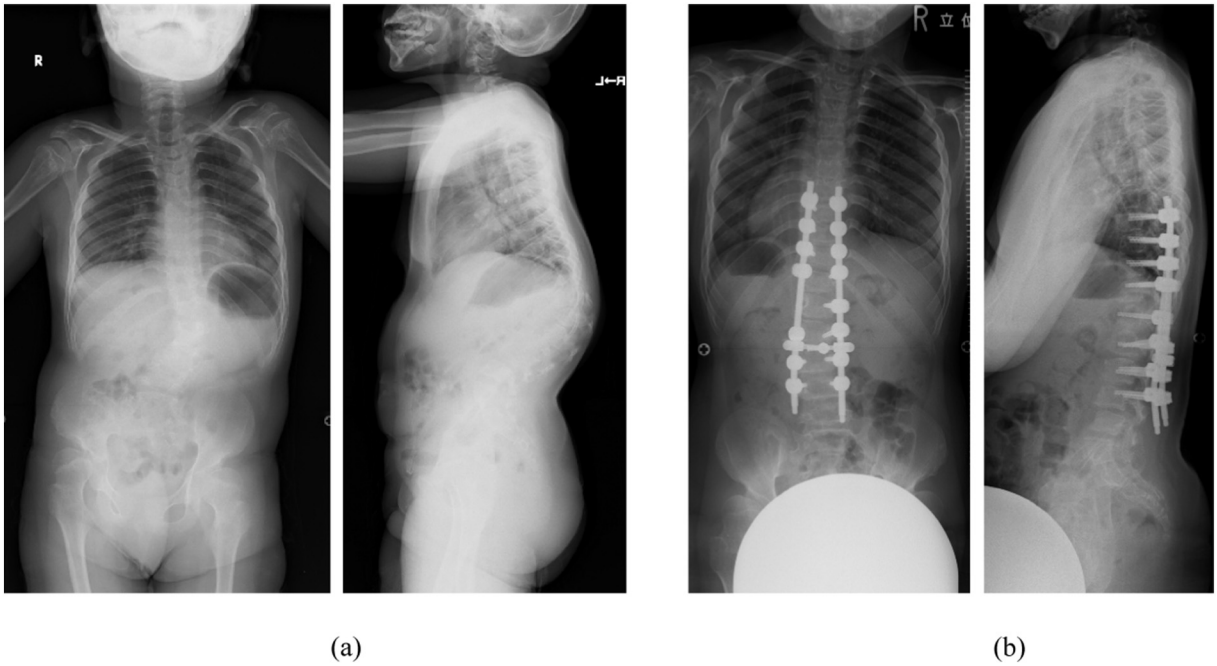

Fig. 1. Case: a 12-year-old girl.
(a) Preoperative Cobb angle of the TL curve was 50° and TL kyphosis was 71°. (b) We performed posterior spinal fusion with an all pedicle screw construct from T9 to L4. Postoperative Cobb angle of the TL curve improved to 5° and TL kyphotic angle improved to 12°.

Fig. 2. (a) Preoperative photographs show severe scoliosis with hump and kyphosis.
(b) Postoperative photographs demonstrate an improvement in body posture.

due to mild back-out of the pedicle screws in L4, but bony union was achieved 3 years after surgery (Fig. 3(a), (b)) without any apparent adverse effects. Postoperative Scoliosis Research Society-22 domain scores were 2.4, 5, 3.6, 3, 3.5, 4.5 and 3.6 for function, pain, self-image, mental health, subtotal, satisfaction, and total, respectively.

4. Discussion

In the present case of a patient with MPS-I receiving posterior spinal fusion for severe TL kyphoscoliosis, pedicle screw fixation remarkably improved radiological findings and achieved good clinical results without severe perioperative complications.

TL kyphosis and other spinal deformities are a characteristic feature of MPS patients. Similarly to congenital kyphosis, surgery for MPS is indicated when the deformity is progressive in order to prevent complications [5,6]. Early surgical management is therefore recommended, especially when sagittal balance is disturbed, as untreated TL kyphosis progression may result in late neurological deficits [7].

Surgical spinal correction in MPS patients is challenging due to the risk of progressive postoperative deformity; McMaster and Singh reported a higher risk of pseudarthrosis and neurological complications after posterior-only procedures for congenital sagittal deformities [6]. Circumferential arthrodesis combining anterior autogenous tibial strut graft and posterior arthrodesis remains a safe procedure to restore anterior height and to achieve solid fusion. Such combined procedures may also reduce the risk of neurological compromise [8–10]. Kariman et al. recommended circumferential fusion for MPS patients to achieve stable correction [11], although the invasiveness of this procedure was high. In our case, lumbar lordosis decreased slightly because of mild back-out of the pedicle screws in L4, but satisfactory bony union was achieved 3 years postoperatively.

Surgical spinal correction for MPS is also difficult due to the risk of

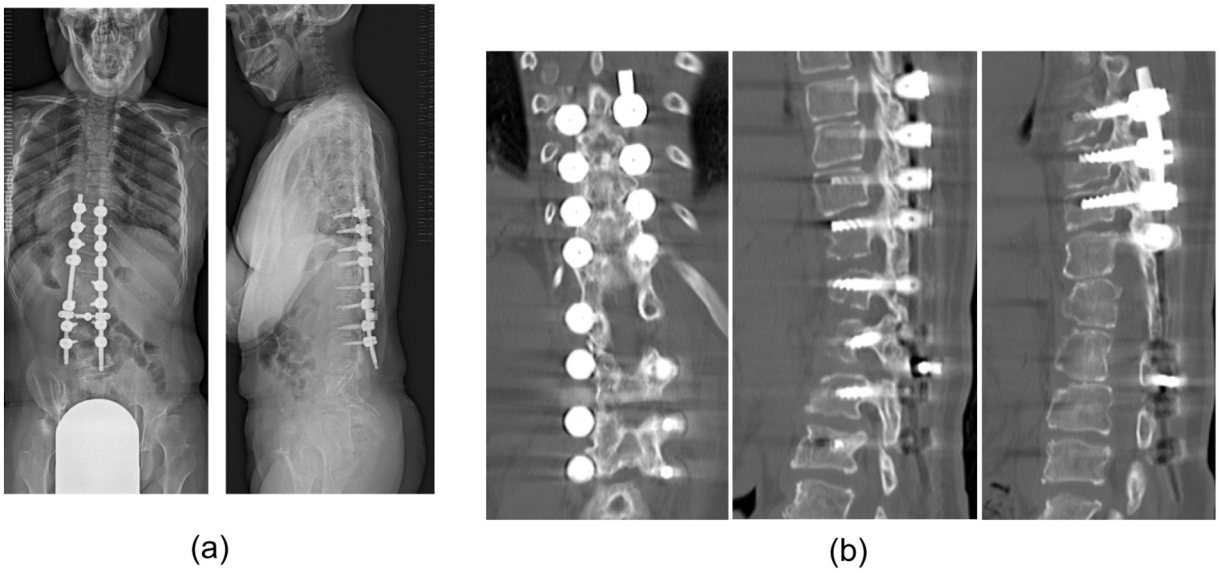

Fig. 3. (a) The pedicle screws in L4 backed out and lumbar lordosis became slightly decreased. (b) Postoperative computed tomography shows achievement of bone union 3 years after surgery.

perioperative complications. Van der Linden et al. described an overall complication rate for surgical intervention of 13.5% [12], and 2 of 13 patients reportedly died from respiratory complications stemming from respiratory insufficiency [11]. Good spinal correction was achieved in our case without severe perioperative complications.

5. Conclusion

Based on the present case, posterior spinal fusion represents a strong treatment option for severe spinal deformity in MPS patients.

Disclosure

We received no specific funding for this study.

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