

Surgical Treatment of Scoliosis in Marfan Syndrome: A Case Report

Iamze Taboridze, Associate Professor

Lali Aladashvili, Associate Professor

Grigol Robakidze University, Tbilisi, Georgia

Irakli Vardzukashvili, PhD student

David Agmashenebeli University of Georgia, Tbilisi, Georgia

Abstract

Marfan syndrome (MFS) is a connective tissue disorder of variable inheritance that affects multiple organ systems. Scoliosis affects around 60% of Marfan patients and may progress rapidly during growth spurts, leading to marked deformity, pain and restricted ventilatory deficit. The purpose of this paper is to describe a 13-year-old female patient with Marfan syndrome and surgical treatment of scoliosis. The results indicate that posterior-only surgery with instrumented fixation and fusion is effective and safe for the treatment of scoliosis in selected patients with Marfan syndrome.

Keywords: Marfan syndrome, scoliosis, surgical treatment

Introduction

Marfan syndrome is a disorder of the connective tissue that is inherited in an autosomal-dominant inherited disorder caused by mutations in the fibrillin-1 (FBN1) gene located on chromosome 15q21 that typically affects the cardiovascular, skeletal and ocular systems. Although the clinical manifestations of MFS are highly diverse within affected families, the primary skeletal manifestations, such as tall stature, chest wall abnormality and scoliosis are relatively common characteristics of MFS. These primary skeletal manifestations affect the growth pattern in MFS (1, 3).

Scoliosis affects around 60% of Marfan patients and may progress rapidly during growth spurts, leading to marked deformity, pain and restricted ventilatory deficit (4).

Adequate treatment should be provided for those with scoliosis to reduce pain, to improve overall cosmetic appearance and, most important, to improve pulmonary mechanics through reduction of spinal and chest-wall deformities. Patients with curves greater than 40-50° or with associated abnormal sagittal curvature deformities require surgery.

Surgical treatment of scoliosis associated with Marfan syndrome poses a challenge to spine surgeons (2, 5). The treatment of scoliosis in MFS has been reported to pose unique challenges. However, the information on surgical outcomes is sparse. In clinical practice, surgery for scoliosis in MFS is reported to confer higher perioperative risks and instrumentation-related complications compared with adolescent idiopathic scoliosis because of atypical and rigid curve patterns and the underlying desmogenic disorder (5).

Case presentation

A 13-year-old female patient with Marfan syndrome was first seen in the outpatient department in 2015. The patient presented skeletal abnormalities including increased height, extremity length, ligamentous laxity and scoliosis; deformities of the lens of the eye and cardiovascular abnormalities and positive family history were observed (Fig.1).

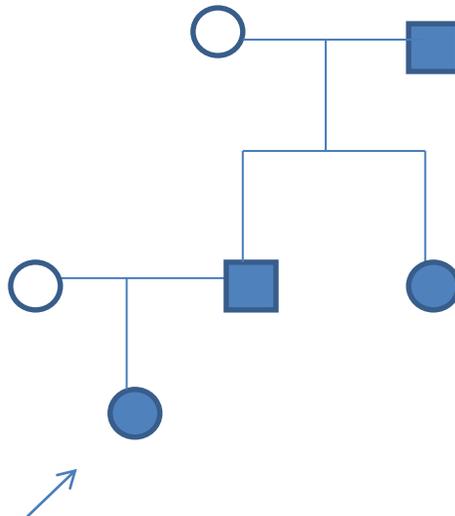


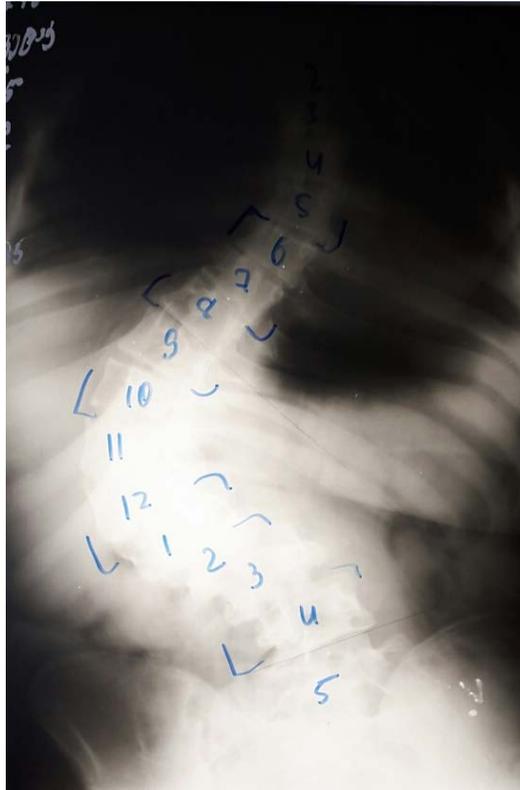
Fig.1. Pedigree of the family

Patient presenting with a scoliosis of the thoracolumbar spine

She reported of an augmenting side bolster on her loins. X-rays showed a scoliosis of 85° Cobb left-convex with an apex at the 10/11thoracvertebral body and hemi-vertebra Th11-Th12. There were no signs of neurologic symptoms.

X-ray in dynamics showed the progressive deformation of the thoraco-lumbar vertebrae in a patient with Marfan syndrome; there is loss of the normal vertebral concavity associated with progressive antero-lateral growth reduction with exaggeration of vertebral flattening associated with the development of marginal osteophytes Th10, Th11. Ligamentous laxity is

also a cause of the precocious axial osteoarthritis in Marfan syndrome (Fig. 2).



(Fig. 2). X-ray in dynamic showed the progressive deformation of the thoraco-lumbar vertebrae in dynamics in a patient with Marfan syndrome, there is loss of the normal vertebral concavity associated with progressive antero-lateral growth reduction with exaggeration of vertebral flattening associated with the development of marginal osteophytes Th10, Th11. Ligamentous laxity is also a cause of the precocious axial osteoarthritis in Marfan syndrome.

Operative procedure was planned in one steps. First, a ventral release was done at the level L1/L2, L2/L3, L3/4, L4/L5. The dorsal fusion with CD instrumentation of L1, L2, L4, Th7, Th8, Th9, Th10, Th12 with spinal husk fixation in the right position was done. After neurologic control in anesthesia, the fusion was prepared by autografts (Fig. 3).



(Fig.3): Operative procedure was planned in one steps. First, a ventral release was done at the level L1/L2, L2/L3, L3/4, L4/L5. The dorsal fusion with CD instrumentation of L1, L2, L4, Th7, Th8, Th9, Th10, Th12 with spinal husk fixation in the right position was done. After neurologic control in anesthesia, the fusion was prepared by autografts.

Before the operation Cobbe angel was 85° and the patient had psychological problems for her spine deformity, but after the operation Cobbe angel was 25° (Fig.4,5,6,7). The patient returned to the normal rhythm of her life.



Fig.3, 4. Before the operation



Fig.5, 6. After the operation:

Discussion:

The treatment of scoliosis in MFS, taking into account the individual challenges of the underlying desmogenetic disorder, can be performed with a moderately increased risk for surgical complications compared with adolescent idiopathic scoliosis (5).

Although phenotypic expression can be variable, affected patients often present with skeletal abnormalities including increased height, extremity length, ligamentous laxity, and scoliosis. Deformities of the lens of

the eye and cardiovascular abnormalities as well as positive family history is observed [4]. Our patient presented with all the mentioned symptoms.

The Marfan syndrome scoliosis patients had similar correction results, while having longer operation duration and more blood loss compared with adolescent idiopathic scoliosis (8).

As compared to anterior release combined with posterior spinal fusion, posterior-only spinal fusion could yield comparable clinical outcomes for scoliosis associated with Marfan syndrome (1).

In this case, the report before the operation Cobbe angle was 85° and patient had psychological problems for her spine deformity, but after the operation Cobbe angle was 25° and the patient returned to the normal rhythm of her life.

Conclusion

The results indicate that posterior-only surgery with instrumented fixation and fusion is effective and safe for the treatment of scoliosis in selected patients with Marfan syndrome.

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