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CASE REPORT

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Proximal realignment surgery for unilateral chronic patella dislocation in Morquio syndrome: a case report

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Mucopolysaccharidosis IVA (MPS IVA: Morquio A syndrome) is a lysosomal storage disorder caused by a deficiency of N-acetylgalactosamine-6-sulfate sulfatase. Patients with MPS IVA appear healthy at birth. Morquio-specific radiographic changes can be observed prior to clinical signs and symptoms. Patients are usually affected by a severe joint degeneration from the 2nd or 3rd decade. Hyperlaxity of the joints is prominent due to the excess of intermediate metabolites. We report a patient with inherited dwarfism, in which a proximal soft tissue realignment procedure was performed to treat chronic patellar dislocation.

Key words: Birth defect; inherited dwarfism; Morquio syndrome; mucopolysaccharide.

Mucopolysaccharidosis IVA (MPS IVA: Morquio A syndrome) is an autosomal recessive disease classified in the group of mucopolysaccharide storage diseases.[1] Cardinal clinical features were first described by Morquio and Brailsford in 1929.[2] The reported incidence of MPS IVA is variable, although average incidence is 1 in 140,000 in the Western population.[3]

Patients with MPS IVA appear healthy at birth. Morquio-specific radiographic changes can be observed prior to the appearance of clinical signs and symptoms. The initial signs and symptoms in most MPS IVA patients are identified by the age of 3 years and the classical phenotype appears at approximately the age of 6, including short trunk dwarfism (due to spondyloepiphyseal dysplasia), odontoid hypoplasia, pectus carinatum, coxa valga and valgus knees. Hyperlaxity of the joints and corneal opacities are regular findings due to the excess of intermediate metabolites.[4,5] A higher mortality rate in early adulthood has been observed in patients with Morquio syndrome because of neurological and cardiopulmonary dysfunctions; nowadays the length of survival has expanded till the sixth decade with improved comprehensive care. In addition, improvements in anesthesia and extensive preoperative examinations, including echocardiography and pulmonary function tests, prevent disastrous consequences during surgical treatment.[6]

Joint hyperlaxity, early cartilaginous changes due to abnormal deposits, abnormal mechanical stresses, and joint dysplasias cause early arthroses in the majority of patients and may lead to the necessity for early arthroplastic joint replacement surgery.[7]

We report a patient with MPS IVA, who presented with chronic patellar dislocation and was treated with a proximal soft tissue realignment procedure.

Case report

A 13-year-old girl with Morquio syndrome was admitted to our clinic with delayed physical growth,

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deformity in the knee and difficulty in walking. The patient was initially diagnosed at walking age with unilateral patellar dislocation but the diagnosis was neglected by the family due to socio-economic problems. She was the first born child of a non-consanguineous marriage. The patient also had a brother with spondyloepiphyseal dysplasia. Our patient was 100 cm tall and weighed 17 kg.

Her family noticed a waddling gait when she was 5 years old, which was progressive in nature. She was observed for the interim years at another institution. The patient presented with a complaint of waddling gait and deformity, especially in the right knee, both in flexion and extension. Additionally, she had moderate pain in the right knee and right lower extremity muscle weakness.

General physical examination revealed a short trunk and neck associated with pectus carinatum. The patient had 3 cm muscle atrophy in the right knee compared with the left, patellar dislocation in flexion, subluxation in extension and bilateral genu valgum (right 18 degrees, left 21 degrees). The Q-angle of the right and the left knee were 20 degrees and 18 degrees, respectively. The patient had full range of motion in the left knee and both hips with excessive joint hypermobility and 30 degrees of flexion contracture in the right knee. Muscle testing was normal in the left knee, both in flexion and extension and 4/5 weakness was noted in the extension of the right knee.

Radiological examination showed odontoid hypoplasia, flattening of vertebral bodies, wide intervertebral disc spaces, deficient acetabular configuration with acetabular dysplasia, deformed proximal femur with incongruous hip, and genu valgum deformities in both knees. Anteroposterior, lateral and tangential radiographs of the right knee showed patellar dislocation. Sulcus angle of the right knee was 144 degrees and the left knee was 132 degrees. Insall-Salvati index was 1.1 in the left knee.

After discussion with the patient and her parents, a proximal extensor realignment procedure with vastus medialis obliquus (VMO) advancement distally and laterally with medial reefing supported by lateral release was performed on the right knee (Figs. 1a-c). Patellar tracking was reevaluated intraoperatively.

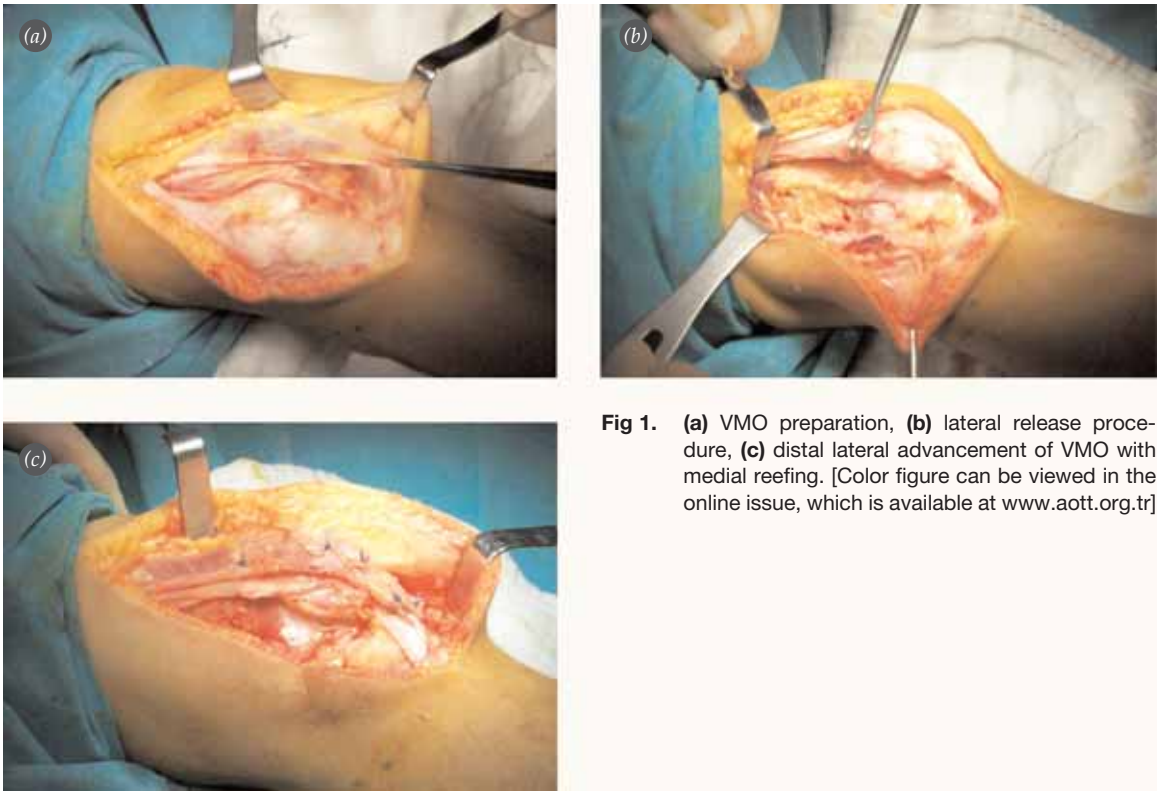

Fig 1. **(a)** VMO preparation, **(b)** lateral release procedure, **(c)** distal lateral advancement of VMO with medial reefing. [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

Intraoperative Q-angle was 12 degrees. The drain was removed within 24 hours. The patient was immobilized with a plaster cast for 15 days, followed by a knee immobilizer for 4 weeks. During the immobilization period active straight leg raising exercises were begun, followed by active range of motion and strengthening exercises. At the 3rd postoperative month, full range of motion was gained. Radiographs taken in the first year follow-up showed the proper maintenance of patellar alignment and tracking (Fig. 2).

Fig 2. Lateral view in flexion at the 1 year follow-up.

Discussion

Morquio syndrome is an autosomal recessive inherited error in the mucopolysaccharide metabolism, characterized by the presence of keratosulfate in urine and Reilly granulations in the cytoplasm of leukocytes. Skeletal manifestations of this condition include odontoid hypoplasia, trunk dwarfism, platyspondyly and wide disc spaces. A pelvis with narrowing at the acetabulae, coxa valga, widening of pubic symphysis, and genu valgum have also been described.[2,3,6] Other manifestations are a shortening of the metacarpals, small carpal bones and distal radial, as well as ulnar dysplasia. Generalized ligamentous laxity is prominent in most cases. Most of the characteristic clinical and radiographic changes were present in our patient.

Patients with inherited dwarfism, such as spondyloepiphyseal dysplasia and Morquio syndrome, pose challenging problems to orthopedic surgeons due to deformities that develop in the first decade of their lives. A review of the literature revealed reports on arthroplasty for the treatment of hip and knee arthropathies after severe cartilage degeneration or osteotomies for genu valgum deformities.[3,6-8] Procedures described for the treatment of recurrent subluxation or dislocation of the patella can generally be divided into five categories: (1) release of the tightened lateral retinaculum, (2) proximal realignment of the extensor mechanism, (3) distal realignment of the extensor mechanism, (4) combined proximal and distal realignment of the extensor mechanism, and (5) patellectomy combined with realignment of the extensor mechanism. Indications for soft-tissue procedures include instability secondary to medial laxity with or without trochlear dysplasia, fine-tuning of distal realignment procedures, and instability in skeletally immature patients. Indications for distal realignment include patellar instability secondary to malalignment as indicated by a Q-angle of more than 20 degrees and anterior tibial tuberosity-to-trochlear groove distance of more than 15 mm, as well as patellar instability with inferior and lateral chondromalacia. This technique should not be used in children with open proximal physes.[9]

We found no published reports in the English literature on treatment of patellar dislocation in patients with Morquio A syndrome.

Altered biomechanics resulting from bone deformation and joint dysplasia can lead to degenerative changes, especially overloading of lower extremities, in patients with Morquio A syndrome. Our patient presented with complaints of deformity, moderate pain and weakness in the right knee due to chronic patellar dislocation. We decided to perform a proximal realignment procedure with the aim of realigning the extensor mechanism to correct the flexion deformity and prevent early deformation and degeneration of the right knee. The one year follow-up showed satisfactory result with full range of motion.

We can conclude that the realignment for the unreduced patellar dislocation in this patient with Morquio A syndrome showed a satisfactory short-term result.

Conflicts of Interest: No conflicts declared.

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