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Case Report

Bilateral Medial Patellofemoral and Medial Patellotibial Ligament Reconstruction in a Patient with Nails Pattela Syndrome – A Rare Case and Review of the Literature

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Abstract

Nail-patella syndrome (NPS), is an infrequent inherited dysplasia that primarily involves structural underdevelopment of the nails and patellae. Patellar instability is a prevalent manifestation in NPS and, in many cases, necessitates stabilisation through surgical means. Although the majority of published work on the knee involvement in this syndrome concentrates on its clinical presentation and radiographic features, there is limited literature addressing surgical management. This case report details a 19-year-old male diagnosed with NPS, who presented with bilateral anterior knee pain and demonstrated recurrent patellar dislocations affecting both knees. The present study describes a patient with NPS treated surgically by reconstruction of the medial patellofemoral (MPFL) and medial patellotibial ligaments (MPTL) bilaterally with excellent clinical and functional results.

Keywords: Knee; Nails Patella Syndrome; Patellofemoral Reconstruction

Introduction

Nail-patella syndrome (NPS) is an uncommon hereditary disorder that involves a spectrum of abnormalities affecting tissues derived from both ectodermal and mesodermal lineages [1].

The prevalence of NPS is approximately 1 in 500,000, with a high degree of penetrance and variable expression [2].

In NPS, the musculoskeletal system is prominently involved, with patellar abnormalities reported in nearly 90% of affected individuals. Complete absence of the patella, however, occurs in only about one-fifth of cases. More frequently, the patellae are hypoplastic, leading to knee instability characterised by recurrent subluxation or dislocation.

In this context, episodes of patellar instability are commonly linked to patellar absence or marked attenuation of the medial patellofemoral ligament (MPFL). A range of surgical strategies has been described to address recurrent patellar instability in individuals with this condition [3].

This study presents a case of a patient with NPS experiencing recurrent patellar dislocation, who underwent bilateral surgical reconstruction of the MPFL and MPTL with excellent clinical and functional outcomes [4].

Case Report

A 19-year-old man presented in February 2018 at the Hospital ,after multiple evaluations at other institutions, due to recurrent

bilateral patellar dislocation significantly impacting his quality of life and daily physical activities. On physical examination, he presented with right shoulder instability, limited extension in both elbows, reduced grip strength, left-sided quadriceps atrophy, and more pronounced patellar instability on the left. Specific knee examination revealed habitual knee dislocation in flexion, irreducible, assuming a lateral position. He had full and painless range of motion bilaterally, with no ligamentous instabilities and inconclusive meniscal tests.

Anteroposterior radiographs demonstrated lateral patellar subluxation without significant patellar hypoplasia (Figure 1).

Figure 1: Anteroposterior radiographs demonstrated lateral patellar subluxation without significant patellar hypoplasia. Type III Wiberg – patellar morphology.

CT scan revealed a tibial tubercle–trochlear groove (TT-TG) distance at the upper limit of normal (16/17 mm), increased patellar tilt upon contraction (38°/41°), patellar subluxation under contraction (10/14 mm), and a dysplastic (nearly flat) trochlea (Figure 2).

MRI of the left knee showed "signs of femoropatellar instability, with a dysplastic femoral trochlea associated with a dysmorphic patella, medial facet hypoplasia, and lateral tilt". No changes in the remaining ligamentous and meniscal structures (Figure 3).

Figure 2: CT scan revealed a TT-TG normal, increased patellar tilt upon contraction and a dysplastic (nearly flat) trochlea.

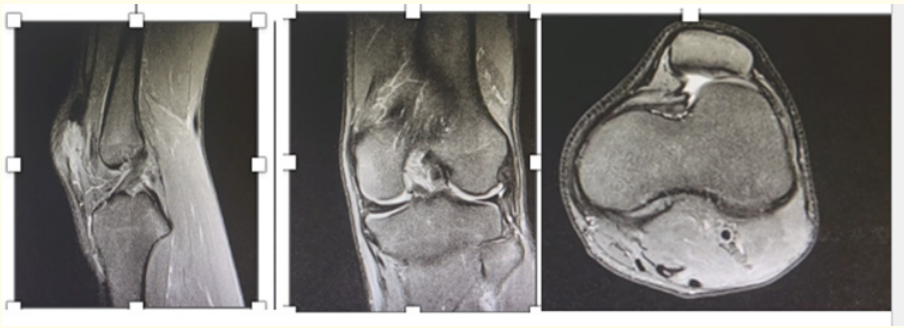

Figure 3: MRI of the left knee showed signs of femoropatellar instability. The menisci, cruciate ligaments, and collateral ligaments were intact.

The patient underwent an initial surgical intervention on the left knee, which included release of the lateral retinacular, transposition of the vastus lateralis and combined reconstruction of the medial patellotibial ligament and reconstruction of the MPFL.

Surgical treatment

The patient was positioned supine, and a pneumatic tourniquet was applied to the operative limb. A longitudinal incision was performed over the anterior aspect of the left knee. After the patient was anesthetized, passive mobility was tested, which revealed a dislocation of the rotula in flexion that reduced in extension (Figure 4). The lateral retinaculum was tight and so a retinacular release was performed and a proximal transposition of the vastus lateralis was also performed to avoid the deforming forces responsible for dislocating the patella. The medial patellofemoral ligament structure was not found; therefore, the combined MPFL and MPTL reconstruction was reconstructed using the combined technique . The gracilis tendon was harvested and prepared. The tibial-patellar ligament was reconstructed using an interference screw fixation and swive lock anchor in the tibia, and MPFL reconstruction was performed with an interference screw fixation in the femur (Figure 5).

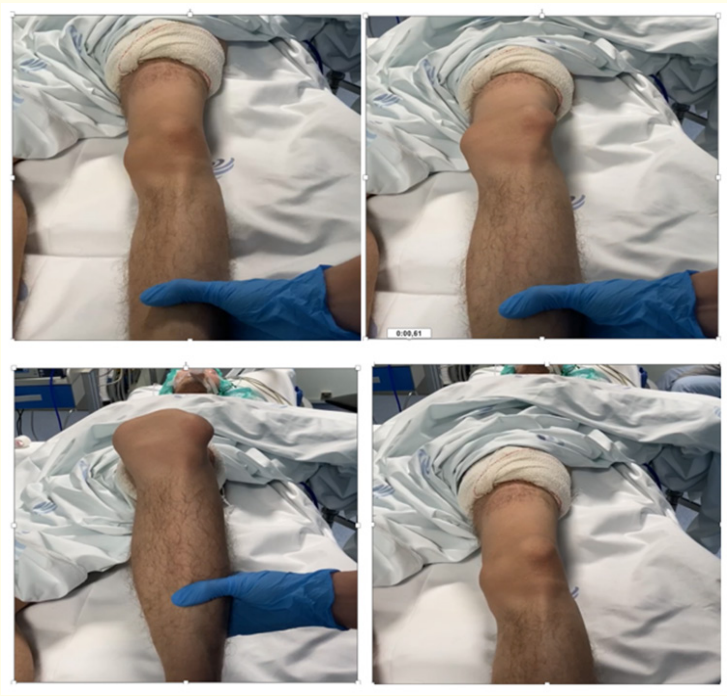

Figure 4: After the patient was anesthetized, passive mobility was tested, which revealed a dislocation of the rotula in flexion that reduced in extension.

Figure 5: The gracilis tendon was harvested and prepared. The tibial-patellar ligament was reconstructed using an interference screw fixation and swive lock anchor in the tibia, and MPFL reconstruction was performed with an interference screw fixation in the femur.

Outcome

Six months postoperatively, the patient had excellent clinical outcomes, with the patella maintaining its position in the trochlea. He exhibited a minor extension deficit of 5° and flexion up to 140°. At the one-year follow-up, the patient demonstrated complete restoration of knee range of motion and remained asymptomatic. Given the successful functional outcome, the patient underwent the same surgical procedure on the contralateral knee one year after the initial surgery (Figure 6).

Figure 6: Intraoperative images of the combined MPFL and MTPL reconstruction of the contralateral knee, right knee.

Discussion

NPS is an uncommon hereditary condition defined by a characteristic constellation of musculoskeletal findings, notably nail dysplasia, marked hypoplasia or absence of the patellae, underdevelopment of the radial head and capitellum, and the presence of iliac horns [3].

Patellar involvement is observed in most cases and may include absence or hypoplasia with subluxation. The majority of cases remain clinically silent and do not necessitate operative management.

However, congenital permanent patellar dislocation (CPPD) necessitates surgical treatment [5]. This case describes a young patient with nail-patella syndrome presenting with recurrent bilateral patellar dislocation significantly affecting his quality of life.

Multiple surgical strategies are available for the management of patellar dislocation, and the selected approach is determined by the specific etiological factors involved. A short list of interventions includes double-strand gracilis autograft, combined MPFL repair and medial patellotibial ligament reconstruction, single-strand quadriceps tendon autograft, split adductor magnus tendon transfer, adductor sling techniques, and medial collateral ligament reconstruction [6].

The goal of MPFL reconstruction techniques is to provide effective stabilization without compromising normal knee function [7].

Various autologous grafts, including semitendinosus, gracilis, quadriceps, and semimembranosus tendons, as well as medial retinacular tissues, have been utilized. Any bone tunnels should be large enough to accommodate the graft.

When employing a two-strand tendon construct, patellar fixation is typically performed at the superomedial pole and at the transition between the proximal and middle thirds of the medial patellar margin, or slightly distal to this level [8].

The femoral attachment may be positioned approximately 10 mm proximal and 2 mm posterior to the medial femoral epicondyle, or alternatively about 4 mm distal and 2 mm anterior to the adductor tubercle. A true lateral fluoroscopic projection of the knee allows accurate localisation of the femoral attachment site, commonly referred to as the Schöttle point [9].

In this case, MPFL and medial patellotibial ligament reconstruction using bilateral gracilis tendon grafts resulted in excellent functional outcomes. Postoperatively, the patient had a stable patella congruent with the femoral trochlea.

The immediate postoperative range of motion was acceptable at 80°. By the three-month follow-up, the patient had achieved 130 degrees of knee flexion and was mobilising independently without assistive devices. One year later , he was fully mobile and pain-free No complications, such as fractures, wound infections, or redislocation, occurred.

Conclusion

In conclusion, secondary patellar dislocation is a common occurrence in NPS. The predominant underlying mechanism is disruption of the MPFL, and surgical management should be directed toward restoring the integrity of this ligament through reconstruction.

This case indicates that MPFL and medial patellotibial ligament reconstruction using a gracilis tendon graft is a safe and effective method and can be utilized in NPS cases caused by MPFL rupture.

This case suggests that the combined technique can effectively re-establish physiological patellar tracking in patients with NPS-associated patellar instability, leading to satisfactory restoration of knee motion and functional performance.

Disclosure of Interest

The authors declare that they have no conflicts of interest concercing this article.

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