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Case Report

Total Knee Arthroplasty Without Reduction of the Patella for Genu Valgum With Permanent Dislocation of the Patella: A Case of Nail Patella Syndrome

Teruya Ishibashi, MD, PhD [a], Tetsuya Tomita, MD, PhD [a, b, *], Masashi Tamaki, MD, PhD [c], Toshitaka Fujito, MD, PhD [d], Seiji Okada, MD, PhD [c]

[a] Department of Orthopaedic Biomaterial Science, Osaka University Graduate School of Medicine, Suita, Osaka, Japan
[b] Morinomiya University of Medical Sciences, Graduate School of Health Sciences, Osaka, Japan
[c] Department of Orthopaedic Surgery, Osaka University Graduate School of Medicine, Suita, Osaka, Japan
[d] Department of Orthopaedic Surgery, Hoshigaoka Medical Center, Hirakata, Osaka, Japan

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ABSTRACT

A 75-year-old woman presented with progressive bilateral knee pain and severe genu valgum. She could walk utilizing braces and T-canes, with a 20° flexion contracture and 150° of maximum flexion. During knee flexion, the patella laterally dislocated. Radiographs demonstrated severe bilateral lateral tibiofemoral osteoarthritis and patellar dislocation. She underwent posterior-stabilized total knee arthroplasty without patellar reduction. After implantation, the knee range of motion was 0°-120°. Intraoperative findings revealed that the affected patella was too small, low articular cartilage volume resulted in the diagnosis of nail patella syndrome with the tetrad of nail dysplasia, patella dysplasia, elbow dysplasia, and iliac horn. At the 5-year follow-up visit, she could walk without a brace and had a knee range of motion (10°-135°) with clinically favorable results.

Introduction

Permanent dislocation of the patella (PDP) and severe genu valgum may coexist and exacerbate one another [1]. The Q angle increases due to the valgus knee, which makes patellar reduction difficult [2]. PDP can reduce ambulation ability and knee flexion angles. Unless appropriate treatment is performed for the valgus knee, the affected knee progresses to subsequent lateral compartmental osteoarthritis (OA) [3]. Therefore, it is crucial to conduct a proper diagnosis and perform early interventions. Treatment options for PDP and tibiofemoral compartmental OA with genu valgum include total knee arthroplasty (TKA) in combination with positions of the femoral and tibial components, proximal and/or distal realignments, and medial patellofemoral ligament reconstruction (MPFL-R) [4,5]. Reconstruction of the knee extensor mechanism reduces pain and improves knee function. However, these surgeries are technically demanding and can cause substantial complications. There are few reports of TKA without patellar reduction for PDP and genu valgum [6,7]. In addition, congenital dislocation of the patella (CDP) demonstrates unusual bone morphology, such as rotational change, hypoplasia, and aplasia. These deformities may make it even more difficult to reconstruct the knee extensor mechanism and worsen tibiofemoral compartmental OA.

Herein, we report our experience of a patient with severe valgus knee OA and PDP who underwent TKA without patellar reduction.

The patient provided written informed consent for the publication of the data concerning the case.

Case history

A 75-year-old woman was admitted to our hospital with progressive right knee pain and severe genu valgum. She underwent orthotic treatment for left congenital dislocation of the hip at birth, total hip arthroplasty for ipsilateral hip OA at the age of 55 years,

* Corresponding author. Morinomiya University of Medical Sciences, 1-26-16 Nankokita, Suminoe-ku, Osaka-shi, Osaka 559-8611, Japan. Tel.: +81 66616 6912.
E-mail address: tetsuya_tomita@morinomiya-u.ac.jp

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and revision surgery for aseptic loosening of the cup at the age of 69 years. She had no history of trauma or knee surgery. Her ambulation was limited due to right knee pain and instability, requiring bilateral knee braces and T-canes. The appearance of her lower extremities revealed severe genu valgum, which was uncorrected by manual stress. Manual muscle testing showed grade 3/4 on the right and left knees. The range of motion (ROM) was 20°-150°, and the patella was laterally dislocated during knee flexion. She was unaware of her bilateral dislocation condition. On long-leg anteroposterior weight-bearing radiographs, she demonstrated severe genu valgum of the right knee with a hip-knee-ankle angle (HKAa) of valgus 29.0°, grade IV osteoarthritic changes in the lateral femorotibial compartment according to the Kellgren-Lawrence classification, and laterally dislocated patella. The lateral radiograph demonstrated dysplasia of the lateral femoral condyle. Radiographs on the skyline view at 30°, 60°, and 90° demonstrated the dysplasia of the trochlea with a sulcus angle of 150° and laterally located patella (Fig. 1). Computed tomography of the knee revealed a tibial tuberosity to trochlear groove distance of 41.3 mm. There were no remarkable laboratory tests, suggesting the presence of rheumatoid arthritis. The preoperative knee score was 20/30, and the functional score was 60/60 on the right and left knees, respectively.

TKA was planned to treat severe OA, and both proximal and distal realignments were planned to treat PDP. The operation was performed under general anesthesia. A longitudinal midline incision was made to expose the extensor mechanism. We transversely resected the tightened iliotibial band and cut the lateral retinaculum; however, the patella was still laterally fixed. Therefore, we performed a medial para-patellar approach and everted the patella. The patella was very small (10 mm) and thin (6 mm), with little articular cartilage (Fig. 2). Considering that the patella was too small to function as an extensor mechanism, we decided not to reduce or replace the patella. Femoral alignment was aimed to slightly valgus to prevent mediolateral instability using an intramedullary rod. For the tibia, alignment was aimed perpendicular to the mechanical axis in the coronal and sagittal planes using an extramedullary referencing cutting guide. Femoral rotation was aimed to 4° external to the posterior condyle axis. Tibial rotation was determined using the ROM technique [8]. TKA was performed using a fixed posterior-stabilized (PS) type (TRIATHLON; Stryker Orthopaedics, Mahwah, NJ) #1 femoral component, #1 tibial component, and a PS tibial polyethylene insert with a thickness of 9 mm. After implantation, the passive ROM was 0°-120° and revealed no impingement of the patella. Rehabilitation involved the same protocol as conventional TKA with progressive continuous passive motion, weight-bearing as tolerated, ambulation, and isometric quadriceps exercises on postoperative day 1. The conventional rehabilitation menu was adapted; on postoperative day 2, she walked on full weight-bearing as tolerated and passive ROM exercises. On postoperative day 21, the ROM was 10°-110°, and the long-leg anteroposterior radiograph exhibited HKAa of valgus 16° and valgus 16° on the right and left knees, respectively. The patient was discharged for further rehabilitation. Following a systemic examination because of the small patella, she was diagnosed with nail patella syndrome (NPS) with the tetrad of nail dysplasia, patella dysplasia, iliac horn, and elbow dysplasia (Fig. 3). At 1-year follow-up, the knee ROM was 5°-125°, with HKAa of valgus 13° and valgus 17° on the right and left knees, respectively. Annual follow-up revealed almost fixed-knee ROM (10°-125°) and HKAa of valgus 11° and valgus 16° on the right and left knees, respectively. At the latest follow-up (5 years), her ambulation had improved without a knee brace, and the ROM of the affected knee was 10°-135°, with an extension lag of 10°. Radiographs showed no evidence of loosening or fracture and HKAa of valgus 11° and valgus 17° on the right and left knees, respectively (Fig. 4). The postoperative Knee Society Knee score and Function score improved to 60/50 and 85/85 on the right and left knees, respectively.

Discussion

We experienced two clinically important issues in patients with severe genu valgum and permanent patellar dislocation. TKA

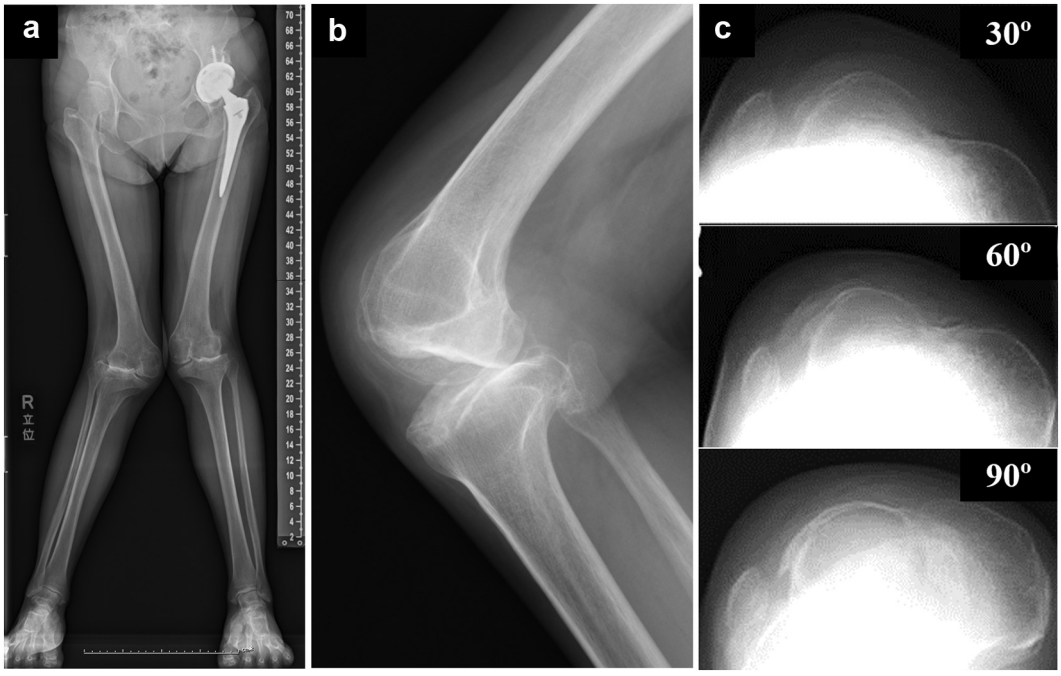

Figure 1. Preoperative radiographs demonstrating bilateral severe valgus knee deformity, lateral compartment osteoarthritis in the long-leg anteroposterior view (a), dysplasia of the lateral femoral condyle in the lateral view (b), and permanent dislocation of the patella in the skyline views at knee flexion of 30°, 60°, and 90° (c).

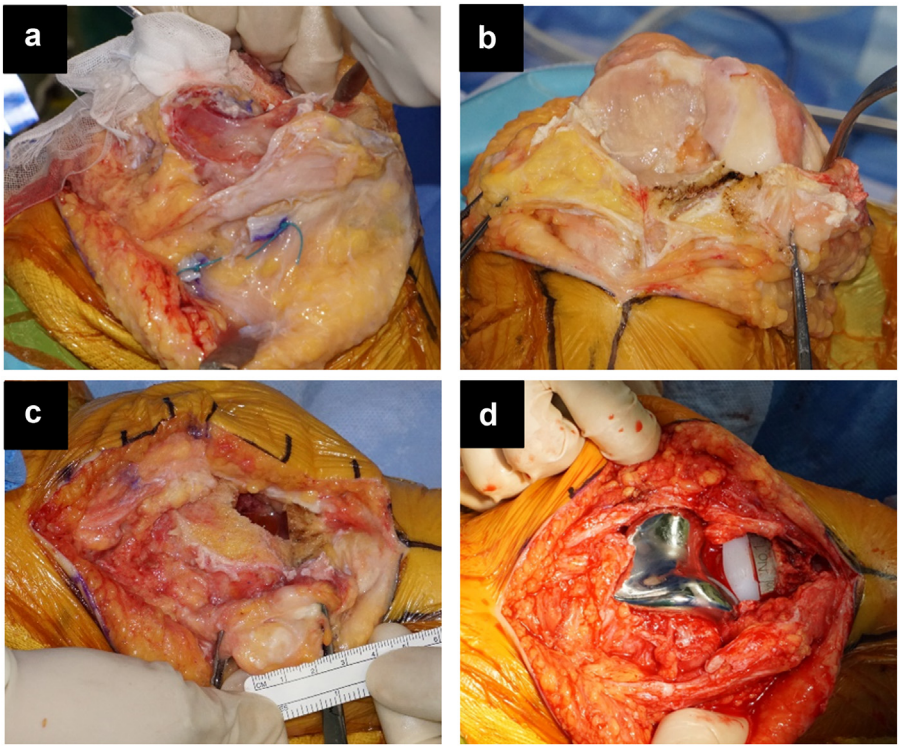

Figure 2. Operative pictures demonstrating the resected iliotibial band which was very tight (a), eburnation of the lateral femoral condyle (b), patella dysplasia (c), and no sign of impingement of the patella after implantation (d).

without patellar reduction is one of the treatment options for severe genu valgum with permanent patellar dislocation. A small patella can be a sign of congenital malformations, including NPS.

First, TKA without patellar reduction for genu valgum with PDP demonstrated good clinical results for 5 years. Patellar reduction in severe genu valgum involving PDP is challenging to treat.

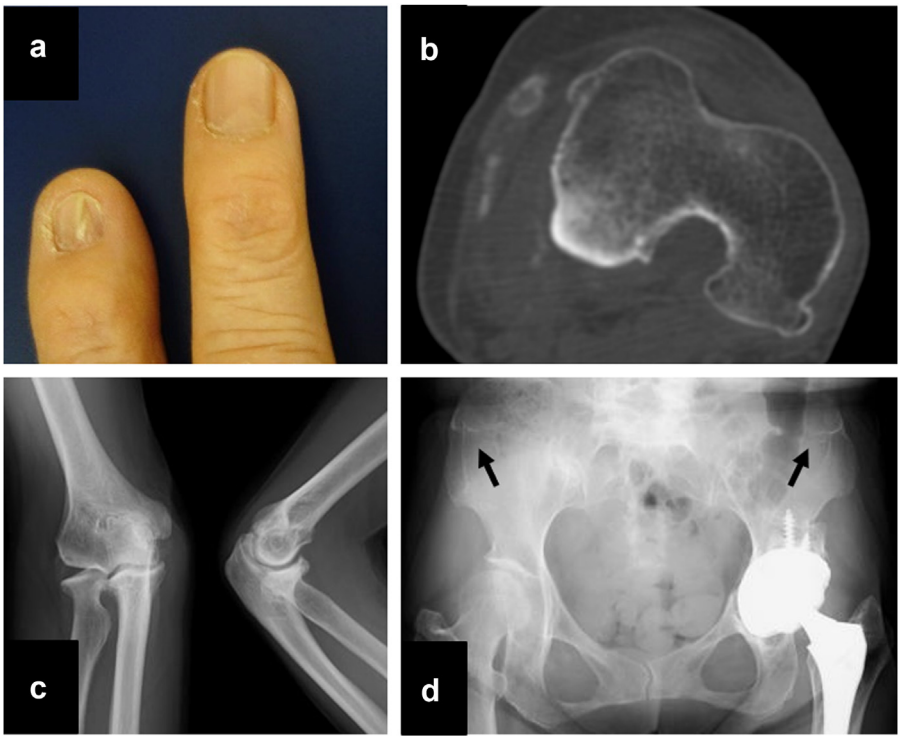

Figure 3. Tetrad sign of nail patella syndrome. Nail dysplasia and triangular lunulae were observed (a). Computed tomography showed a small patella (b). Elbow radiographs showed valgus deformity, dysplasia, and posterior dislocation of the radial head (c). The iliac horn is indicated by black arrows (d).

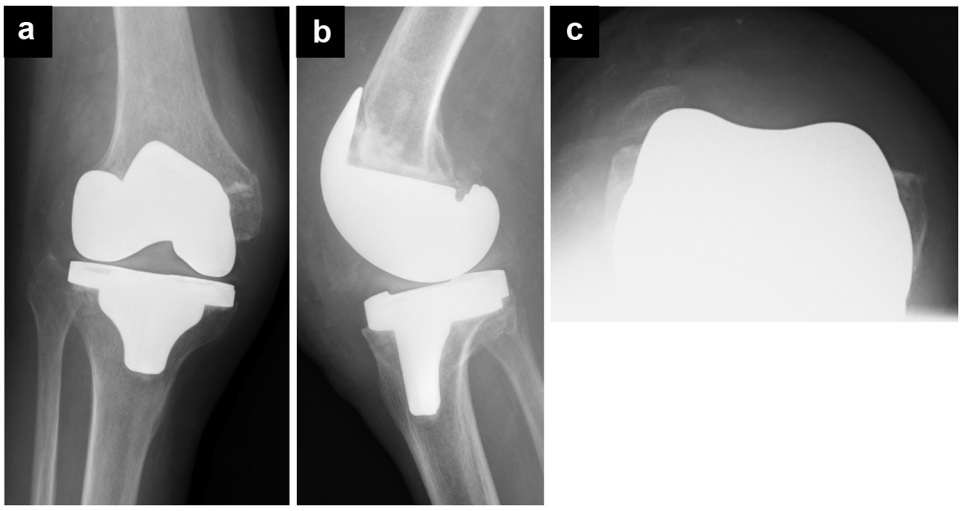

Figure 4. Five-year postoperative radiographs demonstrating no sign of loosening and fracture in the anteroposterior view (a) and lateral view (b), with dislocation of the patella remaining (c).

Considering the knee stability and pain, it is crucial to reduce the patella. Several treatment options in combination with TKA involve proximal and distal realignments, MPFL-R, and intentionally laterally and/or externally rotated positioning of the femoral component [4,5]. Marmor reported that bilateral genu valgum with PDP did not reduce the patella, preventing the loss of knee flexion [6]. In contrast, Bullek et al. reported 3 cases of TKA with proximal realignment for PDP complicated by severe genu valgum [1]. They recommended the realignment of the extensor mechanism to restore normal function and kinematics. In addition, Yamanaka et al. reported a case of bilateral genu valgum accompanied by CDP, in which 1 side underwent TKA with distal realignment, and the other side underwent TKA without reduction of the patella [9]. The results showed that the side with a reduction of the patella experienced a postoperative tibial tuberosity fracture, with knee flexion ranging from 0° to 90°. The side without patellar reduction had a ROM from 45° to 125°. In the case of CDP, several anomalies might exist, such as laterally positioned vastus medialis and anteriorly located pes anserinus, resulting in the semitendinosus and gracilis serving as an extensor [10]. In cases of congenital rather than acquired patellar dislocation, the extension mechanism cannot function as a true extensor because the bone morphology of the distal femur and proximal tibia has demonstrated externally rotated deformity, unlike the normal mechanism. The flexor in the normal knee could likely serve as an extensor in CDP. In this case, there was no remarkable trauma history around the knee joint, or proximal tibial external rotated deformity (tibial tuberosity-trochlear groove distance 41.3 mm), indicating the presence of congenital and PDP. Therefore, no reduction of the patella could worsen the pain and instability of the affected knee because the patella was not a part of the extensor but the flexor.

A small patella and CDP are essential findings for hereditary systemic diseases. Studies have shown that patients with a small patella or patellar aplasia had NPS, small patella syndrome, Meier-Gorlin syndrome, RAPADILINO syndrome, genitopatellar syndrome, and isolated patellar aplasia or hypoplasia [11,12]. Each patient had an accompanying malformation. Patients with NPS have four unique manifestations: nail dysplasia, patellar dysplasia, iliac horn, and elbow dysplasia [13]. Renal disease and open-angle glaucoma have been reported. In this case, postoperative systemic physical and radiographical evaluations resulted in a diagnosis of NPS because of moderate patellar dysplasia. In addition, family history is an important factor in diagnosing these diseases. NPS is an autosomal dominant hereditary disease caused by mutations in LMX1B on chromosome 9, which is important for limb development in vertebrates [14]. In this case, the patient's parents had already died, and no further information was available. One of the sons complained of knee symptoms; however, he did not seek medical attention because of his busy schedule.

Knee instability and patellar dislocation are the most frequent symptoms associated with NPS. The goal of surgical treatment for these conditions in children or adults is to realign the extensor mechanism using proximal and distal realignments and MPFL-R. However, there is no gold standard for cases of delayed diagnosis

Table 1
Review of surgical procedures for osteoarthritis of the knee with nail patellar syndrome.

Author	Patellar condition	Realignment	Osteoarthritis	Arthroplasty	Patellar resurface	Complications
Lachiewicz and Herndon [15]	Dislocation	Medial imbrication lateral release	Medial	TKA	No	No
Louboutin et al. [16]	Dislocation	Lateral approach	Lateral	TKA	No	Patellar tendon rupture
Louboutin et al. [16]	Dislocation	Hauser procedure MPFL reconstruction	Patellofemoral	PFA	Yes	
Curbo et al. [17]	Dislocation	Medial imbrication Lateral release	Medial Kateral	TKA	No	Arthrofibrosis
Yang et al. [18]	Subluxation	No	Medial	UKA (medial)	No	
Vakharia et al. [19]	Dislocation	No	Lateral	UKA (lateral)	No	
Serrano and Serrano [20]	Aplasia	No	Lateral	TKA	No	

PFA, patellofemoral joint arthroplasty; UKA, unicompartmental arthroplasty.

or severe OA. There have been a few case reports involving different treatment strategies (Table 1). Lachiewicz and Herndon first reported PS TKA with proximal realignments and no resurfacing of the patella in patients with NPS affecting medial OA and PDP [15]. Louboutin et al. reviewed the management of patellar problems in adult patients with NPS [16]. One patient with lateral OA without patellar dislocation underwent TKA without resurfacing of the patella. Two months after the surgery, the patient fell and ruptured the patellar tendon. The other patient with patellofemoral joint OA previously underwent several surgical treatments for patellar dislocation. The patient underwent a patellofemoral arthroplasty. Curbo et al. reported on TKA with proximal realignment for bicompartmental OA and patellar dislocation, resulting in the need for manipulation [17]. There were two case reports of unicompartmental arthroplasty without realignment of the extensor mechanism due to the absence of symptoms associated with patellar subluxation. In addition, several reports have demonstrated good clinical results for NPS with patellar aplasia in patients who underwent TKA [18–20]. In this case, the small patella was laterally fixed, and no impingement was observed. No realignment of the extensor mechanism was an option for the patients with NPS to maintain knee flexion.

There were several limitations. First, this is a case report, and TKA without patellar reduction could not be effective in all cases. However, it could be considered a treatment option. Second, this report was conducted 5 years after TKA. A further follow-up would be needed. Finally, the condition of the contralateral knee was thought to affect her gait performance. However, she was satisfied with her gait performance during follow-up and did not wish to have surgery on the contralateral side.

Summary

TKA without patellar reduction is a surgical treatment option for severe genu valgum and PDP. In addition, the small patella could be a key finding in the diagnosis of hereditary systemic diseases, including NPS.

Conflicts of interest

The authors declare there are no conflicts of interest.

For full disclosure statements refer to https://doi.org/10.1016/j.artd.2023.101099.

Informed patient consent

The author(s) confirm that written informed consent has been obtained from the involved patient(s) or if appropriate from the parent, guardian, power of attorney of the involved patient(s); and, they have given approval for this information to be published in this case report (series).

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