The Knee

Three-stage Total Knee Arthroplasty Combined with Deformity Correction and Leg Lengthening Using Taylor Spatial Frames and Conversion to Internal Fixation for Severe Intra- and Extra-articular Deformities and Hypoplasia in a Patient with Hemophilic Knee Arthropathy: A Case Report

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Declarations

Conflict of interest: The institution of KI received funding from Olympus Terumo Biomaterials Corp. (Tokyo, Japan).
Acknowledgements: We would like to thank Enago (www. enago. jp) for editing a draft of this manuscript.
Authors’ contribution: AF drafted the manuscript. KI completed the draft. EK, RH, SM, ZJ, MM, TO, NI, and TE reviewed the draft critically.
Funding: No funding was received for this study.

Abstract

Background: Hemophilic arthropathy is a cause of severe knee deformity, because chronic synovitis due to repeated hemarthrosis affects the area of the epiphyseal plates in juvenile cases. Total knee arthroplasty (TKA) is the standard treatment for end-stage knee arthropathy. However, it is difficult to perform one-stage TKA in patients with severe intra- and extra-articular deformities.

Case presentation: We reported a case of hemophilic arthropathy in a 55-year-old male with leg length discrepancy of 4 cm, limited range of knee motion (−40° extension and 85° flexion), intra-articular deformity (medial proximal tibial angle: 69°; mechanical lateral distal femoral angle: 79°), extra-articular deformity at the distal femoral metaphyseal (30° valgus and 45° flexion deformity), and varus malalignment (% mechanical axis: 33%). We planned a three-stage TKA. Firstly, we performed gradual correction and lengthening of the distal femur using Taylor spatial flame. Six months after surgery, we performed conversion surgery from external fixation to internal fixation. Finally, we performed TKA with rotating hinged type implant. Two years after surgery, physical examination showed a normal gait, leg length discrepancy of 2 cm (the right leg was shorter), improvement in the range of knee motion (0° extension and 100° flexion).

Conclusion: To the best of our knowledge, this presents the first combination of three-stage TKA with correction of femoral deformity and leg lengthening using a Taylor spatial frame and conversion to internal fixation in a patient with hemophilic knee arthropathy and severe intra- and extra-articular deformities.

Key Words

Hemophiliac knee arthropathy, Total knee arthroplasty, Femoral osteotomy, Taylor spatial frame, Knee

1. Introduction

Total knee arthroplasty (TKA) is the standard treatment for end-stage knee arthropathy. Hemophilic arthropathy is a cause of severe knee deformity, because chronic synovitis due to repeated hemarthrosis affects the area of the epiphyseal plates in juvenile cases. This process results in leg length discrepancies, angular deviations, and alteration of structures in the developing skeleton [1-3]. TKA is a standard procedure for end-stage hemophilic arthritis in the knee joint, which has been associated with favorable outcomes [4-6]. However, it is difficult to perform one-stage TKA in patients with severe intra- and extra-articular deformities [7-9].

To our knowledge, this is the first report of three-stage TKA combined with correction of femoral deformity and leg lengthening using the Taylor spatial frame (TSF; Smith and Nephew, Memphis, TN, USA) and conversion to internal fixation in a patient with hemophilic knee arthropathy. The patient presented with intra- and extra-articular deformities and hypoplasia in the distal femur and proximal tibia.

2. Case report

2.1. Patient information

A 55-year-old male was referred to our orthopedic department with complaints of right knee pain and low back pain due to the compensatory changes in the lumbar spine probably caused by a leg length discrepancy. He had a history of severe hemophilia A (factor VIII deficiency). He had suffered repeated intra-articular bleeding in knee joints, and the leg length discrepancy had progressed since childhood. He had undergone left TKA 10 years earlier. Physical examination showed a pelvic obliquity, fall limping gait, leg length discrepancy of 4 cm (the right leg was shorter), limited range of knee motion (−40° extension and 85° flexion in the right knee; −10° and 110° in the left knee), 2011 Knee Society Score of 31 points in the right knee, and Lysholm Knee Scoring Scale of 42 and 92 points in the right and left knees, respectively.

Radiographic examinations showed hypoplasia of the right femur and tibia, intra-articular deformity (medial proximal tibial angle: 69°; mechanical lateral distal femoral angle: 79°), extra-articular deformity at the distal femoral metaphyseal (30° valgus and 45° flexion deformity), loss of joint space at the tibiofemoral and patellofemoral joints (Arnold–Hilgartner classification stage V), and varus malalignment (% mechanical axis: 33%) (Fig. 1). The lengths of the femur and tibia were 371 mm and 356 mm on the right side, and 396 mm and 322 mm on the left side, respectively.

For the treatment of hemophilia, the patient received plasma-derived factor VIII concentrates (3,000 IU; 50 IU/kg) administered intravenously thrice weekly; the treatment was well controlled.

2.2. Preoperative plan

TKA was necessary for this case of end-stage osteoarthritis. Nevertheless, standard (one-stage, non-constrained) TKA was considered difficult because of the presence of severe intra- and extra-articular deformities. Severe intra-articular deformities often require semi-constrained or constrained TKA with stem extension because soft tissue imbalance could occur after bone resection. Severe extra-articular deformities require simultaneous TKA combined with osteotomy or staged-TKA combined with acute correction using osteotomy with internal fixation, or with acute or gradual correction using external fixators [7]. Taking into account the chief complaint of low-back pain, it was necessary to resolve the leg length discrepancy. Radiographic evaluation revealed that an extra-articular flexion deformity in the distal femur and hypoplasia of the right leg were responsible for the leg length discrepancy. Thus, we planned a three-stage TKA: gradual correction and lengthening of the distal femur using external fixators; conversion from external fixation to internal fixation; and constrained TKA with stem extension.

2.3. Operative procedure

2.3.1. First stage – gradual correction and lengthening of the distal femur using external fixators

Firstly, TSF was stabilized in the distal femur, and osteotomy was performed at the center of rotation of angulation. One week after surgery, we initiated correction using a 55-day computer-generated program, which extended the femur length by 20 mm, corrected the extra-articular deformity (varus deformity from 30° to 6°; flexion deformity from 45° to 7°), and led to a change in weight-bearing line of 51% (Fig. 2).

2.3.2. Second stage – conversion from external fixation to internal fixation

Six months after the first surgery, the presence of a callus was confirmed. Thereafter, we performed conversion surgery to internal fixation using a lateral distal femoral osteotomy plate. After removing the eternal fixator, we confirmed stability at the osteotomy site. Subsequently, the TriS® lateral distal femoral osteotomy plate (Olympus Terumo Biomaterials Corp., Tokyo, Japan) was applied to the osteotomy site with the iliac bone graft (Fig. 3). Finally, we released the contracture of the knee.

2.3.3. Third stage – TKA

Fifteen months after initial surgery, bone healing was confirmed. We performed TKA as the third stage of surgery. Considering the risk of fracture at the osteotomy site, it was considered necessary to maintain the plate fixation. We used three-dimensional (3D)-templating to plan the osteotomy, implant size, and alignment (Fig. 4). We selected the smallest size of the NexGen® Rotating Hinge Knee (Zimmer-Biomet, Warsaw IN, USA) because it was the only applicable implant for the severe hypoplastic medullary cavity of the distal femur and proximal tibia among semi-constrained and constrained implants. We used the portable accelerometer-based navigation (Knee Align2®, Zimmer-Biomet) for the planning of osteotomy [10]. We performed TKA with a medial parapatellar approach, quadriceps snip, and medial collateral ligament release. We changed the distal bi-cortical locking screws of the lateral plate to mono-cortical screws to prevent the occurrence of a fracture and interference with the implant placement (Fig. 5).

2.3.4. Perioperative anticoagulation management

The hematologist advised to administer coagulation factors perioperatively depending on the individual pharmacokinetic parameters obtained after a dose test administered before the surgery. Factor VIII concentrate was administered continuously aiming at a factor VIII level of at least 80%. Eighteen days after the first surgery, inhibitor antibodies against factor VIII were detected (maximum: 4BU/ml) and a bleeding tendency was observed. We initiated immune tolerance therapy (factor VIII: 4,500 IU thrice weekly); 2 weeks later, inhibitor antibodies were undetectable and the bleeding tendency had improved.

2.4. Clinical outcomes

Two years after surgery, physical examination showed a normal gait, leg length discrepancy of 2 cm (the right leg was shorter), improvement in the range of knee motion (0° extension and 100° flexion), no back pain, a 2011 Knee Society Score of 69 points in the right knee, and Lysholm Knee Scoring Scale of 93 points in the right knee. X-ray examination showed that there was no loosening of the implant. Moreover, there was no occurrence of complications, such as fracture or infection.

3. Discussion

We reported a case of a 55-year-old man with hemophilic knee arthropathy who presented with intra- and extra-articular deformities and hypoplasia in the distal femur and proximal tibia. Based on the clinical presentation, three-stage TKA combined with correction of the femoral deformity using the TSF and plate conversion was performed. Follow-up evaluation at 1 year after surgery showed good clinical outcomes.

Primary TKA for patients with extra-articular deformity should be limited to those who can be treated with intra-articular osteotomy and soft tissue balancing. Previous studies have reported that intra-articular correction can be achieved via normal surface replacement TKA if the extra-articular deformity is within 20° in the coronal plane on the femoral side or within 30° on the tibial side [8, 9]. Sculco et al. reported that one-stage TKA with corrective osteotomy or TKA with gradual correction is required for cases with greater deformity [10]. In the present case, the patient had hypoplasia of the femur and tibia, intra-articular deformity (69° of medial proximal tibial angle), extra-articular deformity at the distal femoral metaphyseal (30° valgus deformity, 45° flexion deformity), and a leg length discrepancy of 4 cm due to hypoplasia of the lower extremities and flexion deformity. As described at preoperative planning, normal TKA is not indicated for the treatment of such severe deformities. One-stage TKA with a corrective osteotomy may be a one-stage treatment option. However, this approach was considered inappropriate in the present case because the corrective closed osteotomy for coronal and sagittal plane deformities was difficult and could worsen the leg length discrepancy. Thus, we performed staged TKA combined with deformity correction and leg lengthening using an extra-fixator to correct the severe extra- and intra-articular deformities and leg length discrepancy.

If two-stage TKA after gradual correction using an extra-fixator is selected, it is necessary to wait for the formation of a callus and bone maturation. Considering the risk of infection associated with the long-term external fixation [7], TKA should be performed as early as possible. In this case, two-stage TKA was contraindicated for the following two reasons: (1) it was difficult to insert a stem across the correction site due to the hypoplasia; and (2) the expected period for sufficient bone maturation was longer than usual. Therefore, we performed conversion to internal fixation using a plate as the second stage of surgery. Thereafter, after achieving bone maturation, we performed TKA as the third stage of surgery. This approach was not linked to the development of a perioperative fracture or infection.

Previous studies reported the higher accuracy of 3D-templating for the selection of implant size compared with two-dimensional-templating [11, 12]. Our preoperative 3D-templating revealed only one applicable implant among several semi-constrained and constrained implants due to the severe hypoplastic medullary cavity of the distal femur and proximal tibia. Therefore, 3D-templating is necessary for the selection of implants in the treatment of severe hypoplastic medullary cavities and the planning of implant placement.

Although computed tomography-based navigated TKA or robotic-assisted TKA is useful for the treatment of cases with severe deformities, these systems require pin insertion around the knee joint. The occurrence of a fracture through a pin site has been previously reported [13, 14]. In this case, we thought that the risk of fracture through a pin site was high owing to hypoplasia and leg lengthening at the distal femur. A portable accelerometer-based navigation system does not require pin insertion and provides equivalent accuracy to that of a non-computed tomography-based navigation system [10]. Thus, we used a portable navigation system and performed an osteotomy.

The occurrence of bleeding and development of inhibitors, neutralizing alloantibodies against factors

VIII/IX, during surgery are disease-specific and serious problems commonly observed in patients with hemophilia [15, 16]. External fixation is linked to a higher risk of bleeding at the site of pin insertion versus internal fixation. Furthermore, the risk of bleeding during external fixation for leg lengthening is higher than that associated with other procedures involving arthrodesis, fracture [17], or joint contractures [18]. This is because the osteotomy site during leg lengthening is open to surrounding soft tissues. These higher risks may explain the lack of reports on the use of external fixators for leg lengthening in patients with hemophilia. In the present case, bleeding occurred around the pin site and blood testing detected inhibitor against factors VIII after the first stage of the operation. Fortunately, the inhibitor decreased to undetectable level after the administration of immune tolerance therapy. Appropriate monitoring of bleeding and coagulant activity, as well as intensive support by a hematologist, are necessary for performing surgery in patients with hemophilia [19, 20]. Nevertheless, the present case demonstrated that leg lengthening is a possible treatment option for the treatment of leg length discrepancies in patients with hemophilia.

In conclusion, three-stage TKA after correction of deformity and leg lengthening using external fixators and plate conversion may be a treatment option for patients with severe intra- and extra-articular deformities, tibiofemoral hypoplasia, and leg length discrepancy.

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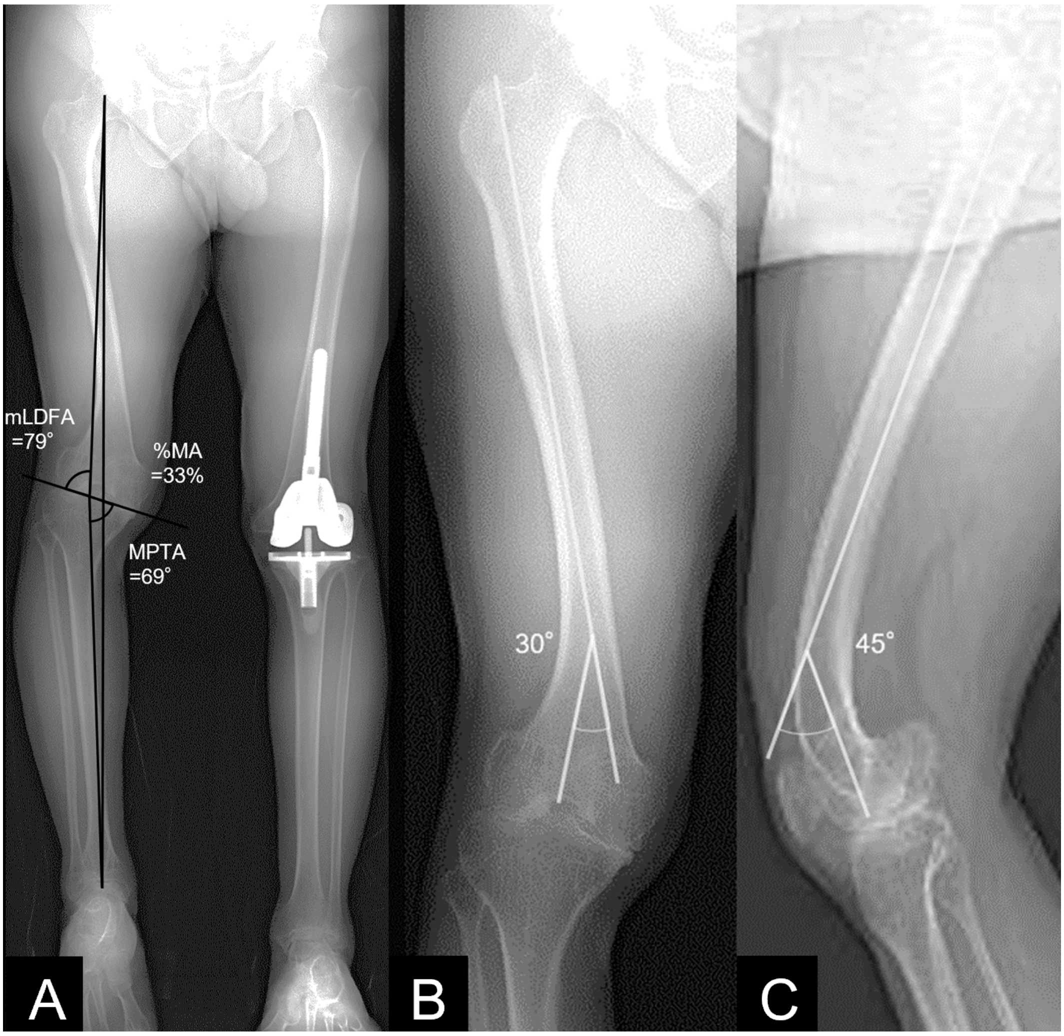

Figure 1.

(A) Anteroposterior whole-leg stanging, (B) anteroposterior thigh, and (C) lateral thigh radiographs showing hypoplasia of the right femur and tibia, intra-articular deformity (medial proximal tibial angle [MPTA]: 69°; mechanical lateral distal femoral angle [mLDFA]: 79°), varus malalignment (% mechanical axis [%MA]: 33%), extra-articular deformity at the distal femoral metaphyseal (30° valgus and 45° flexion deformity), and loss of joint space at the tibiofemoral joint.

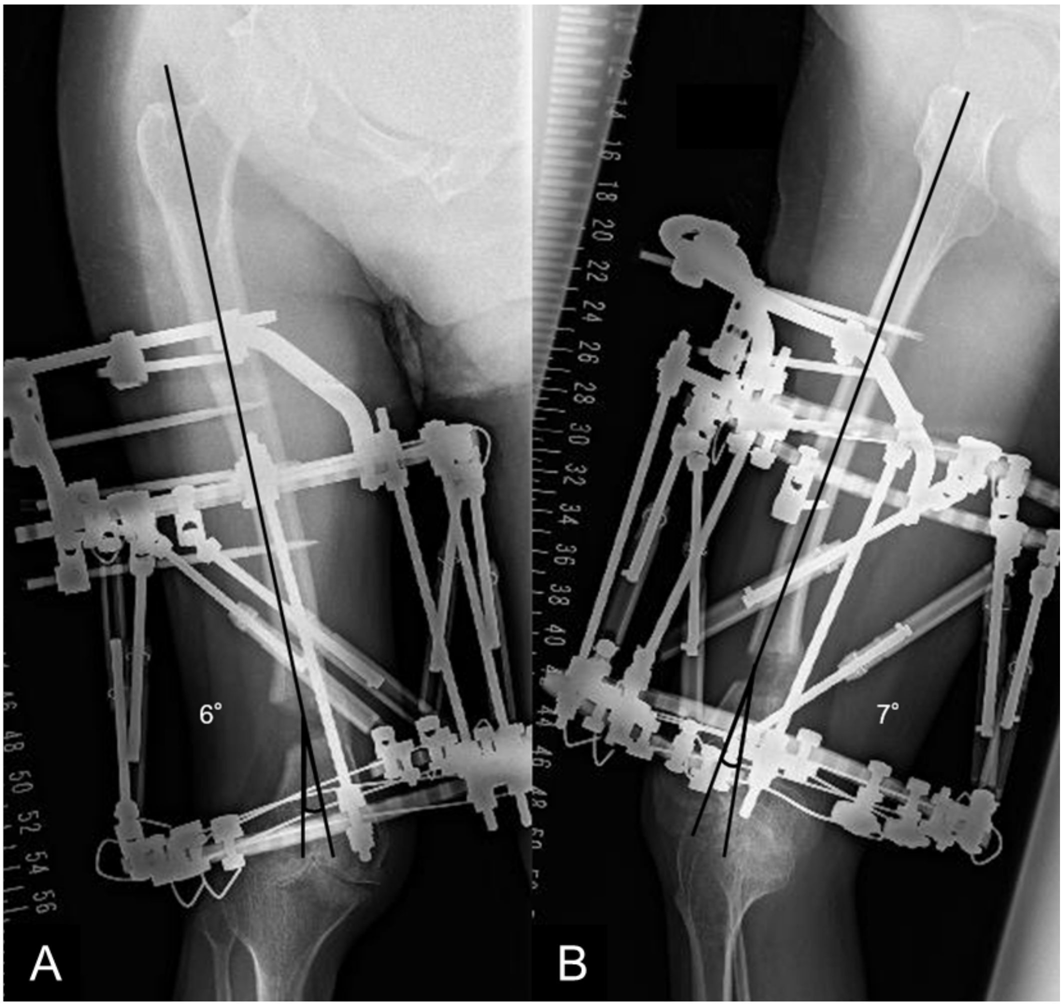

Figure 2.

(A) Anteroposterior thigh and (B) lateral thigh radiographs after the first stage of surgery (gradual correction and lengthening of the distal femur using external fixators) showing that the extra-articular deformity was corrected (varus deformity from 30° to 6°, and flexion deformity from 45° to 7°).

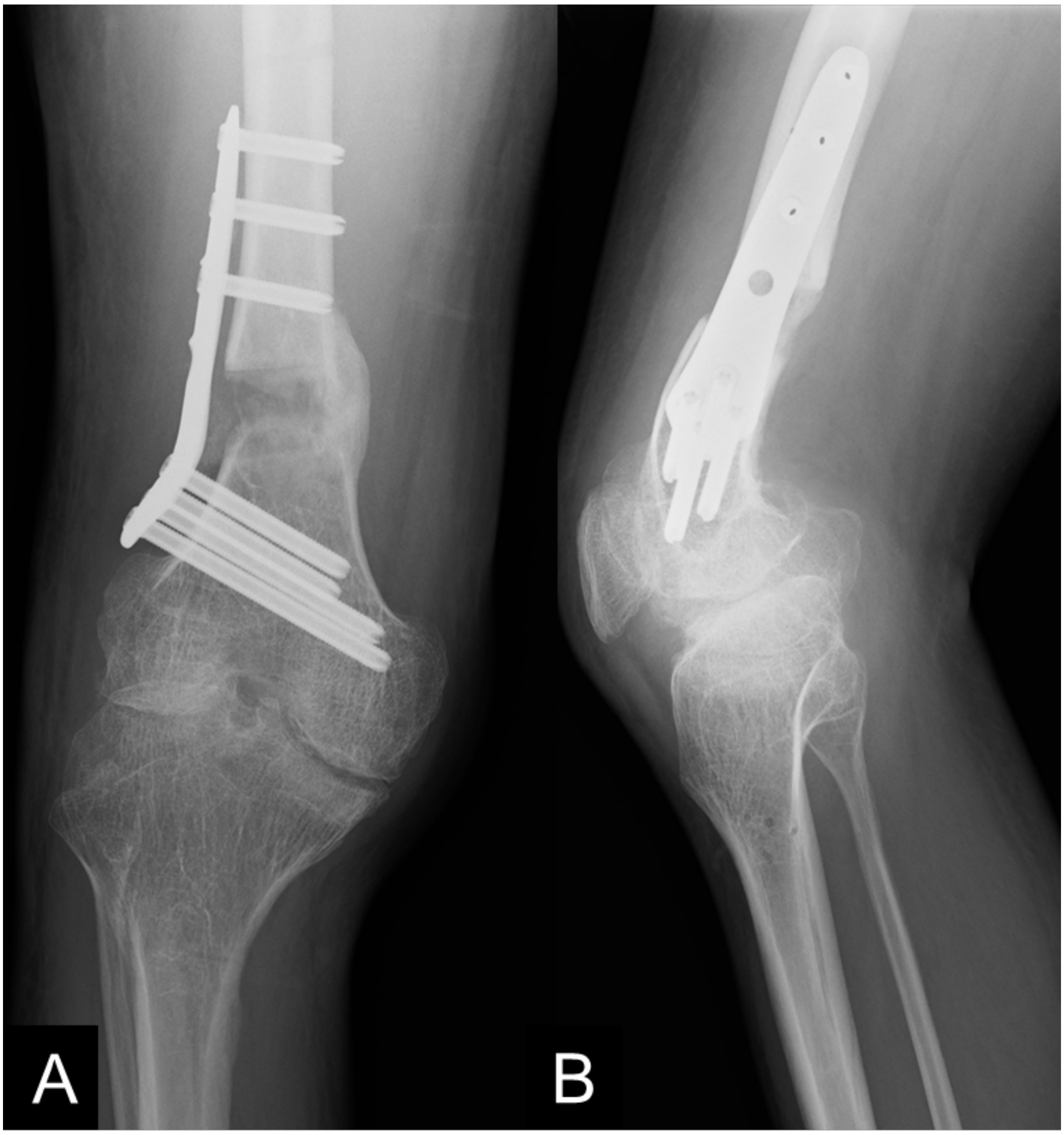

Figure 3.

(A) Anteroposterior knee and (B) lateral knee radiographs after conversion surgery to internal fixation with a plate as the second stage of surgery.

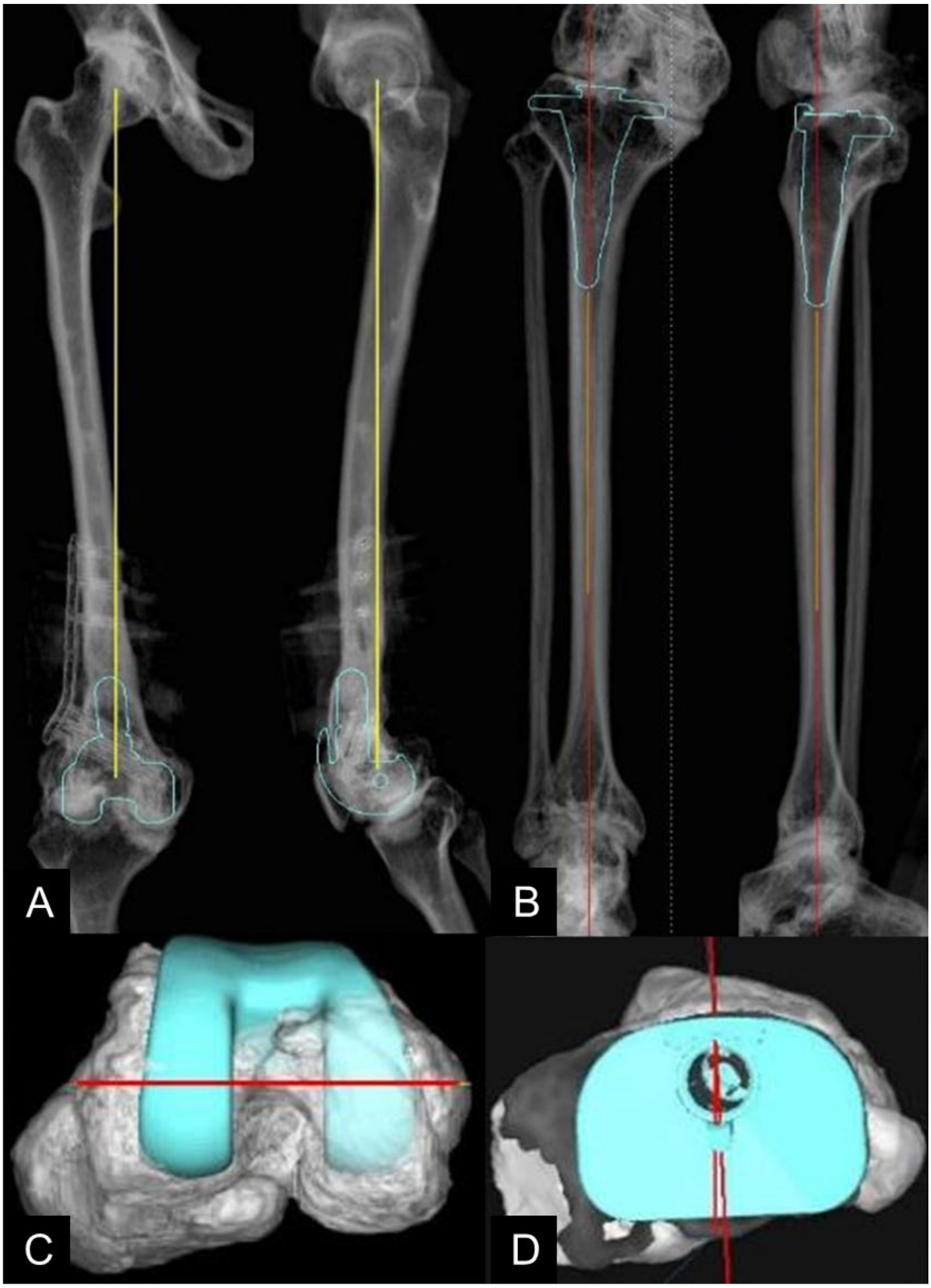

Figure 4.

(A–D) Three-dimensional-templating revealing that the smallest size of NexGen® Rotating Hinge Knee (Zimmer-Biomet, Warsaw IN, USA) was the only applicable implant among semi-constrained and constrained implants for the treatment of severe hypoplasia in the medullary cavity of the distal femur and proximal tibia, and that the tibial component should be installed at 3° varus. (C) and (D) show the amount of resection for the femoral posterior condyle and medial tibial plateau.

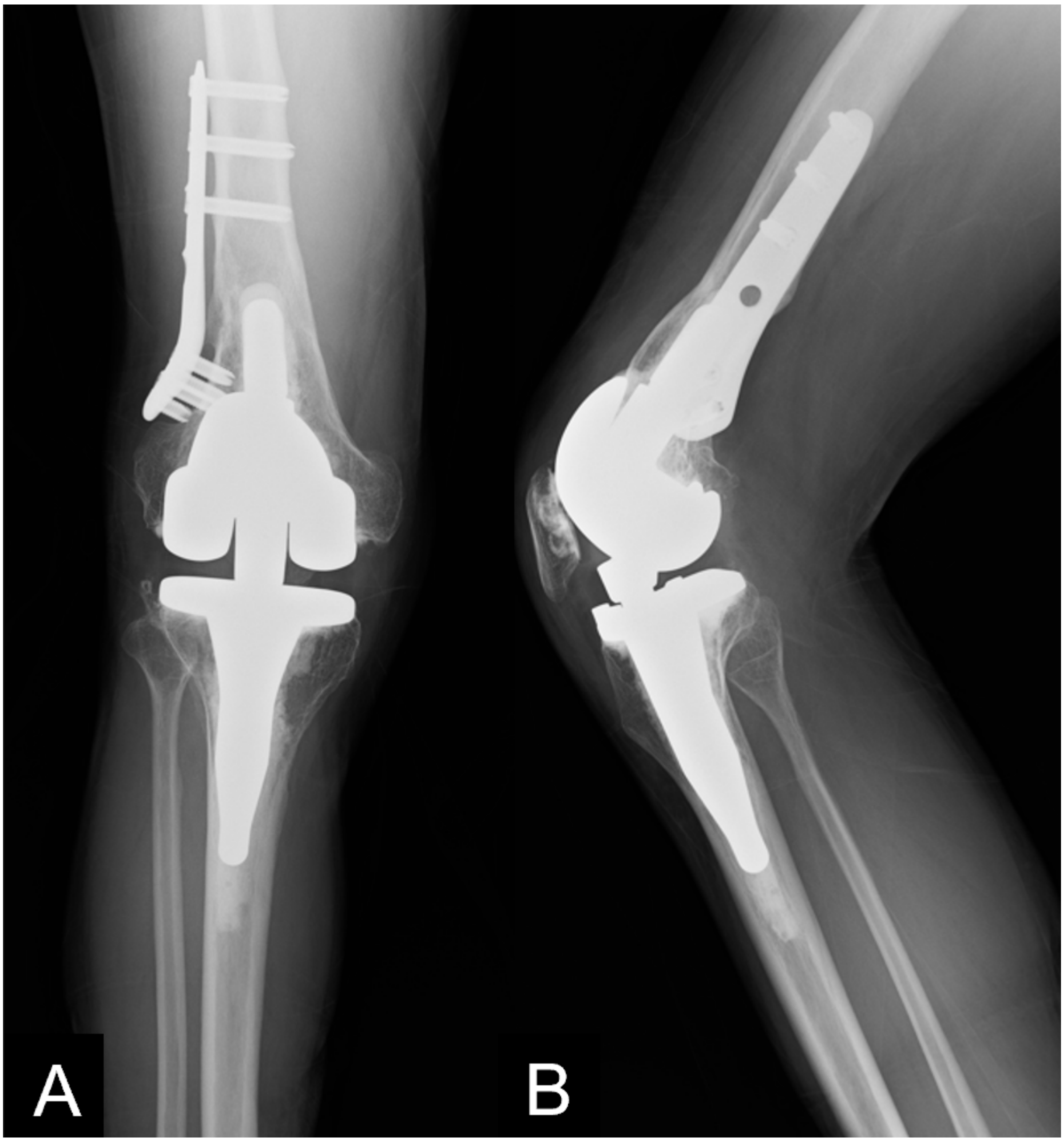

Figure 5.

(A) Anteroposterior knee and (B) lateral knee radiographs after total knee arthroplasty. We changed the distal bi-cortical locking screws of the lateral plate to mono-cortical screws.