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Case report

Staged total knee arthroplasty for bilateral complex knee deformities from Kashin–Beck disease and skeletal dysplasia

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ABSTRACT

This study reported two cases of patients with Grade III Kashin–Beck disease (KBD) with skeletal dysplasia concomitant with complex knee deformity and functional limitation treated by staged total knee arthroplasty (TKA). Detailed pre-operative planning, bone resection, and soft tissue balancing in affected knees were performed in the surgeries in this report. The results demonstrated that TKA could correct lower limb alignment, alleviate knee pain, improve function, and provide good quality of life in people with KBD. Surgical efficacy is still lower compared with treatment for osteoarthritis; contributing factors include weak muscle strength, severe deformity and unequal length of the lower limb, weak extensor apparatus of the knee, and patient-specific factors.

1. Introduction

Kashin–Beck disease (KBD) is a chronic endemic disease that has a high prevalence and morbidity in certain geographical regions of west and north China, Mongolia, Siberia and North Korea [1]. It commonly attacks the growth epiphyseal plates of multiple joints and articular cartilage, causing skeletal deformation and dwarfism in the elderly. The symptoms of KBD include symmetrical multiple arthritic pain and morning stiffness, muscle atrophy, shortened fingers, and enlarged and/or deformed joints with restricted range of motion (ROM) [2–4]. Criteria for diagnosis of KBD are typically based on a history of living in a KBD area; the aforementioned clinical symptoms; and abnormal radiographic changes in the metaphysis, epiphysis, distal phalanges of the hand, and the carpal bones [2,3]. The diagnostic criteria for grading are as follows: Grade I, multiple symmetrical joint enlargement of fingers or other limbs, usually accompanied by pain and slight muscular atrophy; Grade II, worse symptoms than Grade I, usually with significant enlargement of interphalangeal joints and shortened fingers; and Grade III, progression from Grade II, usually with severe symptoms of multiple joint deformity and significant dystrophic limbs or dwarfism [2,5].

Huang et al. reported a high incidence (>85%) of effect on knee joints in patients with KBD, because the knees move most frequently in daily life and are closely related to weight bearing [6]. Only a few studies have focused on surgical treatment for KBD patients. Ling et al. [7] and Liu et al. [8] have reported treating KBD with arthroscopy debridement and osteotomy, respectively; however, both operations could not reverse the severe articular cartilage degeneration in elderly KBD patients. In recent years, total knee arthroplasty (TKA) has been developed into a safe and highly successful alternative operation for treating

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patients with severe osteoarthritis (OA) [9]. Tang et al. reported using TKA to treat patients with mostly Grade I to II KBD [5]. However, to date, TKA has not been documented as an intervention for Grade III KBD and skeletal dysplasia.

The current report presents two Grade III KBD cases with skeletal dysplasia that had severe knee deformities and functional limitations. They were treated with staged TKA intervention with detailed pre-operative planning and individualized technical considerations. The lower limb alignment and knee function were significantly improved, and pain was effectively alleviated by the last follow-up.

2. Case reports

2.1. Case information

Patient A: a 45-year-old man (height: 125 cm, weight: 37 kg, BMI: 23.7) was referred for hospitalization; he complained of pain in both knees, deformation, and limited movement for four years (Figure 1). He came from an endemic area of KBD. Before he came for medical consultation, he had a limping gait and used supportive tools for walking, and took non-steroidal anti-inflammatory drugs that failed to relieve the pain. Physical examination showed short-trunk dwarfism with enlargement of both knees, ankles, and wrists, shortened fingers, and muscle atrophy around these joints. He had valgus deformity of his left knee, and varus deformity of the right knee. He had local tenderness over the medial and lateral knee joint lines and severe pain on weight bearing. The ranges of motion (ROMs) were 15 to 130° flexion in his left knee, and five to 130° flexion in the right knee. Both knees showed instability in the positive collateral ligament stress test and the positive anterior drawer test.

Standard anteroposterior (AP) and lateral radiographs demonstrated retarded growth of the femur and tibia, and enlarged subluxed knee joints with irregular articular space and surface. Full-leg standing radiographs (FLSR) including the whole pelvis showed that his pelvis tilted to the left, and his left leg was approximately six centimeters shorter than his right leg. The FLSR

Figure 1. Photograph (frontal view) of *Patient A* showing that he had enlargement of both knees, ankles, wrists, fingers, and elbows joints, with muscle atrophy around these joints. His pelvic tilted to the left and he had severe valgus of his left knee, and varus deformity of his right knee.

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