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CASE REPORT

Bipolar hip arthroplasty for subtrochanteric femoral nonunion in an adult with autosomal dominant osteopetrosis type II

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Introduction

Osteopetrosis or marble bone disease is a disease of osteoclasts that results in a failure of bone remodeling [1]. It is a generalized skeletal disorder characterized by osteosclerosis, obliteration of the medullary cavity and fragility of the bones [2]. This malignant sex-linked recessive condition is associated with childhood problems and early death. However, the autosomal dominant form is compatible with a normal life span, and as many as 40% of patients may remain asymptomatic [3, 4].

Autosomal dominant osteopetrosis is divided into two subtypes (I and II) [3]. Type I is characterized by increased thickness of the cranial vault, diffuse osteosclerosis of the lumbar spine and pelvis, and symmetrical, long-bone involvement. Type II shows more basal skull involvement, a 'Rugger-Jersey' spine and 'endobones' within the pelvis. Type II is important from an orthopedic perspective because it is associated with an increased rate of fractures, which are difficult to treat [2, 5].

There have been a small number of early reports on hip arthroplasty for hip osteoarthrosis or subtrochanteric femoral fractures including delayed union. However, osteopetrosis is a rare condition [6–9]. Therefore, orthopedic surgeons need to accumulate further case reports to establish an optimal therapeutic strategy for hip fractures in patients with osteopetrosis.

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The study protocol adhered to the ethics guidelines of the 1975 Declaration of Helsinki, and the study was approved by the institutional review board of Sawara Hospital. The patient was informed that this case study would be submitted for publication, and she gave her informed consent.

Case report

A 61-year-old female sustained a right subtrochanteric fracture caused by a traffic accident (Fig. 1). She had been diagnosed with osteopetrosis as a child, though she had never had problematic fractures. She underwent osteosynthesis of her subtrochanteric fracture with a compression lag screw plate at another hospital (Fig. 2). She developed right hip pain 9 months after the operation and could only walk on crutches in her house. She was referred to this hospital. She did not have any signs of infection. She was afebrile, and laboratory findings at the time of referral included a white blood cell count of 3,900/μl and C-reactive protein of 0.55 mg/dl. The alkaline phosphatase level was 239 U/l before the operation at our hospital, and the alkaline phosphatase levels were 528, 378 and 258 U/l at 1 week, 9 weeks and 1 year after the surgery, respectively. The normal range for the alkaline phosphatase level in this hospital laboratory is from 115 to 360 U/l. Radiographs showed subtrochanteric femoral nonunion, and the compression lag screw was cut out of her femoral head (Fig. 3).

There was thickening and sclerosis of the lumbar vertebral end plates (Rugger-Jersey spine), several concentric bands of sclerosis in the iliac wing (bone-within-bone appearance) and massive osteosclerosis at the base of her skull. These findings were typical characteristics of type II autosomal dominant osteopetrosis [10].

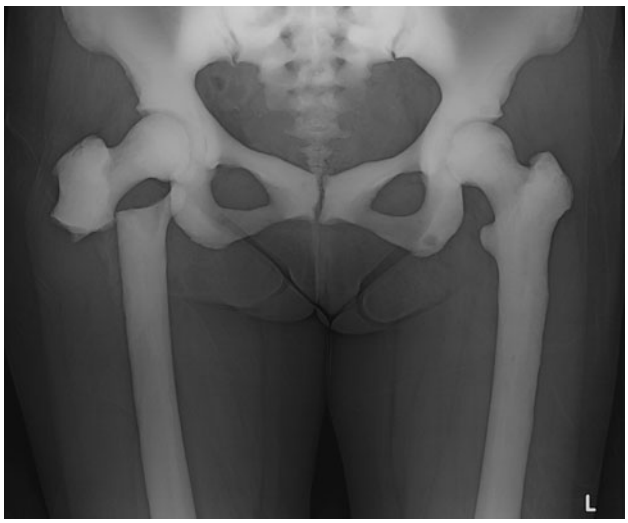

Fig. 1 Radiograph of the pelvis before the operation at another hospital shows a right subtrochanteric femoral fracture and diffuse osteosclerosis of the pelvis with a bone-within-bone appearance in the iliac wing. The medullary canal is obliterated in the femoral diaphysis

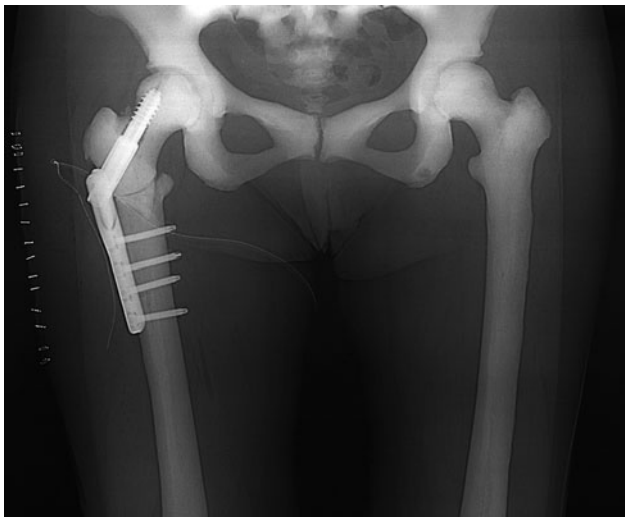

Fig. 2 Radiograph of the pelvis after the operation at another hospital shows a right subtrochanteric femoral fracture with a compression screw plate

She then underwent bipolar head arthroplasty after the removal of the compression screw plate.

A posterolateral approach was used. The compression screw plate was removed. The lag screw was cut out of the femoral head and neck, which revealed synovitis around the lag screw. There was nonunion of the subtrochanteric fracture and instability. The femoral neck was cut with a reciprocating saw. Preparation of the femoral canal was extremely difficult, because the marrow cavity was obliterated with bone that was dense, sclerotic and brittle. There was only the vestige of the removed lag screw and no cancellous bone in the proximal femoral canal.

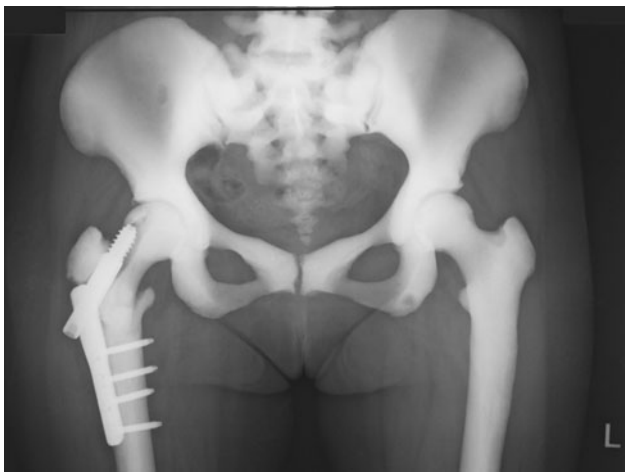

Fig. 3 Preoperative radiograph of the pelvis showing the right subtrochanteric femoral nonunion with a compression screw plate

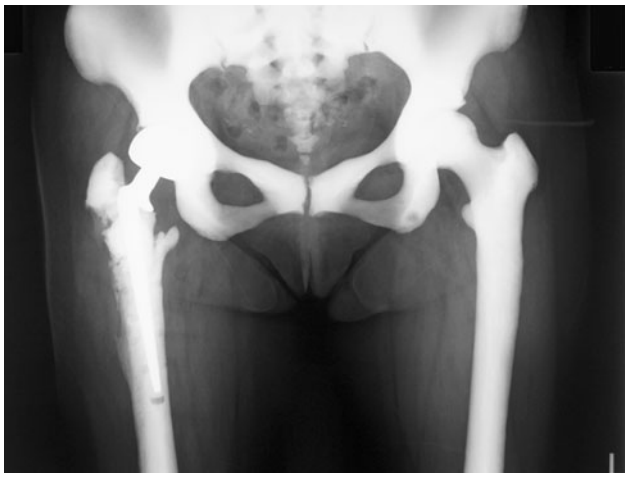

Fig. 4 Postoperative radiograph of the pelvis showing right bipolar head arthroplasty

Therefore, obliteration of the femoral medullary canal made it impossible to create a bony cavity for the stem using the standard surgical procedure. Therefore, a cannulated drill was used to create a bony cavity for the stem. While the K-wire was placed on the center of the femur, the position of the K-wire was checked by fluoroscopy with scrupulous attention. A canal was cut through the K-wire guide in the femur using a cannulated drill. Next, the canal was dilated using a high speed bar and power drill. This created a canal without any perforation or fracture of the cortical bone. Retrograde cementing was accomplished using a cement gun, and a standard sized stem was inserted (Fig. 4). The components of bipolar hip arthroplasty included the cement femoral component [PHS (7-MT) N straight femoral component, Kyocera, Kyoto, Japan] with a 22-mm zirconia ball and a Co–Cr metal outer head with polyethylene inlay.

Fig. 5 Two years after bipolar head arthroplasty, a radiograph showing no remarkable changes in comparison to the findings just after the operation

The patient's condition was satisfactory 2 years after the bipolar arthroplasty. The patient could walk without support, and had neither hip pain nor a limp and no discrepancy in leg length. The Japanese Orthopedic Association hip rating score (JOA hip score) increased from 23 to 76. The JOA hip score includes four categories and 100 points as full marks: pain (40 points), range of motion (20 points), walking (20 points) and activity of daily life (20 points).

A radiograph showed apparently solid fixation of the component with no osteolysis and no migration of the cup. Slight bone remodeling of the femur was observed on the latest X-ray image, with hypertrophic periosteal reaction at the distal screw hole and bone resorption at the stem shoulder (Fig. 5).

Discussion

Osteopetrosis is caused by defective bone resorption possibly related to dysfunctioning osteoclasts [1]. Fractures occur frequently in patients with osteopetrosis, particularly in patients with the autosomal dominant form [11]. The treatment of these fractures has been described in the literature mostly in the form of case reports and small case series [4, 9, 12, 13]. Surgical treatment of osteopetrotic fractures is typically reported to be difficult and associated with many complications: the difficulties associated with surgical treatment include the extreme hardness of the bone, which impedes drilling and cutting, hardware failure, peri-prosthetic fractures, delayed union, pseudarthrosis, re-fracture and peri-prosthetic infection [4, 12–15]. Histologically, mature osteopetrosis fracture callus contains no Haversian organization and has a paucity of osteoclasts [14]. Fracture healing has been studied histologically with specimens from up to 1 year post-fracture [14]. Altered local biomechanics caused by hardware could create problems like peri-prosthetic fracture and re-fracture though screw holes after the removal of an implant [15]. Greene and Tore [16] reported that the pin tracks were still evident on radiographs 2.5 years after pin removal. Birmingham and Mchale [9] accumulated and analyzed past reports and reported that the average time to union is approximately 26 weeks.

Surgical fixation is the recommended treatment for a femoral neck fracture in a child with osteopetrosis [4, 16, 17]. Armstrong et al. [4] recommend surgical fixation combined with a valgus osteotomy as the primary treatment for femoral neck fracture with coxa vara of osteopetrosis. However, there have been few reports describing the treatment for femoral neck fractures in adults with osteopetrosis [18, 19]. Rolauffs et al. treated a 40-year-old male with screws, and 3 of the 4 screws broke. Finally, the patient underwent resection arthroplasty due to osteomyelitis [18]. Schoierer and Hoffmann [19] reported total hip arthroplasty after the failure of osteosynthesis for femoral neck fracture in a 36-year-old male.

Surgical fixation is associated with considerable difficulty and complications in cases of intertrochanteric fracture of osteopetrosis [4, 12–15, 18].

Birmingham and Mchale [9] reported a high complication rate in a cohort series, and the rate of hardware failure is 29%, the rate of reoperation is 29% and the incidence of peri-prosthetic fracture is 14%. On the other hand, there have been few reports of good results in surgical treatment for intertrochanteric femoral fractures [4, 15].

The current patient demonstrated femoral nonunion after surgical fixation using a compression screw and plate, and the compression screw was cut out of the femur head and neck. Fortunately, no breakage of the implants was seen, and there was no sign of infection.

There have been few reports of primary total or bipolar hip arthroplasty for the femur neck, intertrochanteric or subtrochanteric fractures in patients with osteopetrosis [9]. Ashby [12] reported three total hip arthroplasties. Two patients underwent total hip arthroplasty for subtrochanteric femoral nonunion, and one underwent the procedure primarily for a fresh subtrochanteric femoral fracture. One patient showed an incomplete peri-prosthetic fracture at the tip of the femoral stem 3 years later; this healed following 10 months of conservative treatment.

Osteoarthrosis of the hip often occurs in mid-adult life in patients with autosomal dominant osteopetrosis type II. It is thought that the articular cartilage is crushed between the hard subchondral bone of the femoral head and the acetabulum [2]. Advanced hip osteoarthritis sometimes requires arthroplasty. There are several case reports of total

hip arthroplasty for hip osteoarthritis in a patient with osteopetrosis [2, 6, 8, 12, 20]. Most reports emphasize technical difficulties especially on the femoral side, as in the current case. Drills and high-speed burrs, guided by fluoroscopy or navigation, have been used to help create a medullary canal. Some surgeons perform hybrid total hip arthroplasty because of the difficulty in creating a precise femoral cavity and the brittle nature of the osteopetrotic bone [6–8]. Jones et al. [7] reported uncemented total hip arthroplasty using an expansion cup to reduce the risk of an acetabular fracture and a Wagner cone prosthesis to avoid poor cement-bone penetration in the dense sclerotic bone. However, their patient had autosomal dominant osteopetrosis type I. The marrow is filled with bone that is dense and sclerotic, and has a brittle cavity in autosomal dominant osteopetrosis type II, as in the current case [6, 8]. The use of a cementless stem requires creating a precise femoral cavity, and limited contact between the cementless stem and bone may prevent bone ingrowth.

A high speed bar and power drill used under fluoroscopy were used in the current patient to create sufficient space on the femoral side for insertion of the femoral component.

Close attention was paid to avoid the influence of cement from the screw holes. The pressure and speed of the cementing technique were curbed.

In general, total hip arthroplasty is recommended for femoral neck fracture [21], and patients with autosomal dominant osteopetrosis have a full life expectancy [2]. However, there are also some difficulties associated with fixing the acetabular cup of the pelvis if we use the total hip arthroplasty system because of the femoral prosthesis in the patients with autosomal dominant osteopetrosis type II. A bipolar head prosthesis was used on the acetabular side because the articular cartilage of the acetabular was intact. Unfortunately, her normal articular cartilage may become crushed between the hard pelvic subchondral bone and bipolar head prosthesis, and further long-term follow-up study is thus needed.

Conclusions

The short-term 2-year follow-up indicated that the clinical and radiographical results are good. Bipolar head arthroplasty is a useful option to salvage a subtrochanteric femoral nonunion in patients with osteopetrosis. Case reports of other successful treatments are needed in the future to help reach a consensus for the optimal treatment of such patients.

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