Osteopetrosis and total hip arthroplasty

Report of two cases

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Summary. *Two patients with osteopetrosis and osteoarthritis of the hip underwent "hybrid" total hip replacement. There are technical difficulties in seating acetabular and femoral components on hard and brittle osteopetrotic bone with an obliterated marrow cavity. Although the surgical procedures are challenging, this form of arthroplasty is recommended for hip osteoarthritis which is associated with osteopetrosis.*

Résumé. *Nous présentons deux cas d'arthrose de la hanche doublée d'ostéomarmoréose (maladie d'Albers-Schonberg), qui ont été traitées avec une prothèse totale «hybride» de l'ensemble de la hanche. Nous discutons des difficultés techniques de la répartition des composants acétabulaires et fémoraux sur des os de marbre durs et fragiles dont la cavité médullaire est oblitérée. Malgré la difficulté de l'intervention chirurgicale, l'arthroplastie «hybride» de l'ensemble de la hanche est recommandée dans le traitement de l'arthrose de la hanche doublée d'ostéomarmoréose.*

Introduction

Osteopetrosis (Albers-Schönberg disease [1] or marble bone disease) can be divided into three clinical forms; severe infantile autosomal re-

cessive, intermediate autosomal recessive and mild autosomal dominant. The dominant form does not affect life expectancy but may lead to orthopaedic problems [2, 4, 5]. Plating and intramedullary rodding are undertaken for fractures and bone deformities, and joint arthroplasty is carried out for degenerative joints, but these operations are sometimes extremely difficult because of bone fragility and obliteration of the marrow cavity [3, 4, 6].

We have undertaken total hip arthroplasty in two patients with osteoarthritis of the hip associated with osteopetrosis. The operation poses considerable technical difficulties.

Case reports

Case 1

The patient was a 16-year-old girl in whom bilateral congenital dislocations of the hip had been treated with a Pavlik harness. She began to experience pain in both hips and was diagnosed as having the autosomal dominant type of osteopetrosis when she was 13 years of age. At this time, roentgenograms revealed subluxation and osteoarthritis in both hip joints. Her pain increased and when she was aged 16 years she was referred to the Hokkaido University Hospital with a severe antalgic gait and marked decrease in the range of motion in both hips. Her preoperative Harris' hip score was 25 points (gait 9; pain 10).

Radiographic examination revealed the typical features of osteopetrosis and both hip joints showed marked degenerative changes (Fig. 1A). She then underwent total arthroplasty on her left hip and subsequently on the right.

At operation, a postero-lateral approach was used. The femoral neck was cut with an air-pressure oscillating saw with considerable difficulty because of the dense sclerotic bone. The articular cartilage of the femoral head and acetabulum showed marked degenerative changes. Acetabular reaming was carried out and a same-size Harris-Galante porous cup was securely seated with 3 screws. Preparation of the femoral canal was extremely difficult because of obliteration of the

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Fig. 1. **A** (Case 1) Preoperative radiograph of the pelvis showing increased density and concentric sclerotic rings in the ilium. Osteoarthritis of both hips is prominent. **B** (Case 1) Postoperative radiograph of the pelvis showing hybrid total hip arthroplasties

marrow cavity with bone which was dense, sclerotic and brittle. There was no cancellous bone in the proximal femoral canal. Due to the hardness of the bone it was not possible to use a hand reamer or drill, and a high-speed bur under an image intensifier was therefore employed for preparation of the canal. This procedure for acceptance of a Harris Precoat CDH (Zimmer) stem was completed without any perforation or fracture of the cortical bone. A medium sized femoral head was chosen. Retrograde cementing was accomplished using a cement gun and a CDH (100 mm) stem was inserted (Fig. 1 B). Recovery was uncomplicated and 4 weeks after the operation the patient started to walk with partial weight bearing; full weight bearing was permitted at 6 weeks.

Five months after the operation on her left hip, total hip arthroplasty was performed on the right side. The same procedures were followed as during the first operation using an HGP socket (Zimmer) and a Harris Precoat CDH stem (Zimmer). Post-operative radiographs showed leaking of bone cement on the medial side of the femoral shaft but the positions of the acetabular and femoral components were good (Fig. 1 B).

Six years after the operation, the patient's condition was satisfactory. She had neither pain nor a limp, and could achieve a 90-degree arc of flexion in both hips. Her recent Harris' hip score was 84 points (gait 27; pain 40).

Fig. 2. **A** (Case 2) Preoperative radiograph of the pelvis showing increased density of the pelvic bones and the femora. Coxa vara deformity of the left femoral neck and osteoarthrotic changes in the left hip joint are evident. **B** (Case 2) Post-operative radiograph of the pelvis showing hybrid total hip arthroplasty

Case 2

A male aged 50 years had a history of mild pain in his left hip of 6 years duration. A diagnosis of benign autosomal dominant osteopetrosis had been made when he was aged 6. He started to experience severe pain in the left hip during walking in October 1994, a year before his first visit to Hokkaido University Hospital. He was then found to have a marked antalgic gait and a moderate decrease in the range of motion of his left hip. His right hip had a normal range of movement without evidence of degenerative changes. His preoperative Harris' hip score was 54 points (gait 18; pain 20).

Radiographic examination revealed that his left acetabulum was shallow with severe varus deformity of the femoral head (Fig. 2A). In July 1995, the patient underwent total arthroplasty of the left hip. At operation, difficulties in cutting the femoral head and preparing the acetabulum for fixing the socket with screws were similar to those encountered in the previous case. Preparation of the femoral canal was much more difficult because it was completely obliterated by dense, sclerotic bone. With extreme care, reaming to the size needed for the CDH femoral component was successfully accomplished using a high-speed bur, without any perforation of the cortical bone.

A medium-plus size femoral head was chosen. Retrograde cementing was undertaken using a cement gun and the CDH (100 mm) (Zimmer) stem was inserted (Fig. 2B). The patient was allowed to walk with partial weight-bearing 4 weeks after the operation, and he progressed to full weight-bearing 8 weeks postoperatively.

One year after surgery, the patient was doing well and had no signs of postoperative infection. His left hip was painless with a arc of flexion of 90 degrees and his Harris' hip score was 84 points (gait 23; pain 40).

Discussion

Osteoarthritis of the hip and knee is not uncommon in midadult life in patients with the autosomal dominant form of osteopetrosis. It has been suggested that the increased density of subchondral bone in this disorder may be responsible for the early onset of degenerative changes [4, 5, 6]. One of our patients (Case 1) had a history of treatment for congenital dislocation of both hips, and the other (Case 2) had marked varus deformity of the left femoral neck. These abnormalities seemed to contribute to the early onset of osteoarthritis in both patients.

Janecki and Nelson [5] and Cameron and Dewar [3] reported technical difficulties in total hip arthroplasty in patients with osteopetrosis and we experienced similar problems. Preparation of the

femoral canal is extremely challenging because of the hard, brittle bone and obliteration of the marrow canal. In order to avoid fractures, power saws rather than chisels are recommended for cutting the femoral neck. On the acetabular side, a cementless socket is useful because of the difficulties in finding good-quality cancellous bone for effective bone-cement permeation. On the femoral side, a power drill and high-speed burr are recommended for creating sufficient space for insertion of the femoral component, as a regular rasping procedure of the femoral canal is impossible. A cemented femoral component is appropriate because press-fitting the cementless femoral component onto sclerotic, brittle bone is difficult and may cause a fracture of the femoral shaft. A narrow, short femoral component is preferred because of the difficulty in creating a long, wide space in the femoral canal for seating a regular-size prosthesis. Cameron and Dewar [3] reported that they were forced to shorten the femoral component in order to seat the prosthesis in the shaft on both sides of bilateral total hip arthroplasties; we prefer to use a CDH type stem (Zimmer) fixed with cement because it is straight, narrow, and only 100 mm in length. Because of the diminished vascularity of bone marrow and impaired white cell function in osteopetrosis, perioperative antibiotics are indicated for prevention of postoperative infection.

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