Total Hip Arthroplasty with a Revision Stem in Hereditary Multiple Exostoses with Secondary Osteoarthritis

Maximiliano E. Negri, María Belén López Villagra, Guillermo J. Allende
Department of Hip and Knee Joint Reconstructive Surgery, Sanatorio Allende, Córdoba, Argentina.

ABSTRACT
We present a 42-year-old woman with a history of hereditary multiple exostoses (HME), with pain and limited range of motion of the left hip. Radiographic studies showed osteoarthritis added to femoral exostosis with bilateral hip dysplasia and femoral head subluxation. Total left hip replacement was performed using a modular uncemented implant with excellent postoperative results at 7 years of follow-up.

Keywords: Hereditary multiple exostoses; hip joint; osteoarthritis; replacement arthroplasty.

Level of Evidence: IV

INTRODUCTION
Osteochondromas are primary benign osteocartilaginous bone tumors that are typically located around the knee, proximal humerus, and other endochondral ossification bones, and represent the most common primary bone tumor. They can appear as a solitary lesion or as multiple lesions in the context of hereditary multiple exostoses (HME), an autosomal dominant disease caused by a mutation in the tumor suppressor EXT gene family. Their prevalence is one case per 50,000 inhabitants in the general population. 30-90% of patients with HME may have proximal femur osteochondroma, while pelvic osteochondroma affects 15-64% of patients. In 25% of cases, acetabular dysplasia and coxa valga are detected. This has suggested that valgus hip morphology and femoral neck osteochondroma may contribute, independently or synergistically, to the increased risk of lateral hip subluxation and, consequently, to osteoarthritis. Total hip arthroplasty is a valid therapeutic option when this condition is diagnosed. Good preoperative planning must be carried out given the complexity of the joint deformity and the alteration of intraoperative parameters, in order to achieve correct placement of the prosthetic components.

The aim of this article is to communicate an alternative reconstruction option for complex deformities.
CLINICAL CASE
A 42-year-old woman with a family history of HME in her paternal line who consulted, for the first time, for left coxalgia of two years of evolution. He experienced severe pain during weight bearing and limitations on his daily activities. Hip range of motion was 120º of flexion, 30º of abduction, and limited rotations. The Harris hip score was 75. On the pelvis radiograph, bilateral hip dysplasia with subluxation of the femoral head and bony protrusions (exostoses) in both lesser trochanters toward the femoral head were observed (Figure 1). The left hip had a cervico-diaphyseal angle of 165º, acetabular incongruity, and signs of osteoarthritis that were more evident on the CT scan (Figure 2). On radiographs of the knees and ankles, multiple exostoses and tibiofibular ankylosis were visualized (Figure 3).

Figure 1. Panoramic pelvic radiograph. Deformity of both hips is observed, with signs of osteoarthritis in the left hip.
Figure 2. A. Preoperative CT scan of both hips, coronal section, to assess bone stock. B. Three-dimensional reconstruction.
Surgical technique

An uncemented total hip arthroplasty through a posterior Kocher-Langenbeck approach was chosen (Figure 4). A modular cementless S-ROM® implant (DePuy, Johnson & Johnson, Warsaw, IN, USA) with a 28 mm diameter ceramic head and a proximal metaphyseal module was used to allow intraoperative control of the deformity.

RESULTS

There were no intraoperative or immediate postoperative complications. Seven years later, the patient remained symptom-free, with 120° flexion, 40° abduction, 15° external rotation, 20° internal rotation and a Harris hip score of 100. The radiographic appearance of the implant was also satisfactory (Figure 5).

Figure 3. Anteroposterior and lateral radiographs of knees and ankles. Note the multiple exostoses and tibiofibular ankylosis.
Figure 4. Intraoperative image. The dislocation of the femoral head and its irregular contour corresponding to the voluminous cartilage exostosis is observed.

Figure 5. Anteroposterior radiograph of the left hip, 7 years after total hip arthroplasty.
DISCUSSION

HME is an autosomal dominant disease. It tends to be more common in men than in women, and usually affects the femur, tibia with distal and proximal fibula, distal humerus, distal radius and distal ulna. Hip osteoarthritis due to HME is at increased risk of progression because the femoral neck is thickened by the medial exostosis, which may facilitate subluxation of the femoral head. Shapiro et al. reported that eight of 32 patients (64 hips) <20 years with this disease had coxa valga, as did our patient. Increased femoral anteversion has also been reported.

With regard to the surgical treatment of exostoses, there are many reports on the methods used in pediatric patients, which have achieved good outcomes, but there are few reports on the treatment of osteoarthritis in advanced stages in adults. Porter et al. described the hips of 12 patients with HME and reported that only one had undergone total hip arthroplasty, without specifying the evolution.

Moran et al. reported on the placement of a bilateral prosthesis in two patients with hip dysplasia secondary to HME and reported good functional outcomes after two years. In these cases, they used modular implants and performed a femoral osteotomy due to the complexity of the technique.

Vaishya et al. presented a 27-year-old patient with HME and osteoarthritis of both hips and a large femoral and acetabular bone deformity. The patient underwent bilateral total hip arthroplasty using a primary implant. The prosthesis dislocated, therefore the stem was replaced to correct its anteversion and a collar with a higher offset was added. This demonstrates the complexity of this type of deformity and the need to use unconventional implants.

Kim et al. published a case of HME with secondary osteoarthritis treated with total hip arthroplasty using a cementless Wagner® cone stem (Zimmer. Warsaw, IN, USA); the outcome was good and the Harris hip score improved from 35 to 82 in the 2-year follow-up.

In our case, due to the underlying metaphyseal alteration and the increased femoral anteversion, we used a cementless diaphyseal fixation stem with a modular system and a proximal femoral sleeve to control the anteversion as a very good option for the correct positioning of the components, thus achieving the desired stability.

CONCLUSIONS

In patients with HME, deformity of the proximal femur is frequent and facilitates progression to osteoarthritis. Modular cementless total hip arthroplasty is a good option for restoring hip biomechanics. However, it requires proper preoperative planning, which should include a CT scan to assess hip anteversion and valgus.

Conflict of interest: The authors declare no conflicts of interest.

REFERENCES


