Acetabular dysplasia associated with hereditary multiple exostoses

A CASE REPORT

N. A. Felix, J. M. Mazur, E. A. Loveless

From the Nemours Children's Clinic, Jacksonville, USA

Hereditary multiple exostoses is an autosomal dominant disorder characterised by multiple osteochondromata, most commonly affecting the forearm, knee and ankle. Osteochondromata of the proximal femur have been reported to occur in 30% to 90% of affected patients with coxa valga in 25%. Acetabular dysplasia is rare but has been described. This is the first report of a patient requiring surgical intervention.

A girl was seen at the age of nine with hereditary multiple exostoses and when 12 developed bilateral pain in the groin. Radiographs showed severely dysplastic acetabula with less than 50% coverage of the femoral heads and widening of the medial joint space. Large sessile osteochondromata were present along the medial side of the femoral neck proximal to the lesser trochanter, with associated coxa valga.

The case illustrates the importance of obtaining initial skeletal surveys in children with hereditary multiple exostoses to identify potential problems such as acetabular dysplasia and subluxation of the hip.

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Hereditary multiple exostoses is an autosomal dominant disorder with an estimated prevalence of 1 in 50 000.¹ It is characterised by multiple osteochondromata found throughout the skeleton, primarily involving the long bones, the iliac crests, the scapulae and the ribs. The lesions are seen most commonly in the forearm, knee, and ankle.² Deformities at these sites frequently require surgery. The femoral neck is often broad with medial osteochondromata and coxa valga may be present.³ The femoral head may be

partially uncovered, but severe acetabular dysplasia is unusual. We describe a patient with hereditary multiple exostoses with bilateral coxa valga and acetabular dysplasia who required surgical treatment.

Case report

A girl with multiple osteochondromata was first seen at the age of nine years. There was a family history of multiple exostoses affecting her father and a paternal aunt.

Multiple lesions were present bilaterally in the proximal humerus and forearm. The right forearm was shortened with a dislocated radial head and pronation was limited to 20°. Both hands were affected, with shortening of the metacarpals of the left middle finger and of the right ring and little fingers.

Osteochondromata were present on the medial side of the femoral neck, bilaterally. The hips were subluxated with bilateral acetabular dysplasia. The centre-edge angle was -10° on the right and +10° on the left.

The acetabular angle was 40° on the right and 27° on the left. Osteochondromata were present in both knees but a good range of movement was maintained. There was some deformity of the ankles with palpable osteochondromata on the distal tibiae. There was an excellent pain-free range of ankle movement. She had mildly bilateral painful hallux valgus.

The radiological appearance of the hips was a cause for concern, but as she had no pain close observation was recommended. The possibility of surgical correction in the future was discussed.

The patient was seen again at the age of 12 years when she was referred for evaluation of scoliosis. At this visit, she complained of bilateral pain in the groin which was constant when walking, and aggravated by climbing stairs. She was unable to walk for long distances or participate in sports. She could undertake normal daily activities and was not taking medication.

On physical examination, she walked with a normal gait. She had a full range of movement of her spine with no palpable osteochondromata. Hip movements were equal, but limited bilaterally with extension of 0° and flexion to 90°. With the hips in extension, internal rotation was 30° and external rotation 45°. When the hip was flexed to 90°

N. A. Felix, MD, Fellow
Eastwood Orthopaedic Clinic, 99 Remuera Road, Auckland 5, New Zealand.

J. M. Mazur, MD, Orthopaedic Surgeon
E. A. Loveless, MD, Orthopaedic Surgeon
Department of Orthopaedics, Nemours Children's Clinic, 807 Nira Street, Jacksonville, Florida 32207, USA.

Correspondence should be sent to Dr J. M. Mazur.

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internal rotation was limited to 5° and external rotation to 10°. She had pain at the extremes of movement, especially on the right side. She had full motor power, except for weakness of the iliopsoas (MRC grade 3+) on the left side. Sensation was intact and deep tendon reflexes were bilaterally symmetrical.

Anteroposterior (AP) and lateral radiographs showed a right thoracic curve from T6 to L1 of 32° and a left lumbar curve from L1 to L4 of 20°. Radiographs of the hip showed severely dysplastic acetabula with less than 50% coverage of the femoral heads (Fig. 1). There was apparent widening of the medial joint space. Large sessile osteochondromata were present along the medial side of the femoral neck proximal to the lesser trochanter, and there was an associated valgus deformity. CT and three-dimensional reconstruction showed multiple osteochondromata of the pelvis, but none involving the acetabula (Fig. 2). The large osteochondromata present on the medial side of the femoral necks appeared to be causing lateral displacement of the heads.

Surgical treatment was recommended in view of her symptoms and the severity of the acetabular dysplasia. She initially underwent a proximal femoral osteotomy and a Steele pelvic osteotomy on the right side. A varus derotation osteotomy of the proximal femur was carried out using a posterior approach. A bone wedge was removed from the proximal femur including much of the osteochondroma on the medial side. The osteotomy was fixed with a 50 mm 90° blade plate. The patient was then placed in a supine position and the pelvic osteotomy carried out. Through an adductor approach, osteotomies of the pubic and ischial rami were undertaken. Osteotomy of the ilium was carried out in the standard Salter fashion using a bone wedge from the iliac crest. Fixation was obtained using a 6.5 mm

cortical screw and a Steinmann pin. Postoperatively, the patient remained non-weight-bearing for one month followed by partial weight-bearing with crutches.

Two months after surgical treatment on the right side she had similar procedures on the left (Fig. 3). A varus derotation osteotomy of the proximal femur was carried out with removal of a large portion of the osteochondroma and application of a 50 mm blade plate. A Steele triple osteotomy was performed and fixed with two 6.5 mm cortical screws, one in the anterior and one in the posterior column. She was immobilised in a wheelchair for four weeks, and then began gradual weight-bearing.

At follow-up at two years she had no pain in her hips and full function. She was able to run and play without difficulty. Hip movement was improved with extension to +5°, flexion to 140°, and abduction to 60° bilaterally. In both flexion and extension, hip external rotation was 60° on the right and 45° on the left, and internal rotation was 15° on the right and 45° on the left. Radiographs showed union of the osteotomies.

Discussion

Osteochondromata of the proximal femur have been reported to occur in 30% to 90% of patients with hereditary multiple exostoses.^{1,4,5} In 1973 Stelling⁶ felt that limitation of hip movement due to osteochondromata was the main clinical problem in this disorder, but that the development of the proximal femur and acetabulum proceeded normally. More recent studies have shown abnormal development of both the proximal femur and the acetabulum. Shapiro et al⁴ noted that coxa valga occurred in 25% of patients, and acetabular dysplasia less often. Bassett and Scott³ described the femoral neck as typically broad, reflecting abnormal

Fig. 1

An AP radiograph of the pelvis showing incomplete coverage of the femoral heads even with the hips abducted.

Fig. 2

CT three-dimensional reconstruction of the pelvis showing multiple osteochondromata with increased valgus and anteversion of the femoral heads.

Fig. 3

An AP radiograph of the pelvis after bilateral varus derotation osteotomies and Steele acetabular osteotomies.

metaphyseal remodelling; coxa valga was common and the femoral head could be partially uncovered. Weiner and Hoyt⁷ found increased valgus and anteversion of the femoral neck associated with osteochondromata adjacent to the lesser trochanter in all 25 patients whom they reviewed. One had a septic dislocation of the hip which was attributed to severe valgus and anteversion of the proximal femur.

Although abnormalities of the femoral neck have been commonly described in hereditary multiple exostoses, there have been few reports of acetabular dysplasia. Tachdjian⁸ published two cases of severe coxa valga with acetabular dysplasia and uncovering of the femoral heads. In one case, this deformity was surgically corrected by a varus

intertrochanteric osteotomy. We could find no other reports of surgical treatment for subluxation of the hip or acetabular dysplasia associated with hereditary multiple exostoses.

Osteochondromata located near the lesser trochanter and coxa valga are common in hereditary multiple exostoses. Acetabular dysplasia and hip subluxation occur less frequently. Our case illustrates the importance of obtaining initial skeletal surveys in children with hereditary multiple exostoses to identify potential problems. Children with severe coxa valga and acetabular dysplasia should be closely monitored to determine if intervention is required. Coxa valga and increased anteversion of the femoral neck may be treated by a varus derotation osteotomy. Medial osteochondromata can be removed. If acetabular dysplasia is present, a pelvic osteotomy may be required to provide adequate coverage of the femoral head.

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