Acta Orthop Scand 57, 173–175, 1986

Dysplasia epiphysialis hemimelica of the hip

A case report

Dysplasia epiphysialis hemimelica most commonly presents as overgrowth of part of an epiphysis. A case of dysplasia epiphysialis hemimelica, presenting with hip pain secondary to an isolated epiphyseal exostosis of the femoral head, is reported. Excision of the osteochondroma produced complete relief of symptoms. This forme fruste of dysplasia epiphysialis hemimelica has not previously been described in the hip joint.

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Dysplasia epiphysialis hemimelica (DEH) is a rare condition that most commonly presents as overgrowth of part of an epiphysis and may cause gross disorganization of affected joints. Patients usually present with deformity, muscle wasting, restricted joint movement, and occasionally pain (Trevor 1950, Kettelkamp et al. 1966, Connor et al. 1983) in the first decade of life, though presentation as late as 63 years (Heiple 1961) is known. Fairbank (1956) predicted that many of the joints affected by DEH would develop secondary arthrosis.

We present a case of DEH that is unusual both in its site and limited destribution and in its symptomatology.

Case history

A man aged 38 was referred with a 10 year history of intermittent aching in the region of his right greater trochanter. Over the year preceding referral, his symptoms had worsened such that he complained of almost constant pain by day and frequently at night. He denied injury or childhood illness involving the hip. There were no symptoms relating to any other joint, and he was otherwise entirely well.

Examination of the right hip revealed loss of the last 15 degrees of flexion, 20 degrees of abduction and 15 degrees of internal rotation when compared with the left hip. There was no leg length discrepancy or visible external deformity.

Radiographs of the right hip were available from 9 years and 1 year prior to his referral. They had been reported as showing gross lipping of the femoral head with good preservation of the joint space. The radiograph taken at his consultation again showed a normal joint space. However, it was noted that the "lipping" of the femoral head was not a rim of superior osteophytes, but appeared to be a discrete bony mass arising posterolaterally from the head in the region of the capital femoral epiphysial scar (Fig. 1). A linear lucency at the base of the mass suggested a possible fracture of the exostosis. Reactive changes in the lateral lip of the acetabulum implied impingement of the exostosis on the acetabular rim when the hip was abducted.

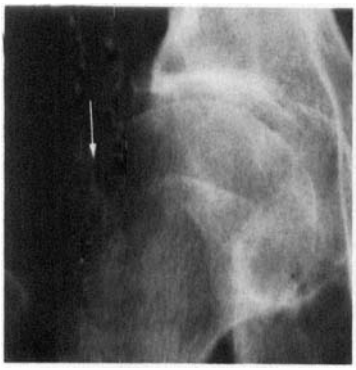
A diagnosis of an epiphyseal exostosis was made. No other exostoses were seen in radiographs of his right distal femur, knee, tibia and fibula, ankle, foot and wrist.

At operation, performed via a posterolateral approach, a tongue-shaped exostosis was found tightly applied to the posterosuperior aspect of the femoral neck on opening the posterior capsule. The exostosis came to within 0.5 cm of the articular surface of the femoral head, but the cartilage itself was healthy. There was evidence of moderate synovitis with an effusion in the hip joint. The exostosis was excised and the capsulotomy repaired.

After 48 hours of bed rest, he was mobilized, initially partially weight bearing, and commenced full weight bearing at 10 days. By 3 months he had regained full abduction and flexion, but internal rotation was still restricted by 15 degrees. When reviewed 2 years after surgery, he was symptomfree, and radiographs showed no evidence of regrowth of the exostosis or of progressive degenerative change (Fig. 2).

Histology

On microscopy, the specimen was seen to consist of mature bone with a small cartilage cap



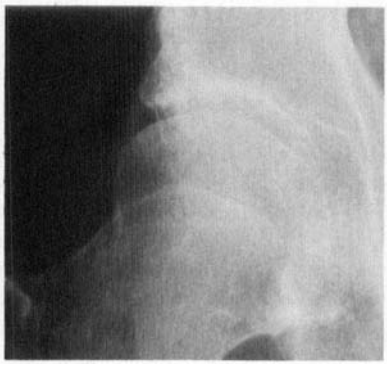


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Figure 1. Radiograph of the right hip at presentation showing a tongue-shaped exostosis arising from the posterosuperior aspect of the femoral neck. The arrow indicates the site of the fracture across the pedicle of the exostosis. Apart from some loss of joint space laterally, the joint is well preserved.

Figure 2. Radiograph taken 2 years after removal of the exostosis. There is no sign of regrowth nor of progressive degenerative change.

Figure 3. Photomicrograph of a section through the osteochondroma showing mature cartilage overlying a plate of bone. There is no evidence of growth activity in the cartilage nor of endochondral ossification (as would be expected in an osteophyte). At one end the bone plate is incomplete (arrow): fibrovascular tissue extends through the plate over bone that is denuded of cartilage. (H + E × 120).

and modules of cartilage on the external surface of the bone at one end. Most of the cartilage was resting on a cortical plate, but a part was undergoing resorption at the base. There was no evidence of active cell proliferation or of organized columns of hypertrophic chondrocytes. This organization of quiescent cartilage and bone is more like that of an osteochondroma than an osteophyte (Fig. 3). The cortex of the bony stalk was incomplete, and fibrous tissue with a poorly organized synovial membrane covered the external surface, confirming the presence of a fracture through the base with fibrous healing.

Since the histology confirmed that the exostosis was an osteochondroma, which radiographically had arisen on the epiphyseal side of the capital femoral epiphyseal scar, the diagnosis of DEH was substantiated.

Discussion

The first description of DEH was by Mouchet and Belot (1926), who called it "tarsomégalie". However, it was not recognized as a distinct clinical entity until 1950, when Trevor reported 10 cases of, what he called, "tarsoepiphyseal aclasis." Fairbank (1956) proposed the name "dysplasia epiphysialis hemimelica" when reporting a further 14 cases. DEH is defined as a developmental disorder of childhood in which there is asymmetric cartilaginous overgrowth of one, or more than one, epiphysis or a tarsal or carpal bone (Kettelkamp et al. 1966). Despite the term "hemimelica", the lesions are not always hemimelic in distribution (Kettelkamp et al. 1966, Luck and Smith 1972). Metaphyseal abnormalities are also not uncommon (Kettelkamp et al.1966, Fasting and Bjerkreim 1976). Because of these discrepancies the terms "epiphyseal osteochondromata" (Theodorou and Lanitis 1967) or "dysplasia epiphysialis osteochondromata" (Luck and Smith 1972) have been suggested.

Intraarticular osteochondromata may assume bizarre shapes because they grow in a confined space, or they may break away to form an intra-articular loose body (Noyes and Kivi 1972). A single osteochondroma may appear much as in the present case ; but if several osteochondromata are adjacent to one another, they may fuse to produce enlargement of one side of an epiphysis. If an osteochondroma grows across the epiphyseal plate, then

growth on that side of the epiphysis will be arrested (Luck and Smith 1972).

To date, only about 100 cases of DEH have been reported (Ippolito & Tudisco 1983). The condition may, however, be more common. Severe cases are rare, but are readily recognizable; less severe cases may be confused with loose bodies, osteophytes (as in the present case), or post-traumatic or osteochondritic conditions.

Luck and Smith (1972) suggest that treatment by excision of the abnormal cartilage focus may prevent the development of major deformity and arthrosis.

Acknowledgements

Our thanks are due to Dr. Colin Woods for his help in understanding the histology and to Miss Margaret Proudlock-Dunbar for her secretarial skills.

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