ABSTRACT

Purpose. To assess the radiographic features of 36 hips with hereditary multiple exostoses (HME).

Methods. Hip parameters of 12 males and 6 females (36 hips) aged 2 to 28 years with HME were assessed using anteroposterior radiographs. The recorded features included the sites of osteochondromas, the femoral head/neck ratio, the Reimer’s migration percentage, Sharp’s acetabular angle, the centre edge angle, the femoral neck-shaft angle, and degenerative changes.

Results. 15 of the 18 patients were asymptomatic; 3 complained of pain (2 underwent excision or bone biopsy); no lesion was malignant. Osteochondromas were most commonly located in the femur followed by the ilium; only one was intra-articular. 32 hips had coxa valga; 26 had an abnormal Reimer’s migration percentage; 17 had an abnormal Sharp’s acetabular angle; 12 had an abnormal centre edge angle; 32 had an abnormal femoral neck-shaft angle; and 6 had degenerative changes. Acetabular and femoral dysplasia as well as subluxation are common in patients with HME.

Conclusion. Borderline subluxated hips and those with marked coxa valga and/or acetabular dysplasia should be closely monitored to determine the need for surgery in the future. Subluxated hips should be operated on early, particularly in children and symptomatic adults.

Key words: exostoses, multiple hereditary; hip dislocation; osteochondroma

INTRODUCTION

Hereditary multiple exostoses (HME) is an autosomal dominant disorder, which manifests as multiple osteochondromas and skeletal deformities. In affected children and adolescents, the lesions are most commonly seen in the forearm, knee, and ankle. Deep-seated osteochondromas, such as those of the proximal femur or pelvis, rarely cause pain or

Hereditary multiple exostoses of the hip

Tarek AM El-Fiky, Wang Chow, Yun Hoi Li, Michael To
1 El-Hadra University Hospital, Alexandria, Egypt
2 Division of Paediatric Orthopaedics, Department of Orthopaedics and Traumatology, The Duchess of Kent Children’s Hospital, The University of Hong Kong, Hong Kong

Address correspondence and reprint requests to: Dr Wang Chow, Division of Paediatric Orthopaedics, The Duchess of Kent Children’s Hospital, The University of Hong Kong, Hong Kong. E-mail: wchowa@hku.hk
other symptoms, but deformity around the hip and premature osteoarthritis have been reported.

30 to 90% of patients with HME have osteochondromas in the proximal femur, which may result in coxa valga when the lesions are located near the lesser trochanter. Other deformities include excessive femoral anteversion and acetabular dysplasia. The lesions may occur intra-articularly. We assessed the radiographic features of 36 hips with HME.

MATERIALS AND METHODS

Between 1984 and 2007, 12 males and 6 females (36 hips) aged 2 to 28 (mean, 12; standard deviation [SD], 7) years were diagnosed with HME in our hospital. Hip parameters were assessed using anteroposterior (AP) radiographs. They included (1) the sites of osteochondromas (Table 1); (2) the femoral head/neck ratio (to assess the broadening of the femoral neck based on the widest areas of the head and neck), whereas the mean ratio of 20 hips with unrelated disease in patients aged 2 to 21 (mean, 11) years was used as the control; (3) the Reimer’s migration percentage—the percentage of the uncovered femoral head lateral to the Perkins’s line; (4) the Reimer’s migration percentage—the percentage of the uncovered femoral head lateral to the Perkins’s line; (5) the Sharp’s acetabular angle (Fig. 1)—the angle between the line joining the superolateral and inferomedial part of the acetabulum and another line joining the inferior tips of the pelvic teardrops (normal, ≤45º); (6) the centre edge angle (Fig. 1)—the angle between a line joining the centre of the femoral head to the lateral edge of the acetabulum and a line perpendicular to the inter-teardrop line passing through the centre of the femoral head (normal, ≥20º); (7) the femoral neck-shaft angle—the angle formed between the anatomic axes of the femoral neck and shaft—which is difficult to determine because of its wide sexual, bilateral and inter-population variations (normal, ≤135º for subjects ≥12 years and ≤150º for younger persons); and (7) degenerative changes.

Correlations between the Reimer’s migration percentage and other parameters were analysed using Pearson correlation coefficients. The femoral head/neck ratios in the HME and control groups were compared using Student’s t test.

RESULTS

15 of 18 patients were asymptomatic; 3 complained of pain (2 underwent excision or bone biopsy); no lesion was malignant. Osteochondromas were most commonly located in the femur (Fig. 2a) followed by the ilium; only one was intra-articular (Fig. 2b, Table 1). 32 (89%) hips had coxa valga (Fig. 2c). The mean femoral head/neck ratios were 0.84 (SD, 0.1) and 1.17 (SD, 0.14) in the HME and control groups, respectively (p<0.001, Table 2); 26 (72%) hips had an abnormal Reimer’s migration percentage of ≥20%; 12 (33%) had an abnormal centre edge angle of <20º (Table 3). 32 (89%) hips had an abnormal femoral neck-shaft angle (Table 3). Six hips (4 patients) showed joint space narrowing suggestive of degenerative changes. The Reimer’s migration percentage was significantly correlated with the Sharp’s acetabular angle (r=0.41, p=0.012) and the centre edge angle (r=−0.68, p<0.001;
DISCUSSION

Anteroposterior radiographs alone are inadequate for identifying the number and sites of osteochondromas in HME hips. Some lesions may be radiolucent, especially in skeletally immature subjects. Lesions in the medial femur and peri-acetabulum may alter the hip’s parameters and affect its development, resulting in acetabular dysplasia, but lesions in the lateral femur and ilium usually cause pain and discomfort rather than alter hip geometry. Intra-articular osteochondromas have been reported\(^9,17\), their actual prevalence may be higher if they are asymptomatic.

The severity of the deformity at the proximal

Table 2

<table>
<thead>
<tr>
<th>Parameter</th>
<th>No (% of hips)</th>
<th>Mean±SD (range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Femoral head/neck ratio</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patients with hereditary</td>
<td>36 (100)</td>
<td>0.8±0.1 (0.6–1.1)</td>
</tr>
<tr>
<td>multiple exostoses</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Controls</td>
<td>20</td>
<td>1.2±0.1 (0.9–1.5)</td>
</tr>
<tr>
<td>Reimer’s migration percentage</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Overall</td>
<td>36 (100)</td>
<td>24±8 (8–48)</td>
</tr>
<tr>
<td>Normal</td>
<td>10 (28)</td>
<td>15±4 (8–20)</td>
</tr>
<tr>
<td>Borderline</td>
<td>19 (53)</td>
<td>25±4 (20–30)</td>
</tr>
<tr>
<td>Subluxation</td>
<td>7 (19)</td>
<td>36±7 (30–48)</td>
</tr>
<tr>
<td>Sharp’s acetabular angle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Overall</td>
<td>36 (100)</td>
<td>45°±4° (30°–52°)</td>
</tr>
<tr>
<td>≤45°</td>
<td>19 (53)</td>
<td>42°±4° (30°–45°)</td>
</tr>
<tr>
<td>&gt;45°</td>
<td>17 (47)</td>
<td>48°±0° (45°–52°)</td>
</tr>
<tr>
<td>Centre edge angle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Overall</td>
<td>36 (100)</td>
<td>22°±5° (7°–30°)</td>
</tr>
<tr>
<td>≥20°</td>
<td>24 (67)</td>
<td>25°±3° (20°–30°)</td>
</tr>
<tr>
<td>&lt;20°</td>
<td>12 (33)</td>
<td>16°±4° (7°–19°)</td>
</tr>
<tr>
<td>Femoral neck-shaft angle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Overall</td>
<td>36 (100)</td>
<td>154°±7° (137°–169°)</td>
</tr>
<tr>
<td>Normal</td>
<td>4 (11)</td>
<td>143°±4° (137°–145°)</td>
</tr>
<tr>
<td>Abnormal</td>
<td>32 (89)</td>
<td>155°±6° (141°–169°)</td>
</tr>
</tbody>
</table>

Table 3

<table>
<thead>
<tr>
<th>Patient age (years)</th>
<th>Femoral neck-shaft angle</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Right</td>
<td>Left</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>152°</td>
<td>145°</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>137°</td>
<td>151°</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>156°</td>
<td>160°</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>163°</td>
<td>158°</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>163°</td>
<td>156°</td>
<td></td>
</tr>
<tr>
<td>21</td>
<td>148°</td>
<td>147°</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>157°</td>
<td>161°</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>153°</td>
<td>162°</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>157°</td>
<td>164°</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>169°</td>
<td>160°</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>152°</td>
<td>150°</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>141°</td>
<td>154°</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>156°</td>
<td>151°</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>156°</td>
<td>160°</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>158°</td>
<td>157°</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>156°</td>
<td>154°</td>
<td></td>
</tr>
<tr>
<td>28</td>
<td>143°</td>
<td>147°</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>145°</td>
<td>143°</td>
<td></td>
</tr>
</tbody>
</table>

* Normal

Figure 2  (a) A symptomatic lateral exostosis in the right proximal femur, (b) multiple intra-articular osteochondromas in the acetabulum, and (c) osteochondromas in bilateral medial femurs and coxa valga.
femur can be measured using the head/neck ratio. A decreased ratio suggests the presence of osteochondromas at the femoral neck causing broadening. Acetabular dysplasia is an abnormality of its shape or size, or of the proportion or alignment of the femoral head to the acetabulum. Such cases are few, although abnormality of the femoral neck is common in HME patients. In our study, acetabular dysplasia was common (47% of hips had abnormal Sharp’s acetabular angle and 33% had an abnormal centre edge angle). Both angles are reliable for assessing acetabular dysplasia.

Some consider coxa valga a developmental consequence to acetabular dysplasia. Others attribute it to the presence of osteochondromas at the medial femoral neck preventing the capital growth plate having a normal transit from a horizontal to a more vertical alignment. This accentuates the valgus deformity as further longitudinal growth occurs. Others have suggested that coxa valga is the initial deformity.

In our study, 72% of hips had abnormal Reimer’s migration percentage (19% were subluxated and 53% were borderline), and subluxation correlated with acetabular dysplasia. Although Reimer’s migration percentage did not correlate with the neck-shaft angle, coxa valga and intra-articular lesions may be attributed to subluxation and acetabular dysplasia. A case of surgical intervention for an HME patient with severe acetabular dysplasia (<50% coverage of the femoral head) has been reported, as has a case of septic dislocation owing to HME.

In our study, most patients were asymptomatic and young (only 8 were older than 10 years) and had fewer symptoms than those who were older. Only one patient underwent excision of a symptomatic osteochondroma. The prognosis of borderline subluxated hips is uncertain. In a report of 12 patients with a mean age of 17 years, one patient had a labral tear, one underwent total hip arthroplasty at the age of 54 years, and one developed sarcomatous changes. Our study has limitations in that it was a cross-sectional and not a longitudinal study with a wide age range.

CONCLUSION

Hip radiographs should be obtained for children with HME to identify potential problems. Acetabular and femoral dysplasia and subluxation (coxa valga, abnormal centre edge angle and Sharp’s acetabular angle) are common in these patients. Borderline subluxated hips and those with marked coxa valga and/or acetabular dysplasia should be closely monitored to determine the need for surgery in the future. Subluxated hips should be operated on early, particularly in children and symptomatic adults.

ACKNOWLEDGEMENT

We thank Prof Hafez Sadek from Alexandria University for his help with the statistical analysis.

REFERENCES