ACRODYSOSTOSIS AND PROTRUSIO ACETABULI

AN ASSOCIATION

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Progressive protrusio acetabuli in a patient with acrodysostosis (peripheral dysostosis Type 12) has not, as far as we know, been reported previously. It is recognised that epiphyseal dysplasias may lead to disturbances of acetabular growth, but generally this results in a shallow socket with associated changes in the proximal femoral epiphysis.

CASE REPORT

A 12-year-old girl was referred for orthopaedic review with the complaint that her hand function was deteriorating; in particular, she was finding it increasingly difficult to write, and fine manipulative movements were noted to be clumsy. The clinical appearance of the hands and feet are shown in Figures 1 and 2, and a diagnosis of peripheral dysostosis was confirmed by the characteristic radiographic appearance of the hands (Fig. 3) which included angulatory deformities of the digits secondary to cone-shaped epiphyses. The epiphysis indents the metaphysis, which gradually envelops that portion of the subchondral plate to produce a lamda-shaped deformation of the proximal portion of the middle phalanx.

The patient's legs were of equal length, but she had significantly reduced abduction and rotation of the hips. A pelvic radiograph revealed bilateral protrusio acetabuli (Fig. 4) which was initially asymptomatic but is now progressive and restrictive.

DISCUSSION

The association between acrodysostosis (peripheral dysostosis Type 12) and protrusio acetabuli is not altogether surprising in view of the changes that this type of skeletal dysplasia also produces in the proximal interphalangeal joints of the fingers. Although this association has not been remarked upon previously,

Giedion (1973), in his report about acrodysplasias, included a radiograph of hips with changes similar to those seen in Perthes' disease and with evidence of protrusio acetabuli. In a recent paper about the trichorhino-phalangeal syndrome, Howell and Wynne-Davies (1986) also presented a 27-year-old patient with cone-shaped epiphyses and protrusio acetabuli.

Shore (1926) described the characteristics of peripheral dysostosis Type 12, noting that a painless swelling affects the interphalangeal joints and that the patient is usually of short stature. The changes become evident between the ages of four and 10 years and affect girls four times as often as boys. Eventually the widening and angulation of the digits limits movement. Early epiphyseal growth-plate closure is seen, as well as radiographic alterations including middle phalangeal distortion (Fig. 3), thickening of the phalanges, the metacarpals and the metatarsals, advanced carpal bone age and enlargement of the medial cuneiform. In addition to changes in the proximal femoral epiphyses which resemble Perthes' disease, Maroteaux and Malamut (1968) also described coxa valga.

In our patient there was mild coxa vara (Fig. 4) which is known to co-exist with protrusio acetabuli (Hooper and Jones 1971). The aetiology of primary protrusio acetabuli remains obscure (Alexander 1965), although Gilmour (1939) considered that premature fusion of the triradiate cartilage would prevent the remodelling of the normal prepubertal inward bulge of the ischio-acetabular pillar. The protrusion may either be idiopathic or familial (Rechtman 1936) with some racial variation (Friedenberg 1963), and, as in this case, is generally bilateral with a greater predominance in girls.

Post-traumatic premature fusion of the triradiate cartilage paradoxically produces a shallow acetabulum (Rodrigues 1973; Gepstein, Weiss and Hallel 1984), but the condition is unilateral and results in a widening of the medial wall of the acetabulum (Hallel and Salvati 1977).

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Fig. 1

Fig. 3

Figures 1 and 2 – Clinical appearance of the hands and feet. Figure 3 – A radiograph of her hands confirmed the diagnosis of peripheral dysostosis. Figure 4 – A pelvic radiograph of the same patient showed bilateral protrusio acetabuli.

Fig. 2

Fig. 4

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