A case of multiple epiphyseal dysplasia complicated by unilateral Perthes' disease

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Case report

A 7-year-old Japanese boy was referred to us because of short stature, which was identified in infancy. He was a product of an uncomplicated pregnancy and delivery. The family history was unremarkable. Radiographs at aged 4 years revealed multiple epiphyseal dysplasia (MED) in the wrist, knee, and hip joints (Figure 1), but no involvement of the spine.

On examination, the patient was found to be of normal intelligence; he had a standing height of 104 cm (–3 SDs of the Japanese standard for his age), a sitting height of 61 cm, and an arm span of 102 cm. He had severe bilateral myopia and bilateral sensorineural hearing loss. Internal rotation of the hip joints was reduced to 35°, and he had genu valgum.

On a follow-up visit at aged 8 years, he complained of pain in the right knee, which resolved in 6 months without specific treatment. Internal rotation of the right hip gradually decreased, but no coxalgia or limp was observed. Serial radiographs showed the cyclic changes of the right femoral capital epiphysis typical of Perthes' disease (Figure 2). On examination at aged 14 years, he had 5°–10° limitation of motion in flexion, adduction, and internal and external rotations of the right hip joint. He was otherwise healthy and had no complaints.

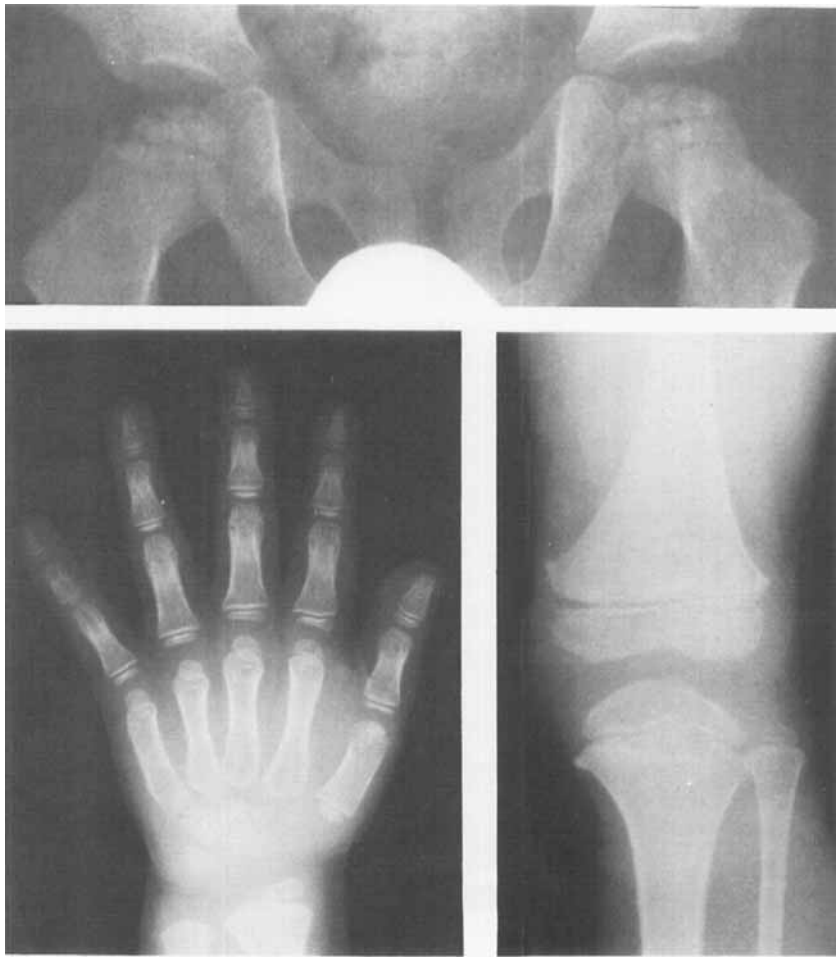
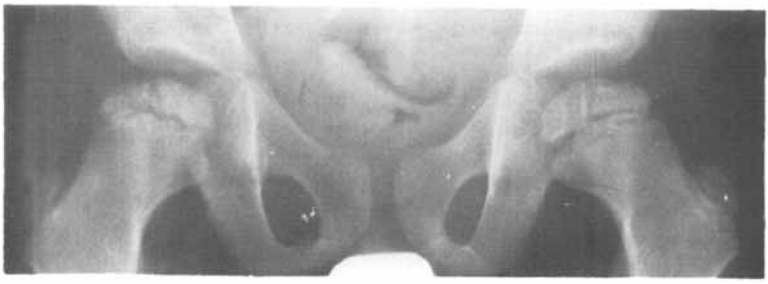
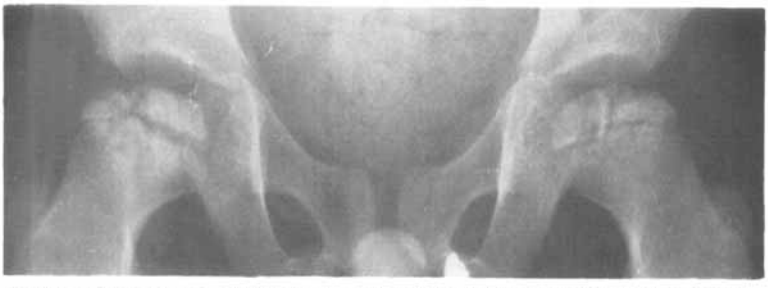


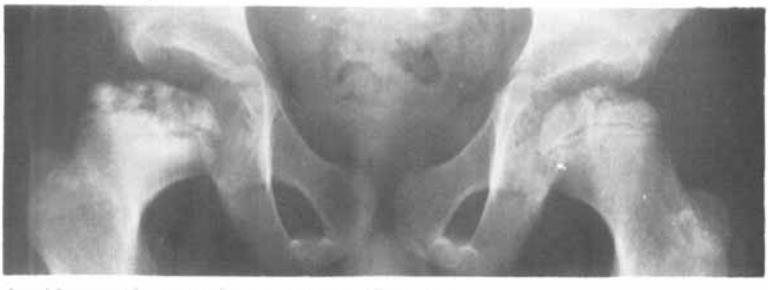
Figure 1. Radiographs at age 4 showing multiple epiphyseal involvement.



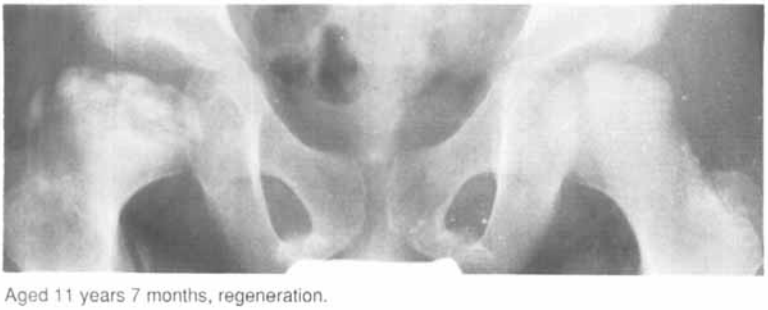
Aged 7 years 2 months, symmetric involvement.



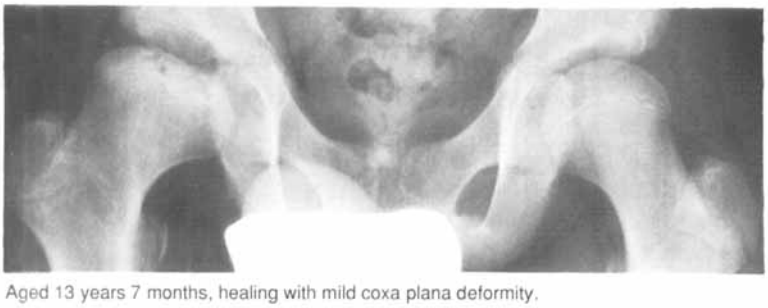
Aged 8 years 3 months, subchondral translucent line in the right capital epiphysis with widening of the epiphyseal line.



Aged 9 years 10 months, fragmentation and flattening.



Aged 11 years 7 months, regeneration.



Aged 13 years 7 months, healing with mild coxa plana deformity.

Figure 2. Serial radiographs of the hips. Cyclic changes of the right hip indicate associated

Discussion

Our case was diagnosed as multiple epiphyseal dysplasia (MED) on the basis of short stature, associated extraskeletal anomalies, and multiple symmetric involvement of the epiphyses. The capital femoral epiphyses before aged 8 years showed symmetric, homogeneous lesions without cystic changes in the metaphyses, which are consistent with MED (Crossan et al. 1983).

However, the radiographic course of the hips after aged 8 years was quite unique. The left hip became nearly normal by the age of 13 years. Such improvement is a feature of dysplasia epiphysealis capitis femoris except that it occurs in children under aged 4 years (Meyer 1964, Harrison 1971). The right hip, by contrast, showed the cyclic changes characteristic of Perthes' disease, i.e., sequential stages of increased density, fragmentation, resorption, and regeneration.

The association of Perthes' disease with MED may be simply fortuitous, because there have been few reports of this association. Wenger and Ezaki (1981) described a case of MED showing acute collapse of both femoral heads in an early adolescence. Mandell et al. (1989) reported the radiographic documentation of the occurrence of Perthes' disease in 10 patients with MED. The incidence of the association, however, may be underestimated, because there are only a few long-term follow-up reports on MED hips.

MED may predispose to Perthes' disease, or there may be a common state of preparedness for the two conditions. Constitutional abnormalities in Perthes' disease have been

reported, i.e., "rostral sparing" (Burwell et al. 1978) and an increased incidence of congenital anomalies (Hall et al. 1979). Based on these observations, Harrison and Burwell (1981) hypothesized that Perthes' disease is a focal expression of a general disorder of skeletal growth. Ponseti (1956), who made a histologic study of Perthes' disease, found changes that suggested an underlying bone dysplasia. However, the hip lesions in our case showed distinct laterality, thus indicating the importance of acquired factors.

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