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Clinical History:

An 8-year-old boy presented with multiple joint pains, which started at the age of about 1-2 years. Both wrists were symmetrically and severely affected.

Figure 1: Posteroanterior radiograph (left hand)  Figure 2: Posteroanterior radiograph (right hand)

What is the diagnosis?

a) Juvenile rheumatoid arthritis.
b) Leukaemia.
c) Juvenile dermatomyositis.
d) Haemophilia.
e) Tuberculous arthritis.

This radiology case was prepared by: Dr. Wilfred C.G. Peh, Associate Professor, Department of Diagnostic Radiology, The University of Hong Kong, Queen Mary Hospital.
Answer:

a) Juvenile rheumatoid arthritis.

Radiological findings

The posteroanterior wrist radiographs (Figures 1-3) show fragmentation, irregularity and deformity of all the carpal bones. Similar changes affecting the distal radial and ulnar epiphyses are demonstrated. There is mild periarticular osteoporosis. Both hands are symmetrically involved.

Figure 3: This figure is identical to Figure 2 with addition of arrows. Irregular fragmentation of the carpal bones (open arrows) and distal forearm epiphyses (arrows) are present.

Discussion

In children presenting with painful joint symptoms, the initial role of imaging is to determine whether the problem is indeed due to a real joint disease or not. Conditions which may mimic joint disease include leukaemia and juvenile dermatomyositis. The term juvenile chronic arthritis (JCA), besides encompassing the disease juvenile rheumatoid arthritis (JRA), also includes other paediatric connective tissue diseases, juvenile ankylosing spondylitis and juvenile psoriatic arthritis.

JRA is relatively common, having a prevalence of 91 to 70 per 100,000 population, and may affect children as young as 1 to 2 years of age. The 3 main clinical subgroups, namely: systemic onset, polyarticular onset and pauci- or mono-articular onset, account for about 70% of cases. Bony erosions, joint space narrowing and bony ankylosis are late radiographic features of JRA. In young children where the bone ends are still cartilaginous, the radiographic clues for the detection of joint disease may be subtle and are easily missed if one is unaware of them. These signs include growth disturbances due to periarticular hyperaemia, typically giving a bulbous appearance to the distal ends of the proximal phalanges; periarticular periostitis; periarticular soft tissue swelling; and periarticular osteopaenia.

The limitation of using plain radiographs to evaluate joint disease in children is the non-visualization of cartilage, the position of which is indirectly implied from the relatively large space between the ossified bone ends. In such circumstances, considerable cartilage destruction often occurs before any bony erosion is radiographically apparent. Magnetic resonance imaging (MRI) provides direct demonstration of the cartilage and surrounding soft tissues in a multiplanar manner. This imaging technique is very promising in the detection, diagnosis and monitoring of disease activity in patients with JRA.

Leukaemia

Clinically, patients with leukaemia may present with painful joints. The most important radiographic feature of leukaemia is a lucent band in the metaphysis of the involved bone. If present symmetrically in a child over 2 years of age, this sign is considered pathognomonic. Other signs of leukaemia include periosteal reaction, metaphyseal destruction and pathological fractures. None of these radiographic features were present in this case.

Juvenile dermatomyositis

Although the joints are usually normal or only minimally affected in juvenile dermatomyositis, muscle involvement often results in multiple joint pain. Muscle or soft tissue calcification represent late changes of this disorder. MRI
is the most sensitive means of detecting early muscular involvement and is useful in guiding the selection of the most appropriate biopsy site. The radiographic changes in this case are not compatible with those of juvenile dermatomyositis.

Haemophilia

The joint changes in haemophilia are due to repeated episodes of intra-articular bleeding. Large joints such as the knee, elbow, ankle, hip and shoulder are most commonly affected. Intra-osseous bleeds may produce lucent defects. There may also be periarticular osteoporosis and coarsened trabecula. Sometimes, dense soft tissue swelling due to deposition of haemosiderin in thickened synovium may be seen periarticularly. In this case, the clinical history, sites of involvement and radiographic appearances exclude the diagnosis of haemophilia.

Tuberculous arthritis

Tuberculous arthritis may be associated with pulmonary tuberculosis. The frequently involved peripheral joints are the hips, knees, hands and feet. The disease process is usually of an insidious onset, with the joint space being unaffected till late in the disease course. Periarticular osteopenia often precedes destructive changes. Erosions are typically present at the peripheral non-contact points of the joints. Tuberculous arthritis is classically monarticular, and can be excluded in this case where involvement is bilateral and symmetrical.

References


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