Case 7625

Perthes disease with metaphyseal cyst
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Patient: 4 years, female

Clinical History:
A 4 year old girl was referred for evaluation of the right hip pain.

Imaging Findings:
A 4 year old girl presented with progressively worsening right hip pain for 3 months. She had difficulty in walking and there was no evidence of fever or erythema on the painful hip joint. Physical examination revealed decreased internal rotation and abduction of the right hip joint. She was limping instead of smooth walking. Her white blood cell count and sedimentation rate were in normal limits. Radiographs of the right hip showed the irregularity and collapse of the femoral epiphysis, and also a radiolucent area with a faintly sclerotic sharp margin in the posteromedial part of the femoral metaphysis adjacent to physis zone (Fig 1a,b). MRI examination of the pelvis revealed fragmentation and collapse of the capital femoral epiphysis and the absence of fat signal in T1 weighted images indicating the presence of osteonecrosis on the right hip joint (Fig 2). Metaphyseal lesion had low signal intensity in T1 weighted images and high signal intensity in inversion recovery images suggesting fluid or oedema in that area. There was no abnormality on the left hip joint (Fig 3a,b). The diagnosis of Perthes disease was made according to the radiological findings. A conservative course of restricted weight bearing was instituted without biopsy.

Discussion:
Perthes disease is the idiopathic avascular necrosis of the proximal femoral epiphysis. It affects children particularly between the age of 4-8 years. It is more frequent in boys than girls in a ratio of approximately 5:1.3. Either hip can be altered or bilateral abnormalities are detected in about 10-20% of cases, rarely in girls [1]. The major clinical signs are limping, pain and limitation of joint motion, particularly abduction and internal rotation of the hip. The hypothesis of clotting abnormalities with vascular thrombosis seems to be the most likely aetiology for the pathogenesis of the disease in recent papers [2]. Radiographic findings have been summarized by Caffey [3] as:
1.) soft tissue swelling on the lateral side of articulation, 2.) smallness, lateral displacement, fissuring and fracture, flattening and sclerosis of the femoral ossification nucleus, and 3.) intraepiphysial gas.
The progression and extent of disease is highly variable. Further compression, disintegration, fragmentation and sclerosis of the epiphysis together with metaphyseal changes can be seen. Metaphyseal cysts and widening and shortening of the femoral neck also can be seen and these are additional manifestations of the disorder.
Metaphyseal cysts are typically located in the anterior femoral metaphysis. Although they are a characteristic part of the radiographic picture of this disease, the pathogenesis of the metaphyseal radiolucencies is debated and there are different hypotheses in the literature. These changes have been described as growth plate disturbances with extension of tongues of physeal cartilage into the metaphysis [4]. Ponseti proposed that changes in Perthes disease result from abnormal forces transmitted across the physis [5]. According to the report of Johnson et al, after osteonecrosis and collapse of the femoral head, abnormal forces are applied through the physis which result in...
compression of the metaphyseal capillary loops. This prevents the normal resorption of uncalcified cartilage, leaving residual islands of uncalcified cartilage in the metaphysis. These metaphyseal lesions may also be non-cartilaginous suggesting metaphyseal cysts, fragmentation of trabecular bone, fibrous or fibrofatty tissue, granulation tissue [6]. In the study of Song et al. of the 30 hips at the avascular stage, 33% showed metaphyseal cyst in MRI scans. Of the 53 hips at the fragmentation stage, 60% showed the metaphyseal cyst [7]. So metaphyseal cysts are not rare lesions and are often seen in the delayed phase of the Perthes disease. In this presented case, metaphyseal cystic lesion was also diagnosed in the fragmentation stage. According to some authors these lesions do not require biopsy proof of their nonneoplastic or noninfectious nature, but may have prognostic significance in having a high association with subsequent severe head and neck deformity [8]. Although, Legg-Calve Perthes disease was described as femoral head disorder by early investigators, metaphyseal changes and subsequent severe head and neck deformity point out that Perthes disease is not a disorder of the femoral head exclusively.

Differential Diagnosis List: Legg-Calve Perthes disease with metaphyseal cyst.

Final Diagnosis: Legg-Calve Perthes disease with metaphyseal cyst.

References:


Figure 1

Description: Collapse and fragmentation of the right femoral epiphysis and a round metaphyseal radiolucency adjacent to physis (arrow). Origin:
Description: Collapse and fragmentation of the right femoral epiphysis and a round metaphyseal radiolucency adjacent to physis (arrow). Origin:
**Description:** Loss of the fatty signal of right femoral epiphysis and collapse suggesting osteonecrosis on the right hip joint and cystic lesion in the femoral metaphysis (arrow). **Origin:**
Description: Cystic lesion in the posteromedial part of the femoral metaphysis adjacent to physis on the right hip joint (arrow). Left hip joint is normal. Origin:
Description: Cystic lesion in the posteromedial part of the femoral metaphysis adjacent to physis on the right hip joint (arrow). Left hip joint is normal. Origin: