Non-Hodgkin's lymphoma of bone causing avascular necrosis of the femoral head

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Summary: A 14 year old boy presented with a painful hip, initially attributed to Perthe's disease although it is uncommon in teenagers. Subsequent investigation showed that the underlying pathology was non-Hodgkin's lymphoma.

Introduction

A rare case of non-Hodgkin's lymphoma of bone, occurring in the femoral neck, is presented. This caused avascular necrosis of the femoral head which was initially diagnosed as Perthe's disease. We emphasize the importance of not making such a diagnosis in the teenage population in whom Perthe's disease is not usual, and in this age group, alternative diagnoses should be considered before ascribing the changes to Perthe's disease. Such aetiologies may include any lesion in the femoral neck which would interfere with the blood supply to the femoral capital epiphysis, and causes may include trauma, bleeding and thrombotic disorders, infections, endocrine disorders and bone dysplasias.

Case report

A previously fit 14 year old boy presented with a painful left hip following a trivial injury sustained during a rugby game. The pain persisted and he was admitted to hospital two weeks later. A plain radiograph taken on admission (Figures 1, 2) showed a sclerotic femoral head and these changes were attributed to Perthe's disease. On physical examination the only abnormal finding was some restriction of movement of the left hip. He was afebrile, and biochemical and haematological investigations were normal.

The pain failed to settle on bed rest and two weeks later he became pyrexial. The erythrocyte sedimentation rate then was 48 mm/h, rising some days later to 112 mm/h. He began to lose weight and his left groin became tender. A further radiograph (Figure 3) revealed a lytic lesion of the femoral neck and apophysis, present in retrospect in Figure 1. At this stage malignancy was suspected and he underwent open biopsy. At operation the femoral head was not fragmented. Biopsy of this showed changes consistent with an avascular necrosis. Histology of the femoral neck biopsy showed non-Hodgkin's lymphoma of bone.

The patient was transferred to our centre where further investigations including bone marrow aspiration, trephine iliac crest biopsy, cerebrospinal fluid examination, chest radiograph, abdominal and pelvic computed tomographic scans and isotope bone scans were all normal. The lymphoma therefore appeared confined to the bone of the femoral neck. The patient underwent chemotherapy and is well with no evidence of local recurrence or disease else where three years after diagnosis.

Discussion

The femoral head is supplied by ascending branches of the medical and lateral circumflex femoral vessels. These form an irregular vascular ring around the femoral neck, closely corresponding to the distal attachment of the joint capsule. This vascular ring receives tributaries from the superior gluteal arteries. Three major groups of cervical arteries arise from the vascular ring and pass upwards along the femoral neck. They give origin to the central and peripheral metaphyseal branches which supply the juxta-epiphyseal region of the metaphysis before they continue as the epiphyseal arteries to supply the epiphysis. The acetabular
artery arises from either the obturator arteries, medial circumflex or branches of these arteries. It enters the hip joint through the acetabular notch and divides into two branches which run subsynovially. Thus a lesion arising in the femoral neck (such as this patient's lymphoma) which compromises the blood supply to the femoral capital epiphysis may lead to avascular necrosis.
Non-Hodgkin's lymphoma of bone (or reticulum cell sarcoma) was first described by Oberling in 1928. It accounts for 5% of all malignant bone tumours; and 5% of all extra nodal lymphomas. Histologically, reticulum cells are the most characteristic cells in this tumour, but there is usually a significant admixture of lymphoblasts and lymphocytes. Our patient is particularly young to have this tumour, 93% occur in patients over 20 years old and 50% occur over the age of 40. The tumour is twice as common in males than females. The tumour usually presents with pain and swelling but the onset is often insidious. Systemic symptoms are unusual and do not appear to affect the prognosis. When this tumour occurs in bone, 50% involve the femur and pelvis, 20% occur in the upper extremities; less commonly the spine, mandible and skull are affected.

The earliest radiological sign is diffuse medullary destruction – a 'moth-eaten' appearance with poorly defined margins. At this stage it is impossible to differentiate the lesion from other neoplastic or infective processes. A periosteal reaction is rare. Cortical destruction occurs eventually and a surrounding soft tissue mass ensues. Sclerotic lesions are uncommon. Twenty seven percent present with a pathological fracture. It has been suggested that some radiological signs such as pathological fracture, 'layering' of periosteal new bone, cortical breakthrough, soft tissue swelling and mass are related to early recurrence and therefore indicated poorer prognosis. Prognosis is considerably better than in other malignant bone tumours. The five year survival rate is 40%–60%, although Bacci et al. recently reported an 88% survival rate for this tumour after an average follow-up period of 86 months. Tumours which have a preponderance of cells with cleaved nuclei (as in this case) have a significantly better prognosis than in those with either non-cleaved or pleomorphic nuclei. Treatment has consisted of radiotherapy and chemotherapy, but in view of the favourable response of malignant round cell tumours to chemotherapy alone, this is now regarded as the treatment of choice. Our patient received intensive chemotherapy for 18 months and remains well after three years.

Lesions of the femoral neck may interfere with the blood supply to the femoral capital epiphysis resulting in avascular necrosis. When such changes are present on a radiograph especially in children over the age of 12 (Perthe's disease is unusual over this age and the metaphyseal lesion in Perthe's is nearly always located well forward on the metaphysis, directly under the site of maximal compression) causes other than Perthe's should be considered.

References