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Case Report

Double pathology, sarcoidosis associated with multiple myeloma: A case report

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ABSTRACT

The association of sarcoidosis with multiple myeloma is not well known. Including this case report, 12 cases of patients with both sarcoidosis and multiple myeloma have been reported in the literature. The skeletal lesions of both conditions have many clinical and radiological similarities, and unless clinicians are aware of the association and the possibility of dual pathologies, the diagnosis of multiple myeloma in patients known with sarcoidosis may be missed. We present a case of a patient known with longstanding sarcoidosis who was found to have multiple lesions on magnetic resonance imaging (MRI) involving the pelvis and both proximal femurs. Histological analysis revealed the presence of both non-necrotising granulomas consistent with sarcoidosis, and sheets of plasma cells consistent with a plasma cell neoplasm.

1. Case presentation

A 50 year old female machinist presented to our general orthopaedic outpatients' department with a 5 month history of right groin and buttock pain of spontaneous onset. She had a background history of hypertension and biopsy-proven sarcoidosis. The pain radiated to the right knee and had progressively worsened over the preceding month. She had not experienced any constitutional symptoms, had no previous malignancies, and had been on a daily dose of 20 mg prednisone and 150 mg azathioprine for 4 years as part of her treatment for sarcoidosis.

Examination revealed an antalgic gait with a painful and stiff right hip. The pain was most severe in flexion and internal rotation.

An anteroposterior (AP) radiograph of the pelvis and lateral radiograph of the right hip demonstrated moderate degenerative changes of both hips (Figs. 1 and 2).

A provisional differential diagnosis of early avascular necrosis (AVN) or femoroacetabular impingement (FAI) was made. Renal function and calcium, magnesium and phosphate (CMP) levels were normal, C-reactive protein was 8.5 mg/l and the erythrocyte sedimentation rate was 40 mm/h. An MRI revealed no evidence of AVN, with areas of intermediate to high signal in the right femoral head and neck, as well as the left femur and pelvis (Figs. 3 and 4). The MRI findings were in keeping with sarcoidosis. Further clinical work up was recommended to exclude other causes such as metastatic lesions or myeloma if clinically indicated.

By this time the patient's symptoms had continued to progress, and a second AP pelvis X-ray (Fig. 5) revealed a lytic lesion in the inferior neck of the right femur with a Mirels' score of 11 [1]. After consultation with both the musculoskeletal tumour and arthroplasty units, it was decided to perform a biopsy of the femoral head/neck and total hip replacement as a single stage procedure.

Uncemented acetabular and femoral components were used, with a ceramic-on ceramic bearing surface. Macroscopic examination of the excised femoral head confirmed a 17×13 mm^2 lytic lesion in the inferior neck. Histological analysis of the femoral head showed hypercellular bone marrow with diffuse replacement of marrow spaces by a population of mature plasma cells. Occasional plasmablasts and multinucleate forms were present (Figs. 6 and 7). Immunohistochemistry demonstrated Lambda light chain restriction (Fig. 8). In addition, scattered non-necrotising granulomas consistent with sarcoidosis were present (Figs. 6 and 7). Special stains for acid-fast bacilli and fungal organisms were negative.

Bone marrow aspiration, biopsy and serum protein electrophoresis confirmed the diagnosis of multiple myeloma (Fig. 9). The patient was referred to the department of haematology, where treatment with high dose dexamethasone and cyclophosphamide was started.

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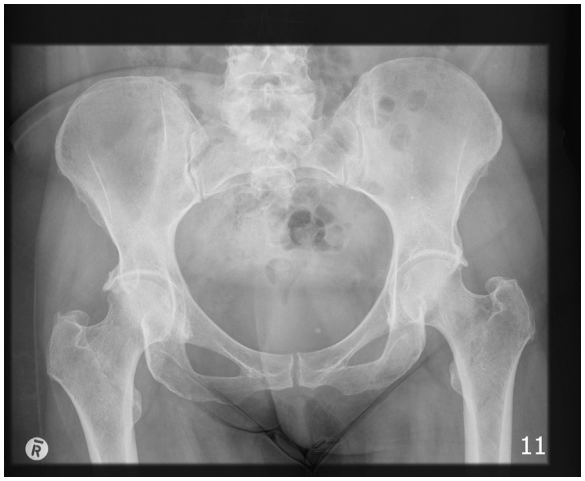

Fig. 1. Anteroposterior radiograph of pelvis at presentation.

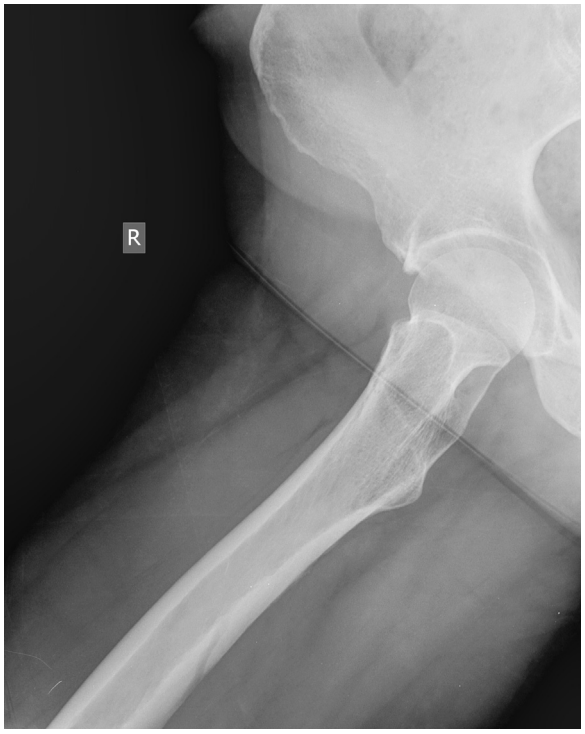

Fig. 2. Lateral radiograph of right hip at presentation.

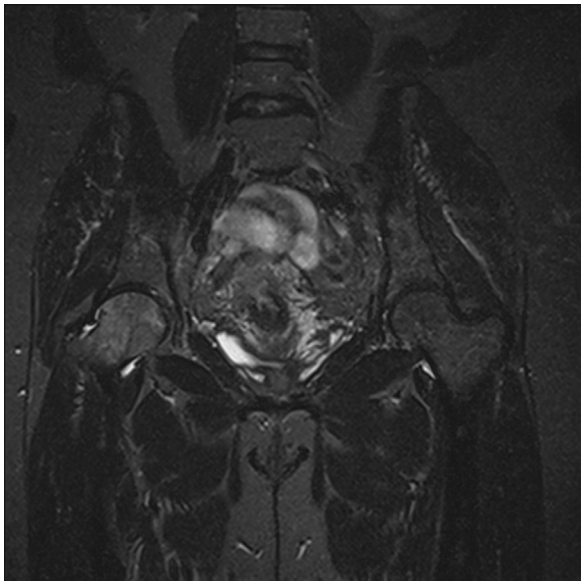

Fig. 3. STIR coronal MRI showing areas of signal abnormality in the right femoral head.

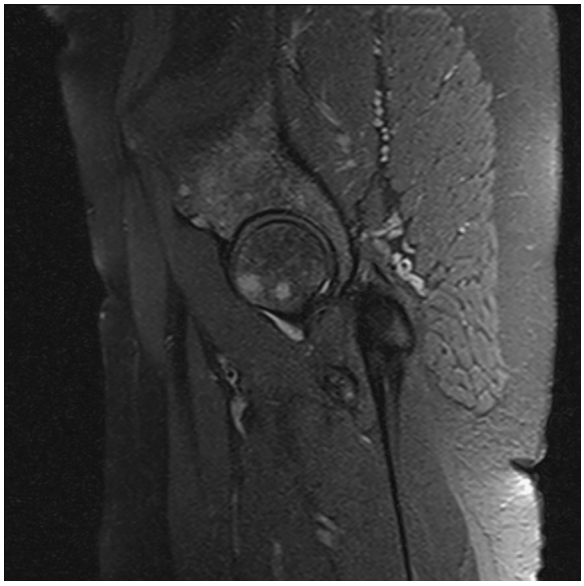

Fig. 4. T2W sagittal MRI showing multiple areas of intermediate to high signal in the right femoral head and pelvis.

At most recent follow-up 5 months post surgery the patient had an asymptomatic right hip with no change in component position (Fig. 10), and no progression of the lesions in her left hip and pelvis.

The patient's clinical and biochemical response to the myeloma treatment has been good, with a drop in the IgG Lambda monoclonal peak from 80 g/l to 30 g/l. Autologous haemopoetic stem cell transplant is planned after maximal response has been achieved with cyclophosphamide and dexamethasone.

2. Discussion

Sarcoidosis is an inflammatory disorder of unknown aetiology characterised by the presence of non-caseating granulomas, with no evidence of other known causes of granulomatous disease. It involves multiple organs, most commonly the lungs, lymph nodes, skin, and eyes, but may be present in any organ system. Involvement of the musculoskeletal system is well described in the literature. Up to 35% of patients will develop an acute or chronic polyarthropathy, and skeletal muscle granulomas occur in 80% of patients, but are largely asymptomatic. Skeletal lesions are typically bilateral and most commonly seen in the small bones of the hand and feet with associated skin lesions. Lesions of the axial skeleton and long bones are considered uncommon, and occur in approximately 5% of patients [2,3].

Large bone lesions may be painful or asymptomatic. Neither skeletal surveys nor radio-isotope scans have proved reliable in screening for skeletal lesions [4]. The lesions are typically small cysts with minimal involvement of adjacent soft tissue, but may present as an active lytic or sclerotic lesion or pathological fracture. MRI features are of indistinct or well marginated lesions of varying sizes with decreased intensity on T1 weighted images and increased intensity on T2 and proton-density fat-saturated weighted images, and lesions may enhance after contrast [5]. There are no pathognomonic features of skeletal sarcoidosis on