CLINICAL CASE - TEST YOURSELF Musculoskeletal Imaging

Uncommon skeletal manifestations of a rather common systemic disease

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PART A

A 32-year-old female patient was evaluated for pelvic and low back pain. Medical history was unremarkable, except from a laborious childbirth two months before. Full blood cell count and biochemical markers were normal. Autoantibody profile, inflammatory indicators, microbiologic stains and tuberculosis cultures were negative.

Imaging work up included radiograph and MR imaging of the pelvis and sacroiliac joints (Fig 1), followed by CT of the chest and abdomen (Fig 2).

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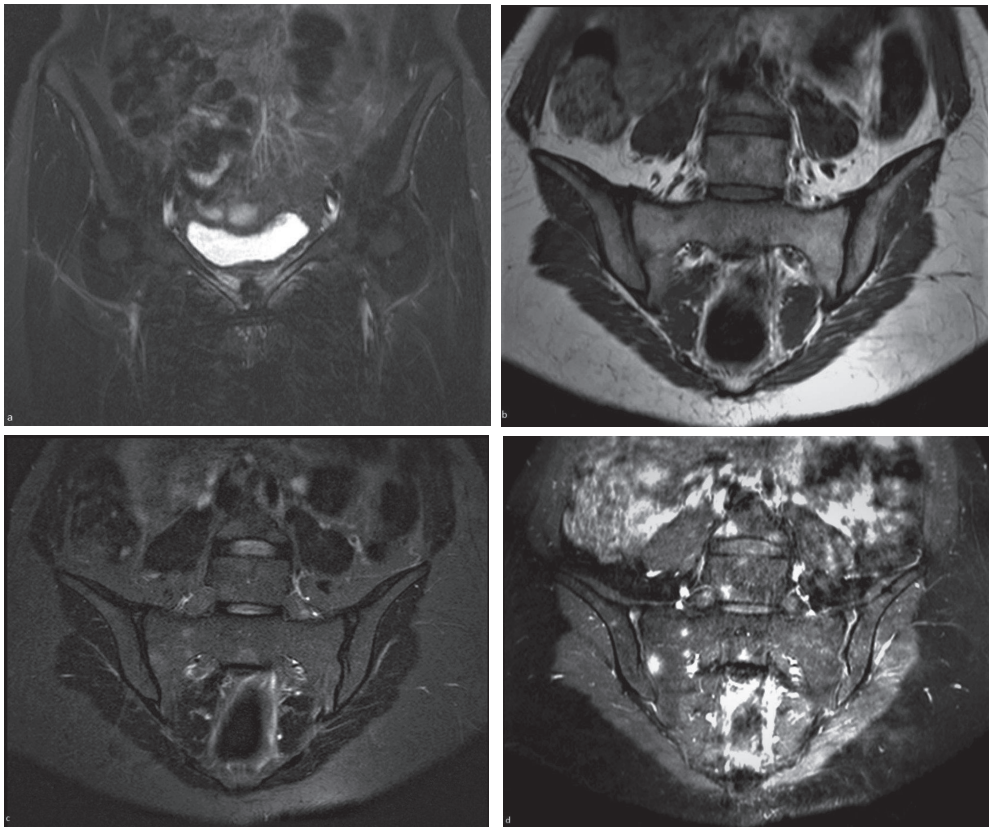

Fig 1. A: *Coronal STIR MR image* ***B:*** *T1w coronal oblique MR image* ***C:*** *Coronal STIR MR image at the same level as above* ***D:*** *Contrast-enhanced T1w MR image with fat suppression enhancement.*

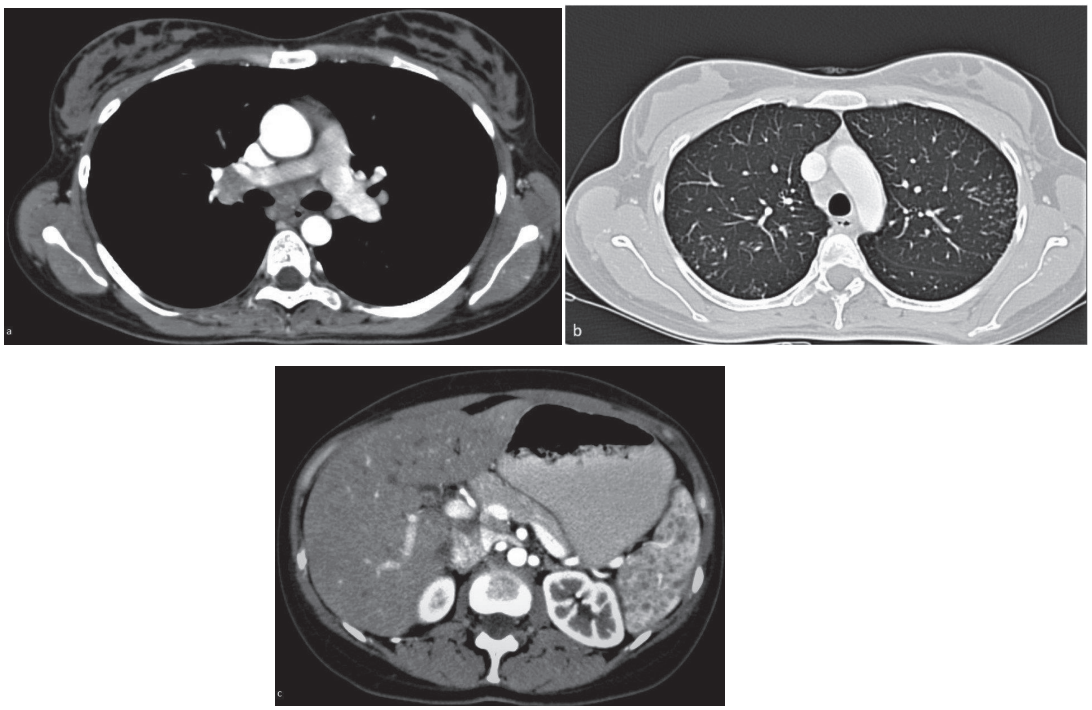

Fig 2. A: *Axial chest CT image (chest window) with IV contrast (arrows),* ***B:*** *Coronal reformatted chest CT image (lung window),* ***C:*** *Axial abdominal CT image with IV contrast.*

PART B

Diagnosis: Disseminated sarcoidosis

Sarcoidosis is a chronic multisystemic disease of unknown cause characterized by the growth of small collections of inflammatory cells (noncaseating granulomas) in any part of the body, most commonly the lungs and lymph nodes followed by liver, spleen and musculoskeletal tissues. Patients, most commonly females, are typically between the age of 20-40 years old [1]. In our patient sarcoidosis was diagnosed in the late postpartum period. It is not clear whether pregnancy relates to onset or flare up of the disease, as may occur in other autoimmune diseases [2].

In current case, small osseous pelvic and lumbar spine lesions were discovered incidentally on an MR imaging study of the pelvis which had been performed because of clinical suspicion of stress fractures. MR imaging showed bone marrow edema and periosteal reaction of pubic bones attributed to postpartum stress reactions which presumably accounted for patient's pelvic pain (Fig 1a). In addition, multiple small lesions, up to 1.3 cm, were disseminated in the bone marrow of pelvic bones and lumbar spine. The lesions had rather indistinct margins and exhibited moderately low signal intensity on T1w MR images (Fig 1b), were faintly visible on STIR images (Fig 1c) and presented significant, homogenous enhancement after intravenous administration of gadolinium (Fig 1d). There was no evidence of cortical destruction. Osseous metastases, lymphoma, granulomatous diseases including tuberculosis and sarcoidosis, were considered in the differential diagnosis so a bone scintigraphy, CT of the chest and abdomen and a bone biopsy, were requested. Chest CT revealed multiple bilateral mainly centrilobular lung nodules, along with enlarged, bilateral mediastinal and hilar lymph nodes (Fig 2 a, b). Despite bulky mediastinal lymphadenopathy the patient did not refer cough or any other respiratory symptoms. Abdominal CT displayed splenomegaly along with multiple hypodense foci, less than 5 mm in size, disseminated within the splenic parenchyma (Fig 2C) and one in the segment VI of the liver, measuring 6 mm. Noteworthy, abdominal CT -bone and soft tissue window- and an anteroposterior X-ray of pelvis and hips did not show any trabecular bone lesions in lumbar vertebrae and pelvis. Triple-phase bone scintigraphy showed increased non-specific focal radiotracer uptake in thoracic and lumbar vertebral bodies and pelvis. A trephine bone marrow biopsy of the iliac crest was negative for neoplastic disease but showed multiple noncaseating granulomas consistent with sarcoidosis.

Musculoskeletal sarcoidosis can affect up to one-third of patients, ranging from arthralgia to widespread destructive bone lesions [1, 3]. Sarcoidal arthropathy and myopathy are considered more common than osseous involvement. MR imaging can demonstrate non-specific arthropathy, tenosynovitis, bursitis, synovitis and multiple, bilateral intramuscular masses in sarcoidal myopathy [1]. Small tubular bone sarcoidosis, is one of the most familiar appearances of musculoskeletal involvement, almost exclusively associated with chronic systemic disease. It typically develops in a symmetrical pattern, most often affecting the second and third phalanges of fingers, preserving the metacarpophalangeal joints. Radiological findings include multiple cyst-like radiolucent lesions or widespread bone erosions along with pathologic fractures. MR imaging is not usually needed for diagnosis, but it can add information concerning marrow involvement and extent of granulomas [2].

With the widespread use of MRI, involvement of the axial skeleton is frequently revealed as incidental finding in patients with sarcoidosis, or even in patients with undiagnosed disease, imaged for other reasons, as occurred in our patient. According to more recent studies, spine followed by pelvis are the most frequent sites of bone disease [3,4]. The real incidence of spinal involvement may be underestimated as bone marrow lesions are usually occult on radiographs and CT but occasionally, they are seen as lytic or sclerotic lesions of the axial skeleton on CT [1,5].

MR imaging can demonstrate spinal, pelvic and large tubular bone lesions with clear or indistinct borders of varying sizes. These lesions usually have decreased signal intensity on T1w MR images and variably increased signal intensity on fluid-sensitive MR images. They may enhance after intravenous administration of gadolinium following a nonspecific pattern, as in our case. Cortical destruction is not a common feature.

Thoracic spine is the most frequently affected,

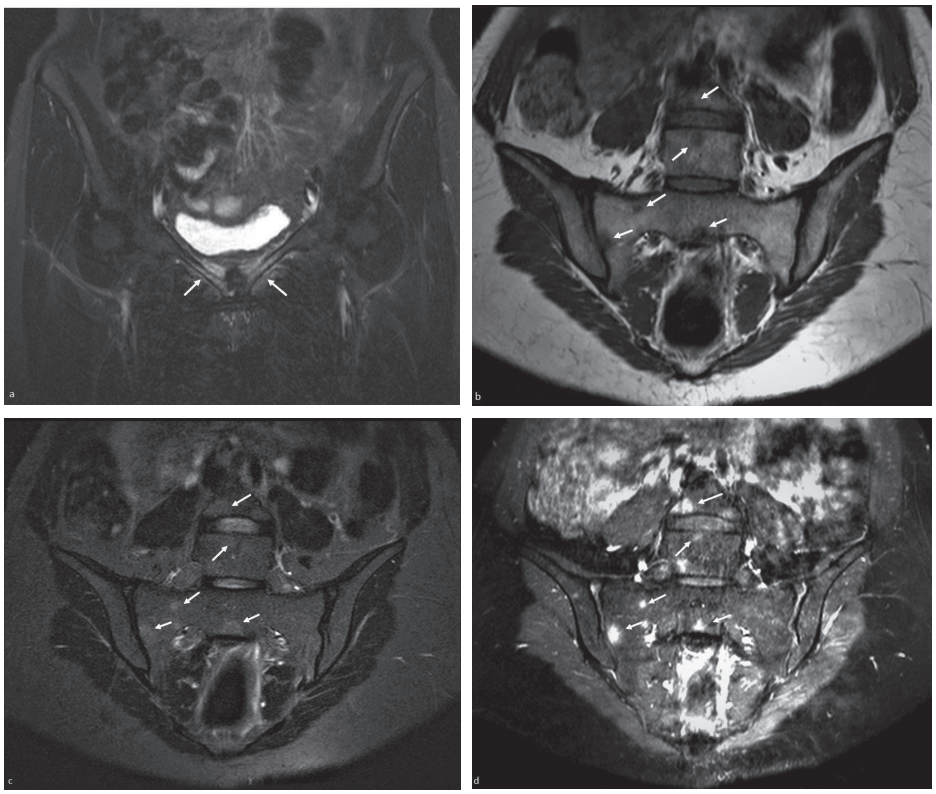

Fig 1. A: *A coronal STIR MR image shows bone marrow edema and periosteal reaction of the pubic bones (arrows). The pubic symphysis exhibits sclerosis of the subchondral bone.* ***B:*** *A T1w coronal oblique MR image shows hypointense foci with indistinct margins in the sacrum and L5 and L4 vertebral bodies (arrows).* ***C:*** *On a coronal STIR MR image at the same level as above these foci are barely visible.* ***D:*** *On a contrast-enhanced T1w MR image with fat suppression the lesions present significant enhancement (arrows).*

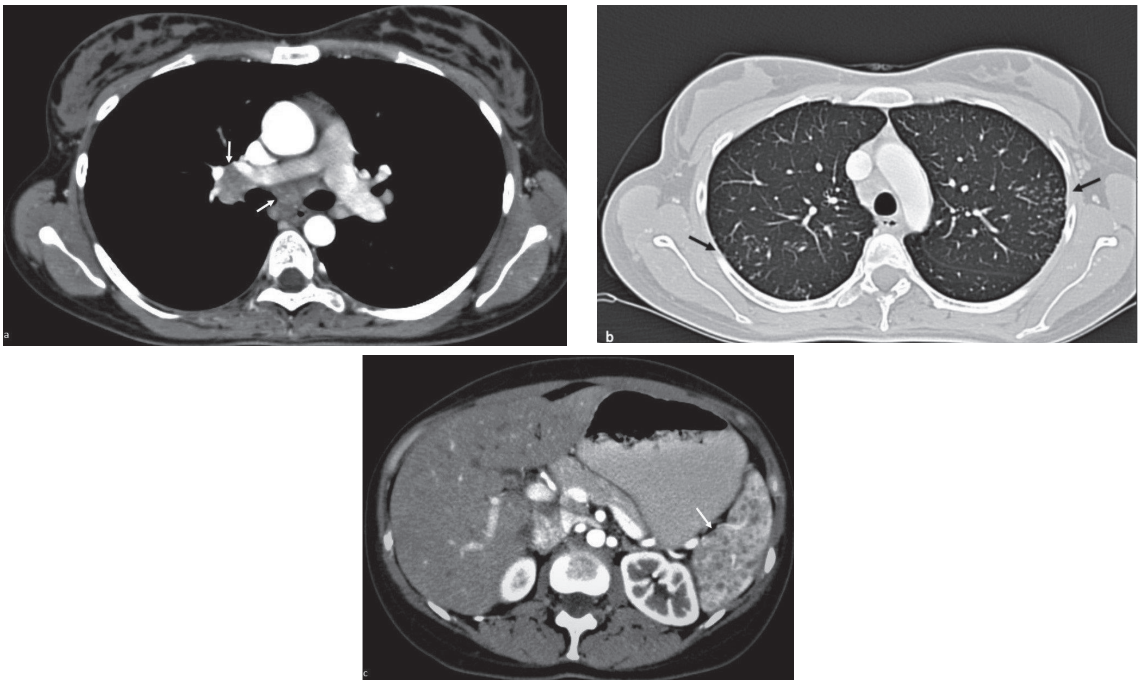

Fig 2. A: *An axial CT image (soft tissue window) depicts enlarged right hilar and subcarinal lymph nodes (arrows).* ***B:*** *An axial CT image (lung window) shows multiple nodules in peripheral bronchovascular distribution (arrows), especially in the left upper lobe.* ***C.*** *Abdominal CT, axial plane, shows multiple hypodense nodules disseminated within the splenic parenchyma.*

although any area of the spine can be involved. Patients with vertebral sarcoid are frequently symptomatic presenting with back pain [4]. MR imaging is sensitive for detection of the lesions in axial skeleton but is not specific even in the presence of proven systemic sarcoidosis. Spinal lesions share similar MR features with metastases or myeloma and histopathological proof is needed for differentiation. Moore et al found that sarcoid lesions in the bone marrow of axial skeleton may have intralesional fat and more indistinct margins, as in our case; moreover, posterior vertebral involvement with accompanying soft tissue mass is considered a reliable criterion for differentiation from vertebral metastases [5]. Intralesional fat has also been associated with involution of intramedullary sarcoid lesions [2]. Presumably, it was the presence of intralesional fat that made these focal lesions less apparent on STIR images in the current case. A ring or a double-ring pattern with fatty element in the central part has also been described in sarcoid lesions of pelvic bones and sacroiliac joints [6]. Diffusion-weighted MR imaging (DWI) might be useful to exclude malignant infiltration whereas DIXON sequences might be optimal to confirm the presence of intralesional fat.

Regarding prognosis, sarcoidosis frequently is a self-limited disease or responds well with corticosteroids. In aggressive cases leading to organ damage, immunosuppressive drugs have been used [4].

In conclusion, a high index of suspicion should be raised in young female patients with multifocal bone marrow lesions even without a previous diagnosis of sarcoidosis [5]. The final diagnosis should be based on a constellation of imaging findings especially from chest, consistent with sarcoidosis, along with consideration of the sex and age of the patient or on histology. **R**

KEY WORDS sarcoidosis, MR imaging, spine, pelvis

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