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CASE STUDIES

Presentation of Ewing's sarcoma as unilateral sacroiliitis

Vikram Muralidhar Haridas[1]*, S Chandrashekara[2], Balebail Gopalakrishna Dharmanand[3]

[1, 2, 3] Chanre Rheumatology and Immunology Center, Basaweswaranagar, Bangalore, India

Abstract

Presence of sacroiliitis (SI) is one of the hallmarks for diagnosing seronegative spondyloarthropathy, especially ankylosing spondylitis. In certain cases, the occurrence of erosions around sacroiliac joint due to other causes can mislead the diagnosis. We are discussing here a rare case of Ewing's sarcoma, which clinically presented as SI.

Case report

A 20-year-old man presented with low back pain in the right lumbar region, which was classically inflammatory in presentation. Family and personal history of the patient did not report enthesitis, oral ulcers, genital ulcers or redness of eyes. Additionally, there was no history of loss of appetite or weight loss. The laboratory findings were as follows: ESR-100 mm/hr (normal<20), CRP-59 u/dl (normal<6), and alkaline phosphatase-100 IU/L (normal <306). X-ray of the pelvis showed erosive right-sided sacroiliitis (Fig 1a). CT results indicated that the articular margins of right sacroiliac joint (SI) were irregular and indistinct with marked subchondral marrow edema more on the iliac bone. Mild adjacent soft tissue edema was also noted, suggestive of right saroiliac arthritis (Fig 2a). Biopsy of the SI joint showed the occurrence of chronic non-specific inflammation.

Mantoux test and human leukocyte antigen (HLA) B27 tests were negative. Diagnosis of tuberculosis (TB) was suspected. Since tuberculosis is a common cause of unilateral sacroiliitis in India, the patient was started on antitubercular treatment (4 drugs for 3 months). However, the treatment response was minimal. Repeated X-ray of SI joint showed the presence of progressive lesion (Fig 1b). Eventually, the patient developed loss of sensation over the buttock and on the posterior aspect of thigh with severe radiating pain. His MRI (Fig 2b) showed that hypointensity lesions on the T1 weighted sequence became heterogeneously hyperintense relative to all other sequences in the right iliac bone, which extended onto the acetabulum, the ischium, and the adjacent right sacral ala. This was suggestive of a metastatic bone disease. Bone scan indicated increased uptake in the right iliac bone, skull, and lumbar vertebral bodies. Features observed in repeated biopsy of the SI joint were suggestive of small round cell tumor, the Ewing's sarcoma of iliac bone.

Discussion

Ewing's sarcoma (ES) is the second most prevalent malignant tumor of bone in children and adolescents following osteogenic sarcoma.[1] Around 90% of the patients with this malignancy are of less than 30 years of age.[2] The etiology of the disease is unknown. However, in 85% of the affected subjects, the pathognomonic chromosomal translocation of t(11:22) is found. Pain (90%) followed by swelling (70%) is the most commonly noted symptom of ES.[3] Akin to the previous findings, the current study also reported the presence of systemic manifestations such as weight loss, fever, and increased sedimentation rate.[1] Although the disease can affect almost any bone and soft tissues, femur (23%) followed by the pelvis (17%) are the commonest sites. Diagnostic delay is common and this could be ascribed to the presence of non-specific symptoms, gradual development of the tumor, and the deep location, especially in cases of pelvic tumors.[1, 2]

Fig 1a: X-ray of SI joint showing subarticular sclerosis on ilial and sacral articular surface suggestive of right sided sacroiliitis. Fig 1b: X-ray of SI joint showing diffuse osteosclerosis of right iliac bone with ill-defined areas of osteolysis with subarticlar sclerosis on the sacral side.

Figure 1a

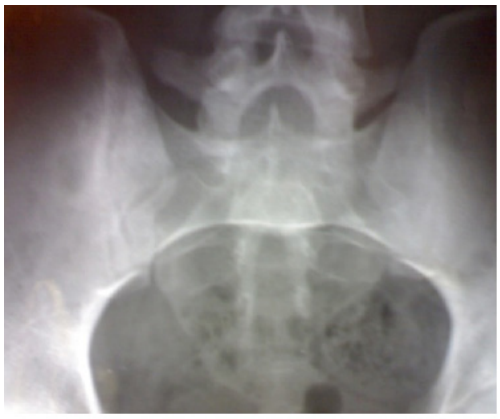


Figure 1b

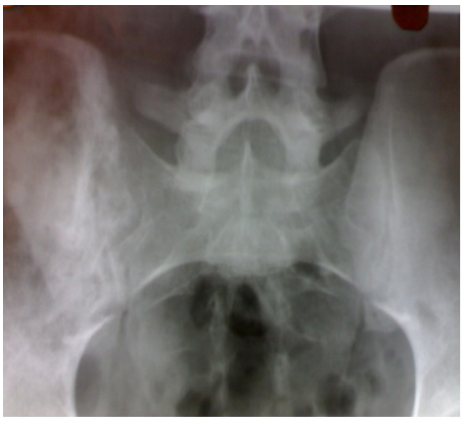


Fig 2a: Axial plain CT of pelvis showing irregular osteolysis of ilial bone and sacral articular surface of right SI joint with adjacent soft tissues. Fig 2b: Short T1 inversion recovery (STIR) MRI image of SI joint showing altered marrow signal in ilial and sacral articular surface of right SI joint, abnormal soft tissue is noted in the periosteal region of iliac bone and superior to right SI joint.

Figure 2a

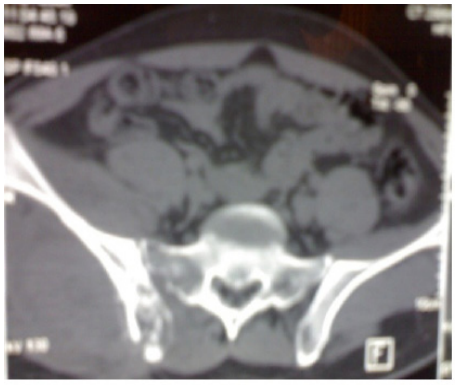
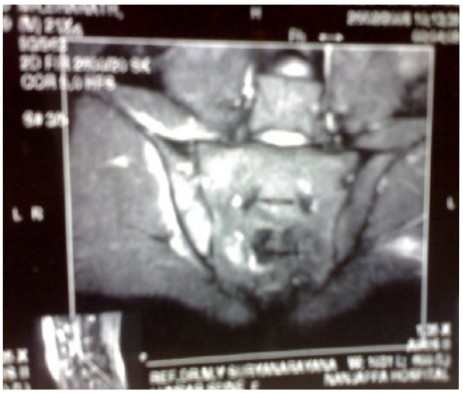


Figure 2b



Radiologically, it was classically described as a central, diaphyseal, lytic tumor, which is often permeative and has a lamellated or `onion skin' periosteal reaction affecting a long bone, and associated with soft-tissue mass. The bone lesions are usually lytic, but may be sclerotic or mixed. Most of the lesions are diaphyseal or metadiaphyseal. However, this classic description is no longer valid. The classic description has only 5% sensitivity, but 99% specificity and 20% prevalence, in an analysis of the plain radiographic appearance of ES of bone.[4] The tumor may be misdiagnosed as benign disorders. As far as we are aware, the number of reported cases of ES of the ilium mimicking sacroiliitis is three.[5, 6] ES of the tibia may mimic osteomyelitis.[7] It may present as postpartum back pain, monoarthritis, and spondylolisthesis.[8, 9, 10] Hence, the plain radiographic abnormalities of ES are non-specific and it may mimic benign disorders. CT scan, demonstrating the pattern of bone destruction and the associated soft-tissue mass, is useful in defining the tumor of ES.[5] The sensitivity of MRI is reported to be more, when compared

to CT, in displaying soft-tissue involvement and bone marrow metastases.[6] Definitive diagnosis of the disease can be made by an open biopsy and histological examination in conjunction with immunochemistry and cytogenetics. Multi-drug chemotherapy combined with surgery and/or radiotherapy is the preferred treatment strategy for ES. Surgery is preferred over radiotherapy, if the tumor is resectable and the function following resection is predicted to be reasonable. The five-year survival rate of patients presenting without metastases is 50-75%.[1,6] In cases with metastasis, the two-year survival is 39%.[6] The factors influencing the prognosis are the location and the size of the tumor, presence of metastasis, and the response to chemotherapy. Prognosis is poor if the primary tumor is in the pelvis and the sacrum, and if the tumor size is larger than 8-10 cm.

In conclusion, ES is a highly malignant tumor of bone and soft tissues affecting children and adolescents. Owing to the non-specific clinical manifestations and radiographic abnormalities, it can be confused clinically and radiologically with infection or inflammation. ES affecting the ilium could be misdiagnosed as unilateral sacroiliitis.

Alternative diagnosis should be considered in unilateral sacroiliitis patients, especially in those with the following history or clinical findings: not responding to adequate dose of analgesics or anti-tubercular drugs, with newly onset neurological deficit, inconclusive biopsy, with symptoms disproportionate to simple sacroiliitis and having very high inflammatory parameters.

Competing interests
The authors declare that they have no competing interests.

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* Correspondence: Dr.Vikram Muralidhar Haridas, Arthritis Super Speciality Center, Hubli, India
haridasvikram@yahoo.co.in

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