Diffuse Skeletal Hemangiomatosis Mimicking Sacroiliitis

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Hemangiomatosis of the skeletal system is a rare disease without specific symptoms and signs. We describe a 20-year-old patient with low back pain, whose plain radiographs of sacroiliac (SI) joint showed irregular sclerotic lesions. The patient was finally confirmed with skeletal hemangiomatosis by magnetic resonance imaging (MRI) and excisional biopsy of the lesion. The present case suggests that if patients with abnormal lesions of the SI joint in the plain radiographs do not have typical inflammatory back pain, advanced imaging is required to make an accurate diagnosis. Our case also emphasizes the importance of MRI and biopsy in establishing the diagnosis.

Key Words. Hemangiomatosis, Sacroiliitis, Magnetic resonance imaging

Introduction

Skeletal hemangiomatosis defined as a neoplastic entity arising from blood vessels, constitutes <1% of all primary bone neoplasms (1). Almost 80% of cases involve the axial skeleton, especially the vertebrae, and are generally asymptomatic (2). We present a patient with skeletal hemangiomatosis mimicking sacroiliitis.

Case Report

A 20-year-old man presented at the rheumatology outpatient clinic having suffered from intermittent low back pain for 2 weeks. The pain was neither worse after long periods of rest nor relieved by exercise. The pain was only exacerbated by active physical exertion without radiation or diurnal variation, that was not consistent with inflammatory back pain. He had no personal history for hospitalization, trauma, surgery, or other disease. There was not uveitis and enthesisopathy. Familial history was unremarkable. He had neither pain after direct compression on over sacroiliac (SI) joint nor limitation of hip or SI joint. Gaenslen test and Patrick test were negative. Neurologic exam demonstrated no abnormal findings.

The HLA-B27, rheumatoid factor and purified protein derivative (PPD) skin tests were negative. Erythrocyte sedimentation rate of 2 mm/h, C-reactive protein of 0.21 mg/dL, alkaline phosphatase of 84 U/L, calcium of 9.6 mg/dL, phosphorus of 2.9 mg/dL, were all within normal limits. Other laboratory investigations, including complete blood count and serum biochemistry were also normal.

X-ray examination revealed irregular sclerotic lesions of the left SI joint, and sclerotic cystic lesions on the both sacral wing and left ilium (Figure 1A), as well as sclerotic changes of the vertebral endplates in the lumbar spine (Figure 1B). Pelvic computed tomography (CT) revealed multiple subchondral cysts of the sacral wing and left ilium (Figure 1C), and subchondral and cortical intramedullary osteosaceous cysts of the SI joints were evident by MRI (Figure 1D).

In an L-spine MRI there were multiple lesions that were hypointense on T1-weighted images and hyperintense on T2-weighted images, and these were scattered between the T9 and S3 vertebrae (Figure 1E). In fat suppression images, the lesions were of high signal intensity, and were markedly enhanced after gadolinium administration (Figure 1F). The le-
sions had the characteristics of highly vascular fatty tumors. These radiologic findings suggested diffuse skeletal hemangiomatosis. To evaluate the involvement of other organs, we performed abdominal and chest CT, which revealed multiple irregularly-shaped fat-containing lesions in the thoracolumbar vertebrae, sacrum, and left iliac bone as well as multiple cystic masses in the spleen.

To exclude other bone malignancies, we performed an excisional biopsy of the left pelvic bone 4 cm proximal and 4 cm posterior to the anterior superior iliac spine. The histological finding of bone lesion shows bony trabecula with dilated blood-filled spaces and some medium-sized abnormal
Figure 2. Histologic appearance of a bone lesion (hematoxylin-eosin ×200 (A) and ×400 (B)) shows bony trabecula with some medium-sized abnormal vessels in fatty marrow spaces.

vessels in fatty marrow spaces (Figure 2), with no evidence of malignancy. The final diagnosis was hemangiomatosis of diffuse skeletal bone. The patient has been followed up after diagnosis without medication and there has been no worsening of symptoms.

**Discussion**

Hemangioma is defined as a neoplastic entity which arises from blood vessels. Although skin is the most common primary site, any organ including bone can be affected. It can occur at all ages but is most common in the fourth and fifth decades of life, and has a female preponderance (female : male = 3 : 1) (3). Its cause is unknown but a history of trauma seems to be involved in some cases in the literature (4).

Involvement of the SI joints is a hallmark of axial spondyloarthritis (SpA) or ankylosing spondylitis (AS) (5). Conventional radiographs remain the most widely accepted and available screening method for SpA. In our case, the irregular sclerotic lesions of the left SI joint in plain radiography were suspected to be axial SpA, but clinical history and physical examination were nonspecific. Because inflammatory back pain is not a highly specific indicator of sacroiliitis, there is a need for valuable imaging techniques (5). Among the imaging techniques, MRI is highly sensitive for detecting bone lesions, but biopsy is often required for definitive differential diagnosis including metastatic cancer (6).

Hemangioma involving the axial skeleton are commonly asymptomatic and may remain undetected for long time (7). They are often discovered incidentally during a radiographic study, whereas hemangioma of the appendicular skeleton are often symptomatic. Very few cases of hemangioma affecting the pelvic bones have been reported (8). No prior case was reported about hemagioma affecting the SI joint. Hemangiomas in the peripheral and pelvic bones are usually symptomatically associated with pain, soft tissue swelling and pathologic fracture (9). Our patient complained of pain in his back for a duration of approximately two weeks after active physical exertion.

Intraosseous hemangioma may have a variety of radiological appearances. Due to its lack of specific features and rarity, it is often difficult to diagnose radiologically pre-operatively (9). CT usually shows well-margined focal areas of decreased bone attenuation with a polka-dot or honeycomb appearance (7). On MRI, signal intensity on T2 weighted sequences is frequently high, while signal intensity on T1-weighted images varies depending on the vascular and fat components of the lesion (7).

The histologic pattern of osseous hemangiomas is characterized by proliferation of anomalous thin-walled blood vessels and sinuses lined by endothelium within the thickened and vertically oriented trabeculae of bone (9). The dilated vascular channels are set in a stroma of fat. The ratio of fat to vascular tissue wrapped between the pillars of bone determines the signal intensity on MRI images (10).

Although special treatment is not required for asymptomatic and small lesion, surgical treatment such as curettage or complete surgical resection and bone grafting is indicated for symptomatic ones. Radiation therapy to ablate the venous channels forming the lesion is reserved for subtotal resection or for an unresectable lesion in a symptomatic patient (8,11). In our case, pain was not severe and no worsening of symptoms without medication. So, he has been followed up without any treatment.
Inflammation of sacroiliac joints is a characteristic feature of patients with SpA including AS (5). Specially, symmetrical sacroiliitis is found in more than 90% of AS patients, and the asymmetric involvement of the SI joints frequently indicates other forms of SPA, most commonly psoriatic arthritis (12). Moreover, asymmetrical sacroiliitis can be shown in other non-rheumatic diseases such as infectious diseases, tuberculosis, sarcoidosis, hyperparathyroidism, and malignancy.

MRI is sensitive imaging method for detecting inflammatory changes and is helpful for differential diagnosis of sacroiliitis. The estimated sensitivity and specificity of MRI in the diagnosis of sacroiliitis are approximately over 90% (13). Structural damage and chronic lesions, such as fatty degeneration and erosions, are best visualized on T1-weighted images. Degenerative changes of the sacroiliac joints are characterized by irregularity and narrowing of the articular space, periaricular bone sclerosis, subchondral cysts, and the absence of inflammatory features such as erosions (14). The active inflammatory lesions are best visualized on STIR images or fat-suppressed T2-weighted. Inflammatory sacroiliac lesions of SpA is characterized as bone marrow edema on subchondral bone surfaces of SI joints, but do not cross anatomic borders. In contrast, infectious sacroiliitis is visualized as unilateral high signal intensity of the subchondral bone surfaces of the sacroiliac joint on STIR images and contrast-enhanced T1-weighted images with surrounding soft tissue. When the edema is unilateral, infectious sacroiliitis must be excluded (15).

In our case, unilateral sacroiliitis is presented as subchondral and cortical intramedullary osseous cysts of the SI joints without bone marrow edema in MRI. If MRI of the SI joints is insufficient of diagnosis, biopsy is often required for confirming diagnosis.

The involvement of SI joints may be mistaken for the inflammation seen in SpA. This case indicates that diffuse skeletal hemangiomatosis can be expressed as a sacroiliitis. Therefore, in cases with non-specific clinical manifestations, further evaluations by MRI or biopsy is needed for accurate diagnosis.

Summary

Non-rheumatologic diseases such as diffuse skeletal hemangiomatosis should be considered when patients with abnormal lesions of SI joint in plain radiographs do not have typical clinical symptoms and signs. It is recommended that further evaluations by MRI or biopsy should be undertaken to make an accurate diagnosis.

References