SAPHO: A Challenging Diagnosis in a Young Woman With Recurrent Osteomyelitis

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SAPHO: A Challenging Diagnosis in a Young Woman With Recurrent Osteomyelitis

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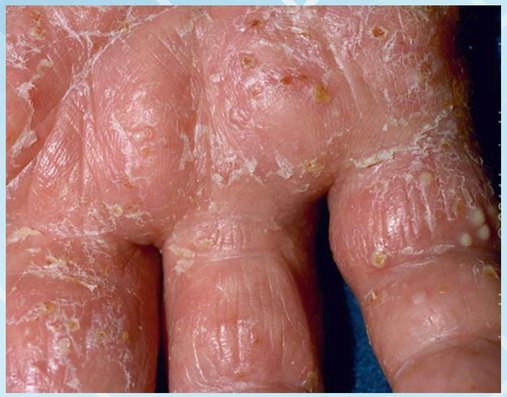
INTRODUCTION

SAPHO, known as synovitis, acne, pustulosis, hyperostosis and osteitis is a rare syndrome and has been estimated to occur in 1 in 10,000 in the Caucasian population. It is a disease of unknown pathogenesis which can present with swelling and bony tenderness as well as osteitis affecting multiple bones.

CASE DESCRIPTION

A 25-year-old female with a history significant for severe hidradenitis suppurativa requiring multiple drainages and frequent antibiotics, mandibular osteomyelitis due to actinomyces, and culture negative osteomyelitis of the clavicle presented with left elbow pain and swelling. This was evaluated with MRI demonstrating a septic joint with surrounding myositis and uncertain osteomyelitis. Incision and drainage including bone biopsy was performed; pathology demonstrated osteomyelitis but cultures were negative. The patient was treated with 4 weeks of intravenous antibiotics. After discharge, the patient returned with recurrent left elbow pain; MRI showed worsening osteomyelitis and blood work showed increasing inflammatory markers. There was concern for osteosarcoma; however pathology again revealed acute osteomyelitis with recurrent negative cultures. With the constellation of symptoms including hidradenitis suppurativa and recurrent culture-negative osteomyelitis particularly including the clavicle, the diagnosis of SAPHO was established. The patient was started on anti-inflammatory medications and methotrexate with eventual progression to adalimumab.

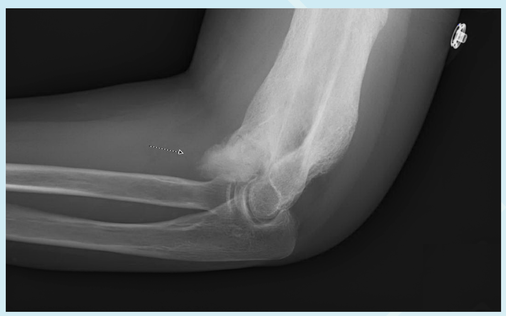


Palmoplantar pustulosis[2]

Diagnostic criteria proposed by Benhamou for SAPHO syndrome diagnosis[1,2]

1. Osteoarticular manifestations in severe acne
2. Osteoarticular manifestations in palmoplantar pustulosis
3. Hyperostosis with or without dermatosis and
4. Recurrent multifocal chronic osteomyelitis involving the axial or peripheral skeleton, with or without dermatosis

SAPHO: synovitis, acne, pustulosis, hyperostosis osteitis



X-ray of the left elbow of our patient read as significant soft tissue calcifications.

DISCUSSION

SAPHO is a diagnosis of exclusion once infectious osteomyelitis, malignancy, and other autoimmune conditions have been ruled out. After these exclusions, the diagnosis can be made if the patient has severe acne, palmoplantar pustulosis, or any case of hyperostosis or recurrent multifocal osteomyelitis with or without dermatologic features. Most commonly affected bones are the anterior chest wall (sternum, clavicle, or sternocostoclavicular joints), the vertebrae, sacroiliac joints and long bones. There are no specific tests for SAPHO, but elevated inflammatory markers may be seen. Plain films and bone scan may help identify additional areas of involvement. Tissue sampling and joint aspiration can help rule out infectious osteomyelitis.

Treatment consists of NSAIDs and for refractory cases: prednisone, methotrexate, bisphosphonates and biologics. Increased awareness of SAPHO can help clinicians arrive at a faster diagnosis with hope of reducing the incidence of osteomyelitis and decreasing unnecessary antibiotic administration and surgical procedures.

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