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CASE FOR DIAGNOSIS

Palmar-Plantar Pustules in a Patient with Sternoclavicular Pain

Pústulas palmoplantares en un paciente con dolor esternoclavicular

Clinical History

A 40-year-old woman with no relevant history was sent to our outpatient clinic for pruriginous pustules that had appeared 5 days earlier on the palms and soles. The patient reported no systemic symptoms, except for mild tenderness over the upper part of the anterior chest wall. She also denied any symptoms of infection in the days before the symptoms developed.

Physical Examination

Examination revealed sterile pustular lesions a few millimeters in diameter on an erythematous base, situated on the palms and plantar arches; some of the lesions were confluent (Figure 1).

The patient also reported pain on palpation of the sternoclavicular joints.

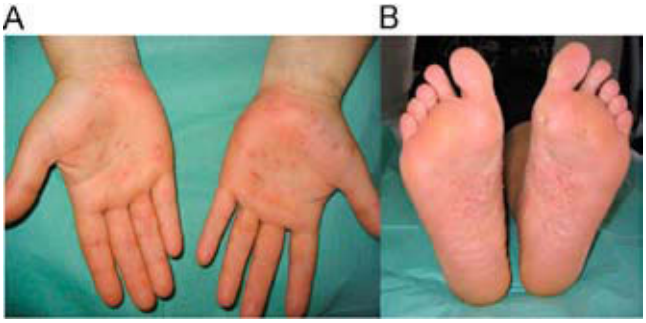

Figure 1 A, Sterile pustules affecting the palms. B, Pustular lesions on the soles.

Additional Tests

Blood tests including complete blood count, biochemistry, erythrocyte sedimentation rate, autoimmunity, and HLA-B27 were normal or negative, as were a pharyngeal swab, chest x-ray, and microbiological culture of the pustules.

Bone scintigraphy with technetium 99 showed increased uptake in the sternoclavicular and sacroiliac joints (Figure 2).

Diagnosis

SAPHO syndrome.

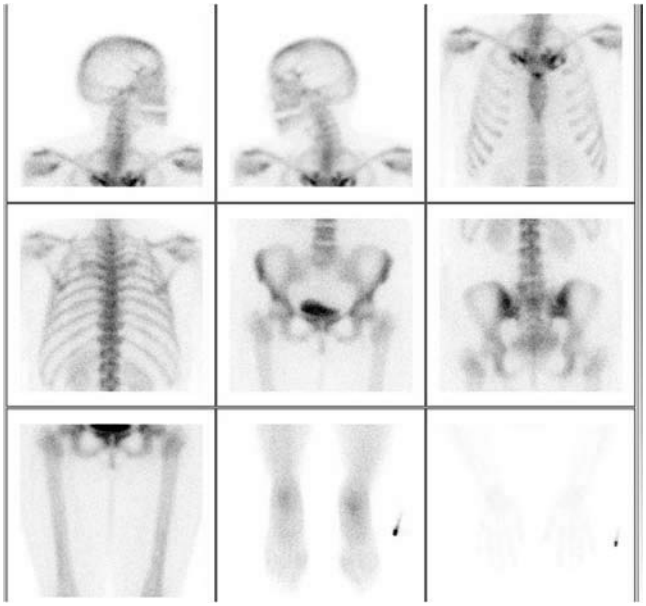

Figure 2 Bone scintigraphy with technetium 99 showing hot spots over the sternoclavicular and sacroiliac joints.

What Is Your Diagnosis?

Course and Treatment

Treatment was started with high potency topical corticosteroids under occlusive dressings and bathing with saline solution for the skin lesions, and oral indomethacin for the joint manifestations.

The clinical course was favorable and the skin lesions had healed almost completely 3 weeks after their onset.

Comment

SAPHO syndrome, described in 1987 by Charmot et al, refers to a group of skin and joint manifestations. The acronym SAPHO describes the main characteristics of this syndrome: synovitis, acne, pustulosis, hyperostosis, and osteitis.[1]

The incidence and prevalence are unknown. The condition has been described most commonly in Japan and northwest and southwest Europe, mainly in children and adults. There is an apparent predominance of cases presenting with acne in men and a predominance of palmar-plantar pustulosis in women.[2]

The etiology is unknown, although a number of theories have been proposed.[3,4] Some authors explain SAPHO as a reactive arthropathy possibly related to *Propionibacterium acnes*, others have related it to a low-virulence microorganism, and a third group consider it to be a type of seronegative spondyloarthropathy.

The most characteristic clinical manifestation is pain and swelling of the anterior chest wall resulting from involvement of the sternoclavicular, costosternal, costochondral, or manubriosternal joints.[3] The condition generally presents symmetrically and bilaterally, and occurs as flare-ups. Patients may complain of low back pain if the sacroiliac articulations are affected, but, unlike other spondyloarthropathies, this tends to be unilateral. Peripheral arthritis is uncommon and is characterized by oligoarthritis or monoarthritis, with the knee the most commonly affected joint. It can also affect the flat bones, especially the mandible.

Skin involvement is variable and includes palmar-plantar pustulosis, acne conglobata or fulminans, hydradenitis suppurativa, pustular psoriasis, dissecting cellulitis of the scalp, Sweet syndrome, and Sneddon-Wilkinson disease.[3]

Diagnosis is based on clinical manifestations combined with compatible imaging studies; a scintigraphy image showing a bull-horn configuration of the sternoclavicular joint is relatively specific to the syndrome.[5] In the blood tests there may be elevation of the acute phase reactants and HLA-B27 can be positive and rheumatoid factor negative in 12% to 30% of cases.

The differential diagnosis includes bacterial osteomyelitis, sclerosing osteitis, osteoarthritis, aseptic osteonecrosis of the medial epiphysis of the clavicle, primary bone tumors, and bone metastases.

Treatment for SAPHO syndrome includes nonsteroidal anti-inflammatory drugs, corticosteroids, colchicine, sulphasalazine, dapsone, methotrexate, intravenous pamidronate, and antitumor necrosis factor-α agents.[6]

Conflicts of Interest

The authors declare no conflicts of interest.

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