Palmoplantar pustules and osteoarticular pain in a 42-year-old woman

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CASE SUMMARY

History

A 42-year-old Caucasian woman was evaluated at the National Institutes of Health (NIH) Clinical Center for bilateral palmoplantar pustular plaques and joint pain. Her dermatologic history was notable for significant acne involving her face and back in her youth. At age 22 years, the patient developed a recurrent rash with pustules on the bilateral palms, which progressed to involve her bilateral soles and was associated with pruritus, skin pain, and fissuring. Treatment with topical steroids, coal tar, psoralen plus ultraviolet A, and ultraviolet B therapy was of limited benefit.

At age 29 years, she developed pain, swelling, and morning stiffness involving her right sternoclavicular joint, fingers, hips, and knees that improved with physical activity. Whole-body scintigraphy revealed increased tracer uptake in the right sternoclavicular joint and left knee. Bone biopsy specimen of the right sternoclavicular joint showed no evidence of tumor or infection. The patient was treated sequentially with nonsteroidal anti-inflammatory drugs, methotrexate, sulfasalazine, and intralesional steroids with modest improvement in pain and stabilization of right sternoclavicular joint swelling.

Abbreviations used:

CRMO:	chronic recurrent multifocal osteomyelitis
IL:	interleukin
NIH:	National Institutes of Health
SAPHO:	synovitis-acne-pustulosis-hyperostosis-osteitis

The patient was a current nonsmoker, but had a 50 pack-year smoking history. Her family history was remarkable for several first- and second-degree relatives with palmoplantar pustulosis or plaque psoriasis.

Physical examination

On examination at the NIH, discrete pink plaques with scattered pustules and overlying coarse scale were noted on the medial aspects of bilateral soles (Fig 1). Coarse adherent white scale was present on the left elbow and fine scale was noted on the palmar surface of the right thumb. The right sternoclavicular joint was notably enlarged and tender to palpation. The left second metacarpophalangeal joint was notable for swelling and synovial thickening. There was tenderness without swelling of the sternomanubrial, left sternoclavicular, bilateral shoulder, left

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third metacarpophalangeal, right second to fourth metatarsophalangeal, and medial right knee joints.

Histopathology

Histopathologic examination of a punch biopsy specimen obtained from the right sole revealed hyperparakeratosis, acanthosis, loss of the granular layer, intraepidermal and subcorneal neutrophils, and neutrophilic pustules (Fig 2).

Significant diagnostic studies

Laboratory investigations were notable for elevated high-sensitivity C-reactive protein of 6.6 mg/L. Noncontrast whole-body magnetic resonance imaging revealed short tau inversion recovery (STIR) enhancement in the soft tissue adjacent to the left medial clavicle and small bilateral knee effusions.

Diagnosis

We diagnosed synovitis-acne-pustulosis-hyperostosis-osteitis (SAPHO) syndrome with palmoplantar pustulosis, hyperostosis of the right sternoclavicular joint, and inflammatory arthritis.

DISCUSSION

SAPHO syndrome is a rare, chronic, autoinflammatory disorder that includes a spectrum of recurrent osteoarticular and neutrophilic cutaneous manifestations. The prevalence of SAPHO syndrome has been estimated at 0.04%,[1] however it may be underreported. A female predominance has been reported and disease may present at any age, however onset is most frequently in childhood or early adulthood.[2] A subset of SAPHO syndrome, known as chronic recurrent multifocal osteomyelitis (CRMO), predominates in children, although it has been reported in adults.

Greater than 60% of individuals with SAPHO syndrome and 30% with CRMO develop cutaneous manifestations.[2-5] The cutaneous manifestations of SAPHO syndrome and CRMO consist of neutrophilic and pustular dermatoses and may present concurrently with, before, or after the onset of musculoskeletal symptoms.[6] The most frequent dermatologic finding is palmoplantar pustulosis, representing 50% to 75% of SAPHO syndrome skin manifestations.[7] Other skin manifestations include severe acne (ie, acne conglobata and acne fulminans), pustular psoriasis, psoriasis vulgaris, hidradenitis suppurativa, pyoderma gangrenosum, pyoderma vegetans, erythema nodosum, Sweet syndrome, and subcorneal pustular dermatosis.[2,8-13] Acne and hidradenitis suppurativa occur more commonly in males in the setting of SAPHO syndrome.[4,14]

The hallmark feature of SAPHO/CRMO is osteoarthropathy, characterized most prominently by hyperostosis and osteitis. Onset of osteoarticular manifestations is usually insidious.[15] The most frequently affected skeletal site in adults is the anterior chest wall (65%-90%). Spinal involvement (33%), sacroiliitis (13%-52%), long bone involvement (5%-10%), mandibular lesions (1%-10%), and synovitis in peripheral joints (30%) can also occur.[7,16,17] In contrast, long bones represent the most frequent site of involvement in children.[3,18-20]

Other systemic features are uncommon, however fever and elevated inflammatory markers can be seen.[4] Inflammatory bowel disease, most often Crohn's disease, has been reported in up to 10% of patients with SAPHO syndrome.[4]

The disease course of SAPHO syndrome is highly variable. Approximately half of cases have a chronic relapsing-remitting course. The remainder exhibit either monophasic disease or a relapsing-remitting course for several years followed by remission.[2] Predictors of chronic disease course include female sex, anterior chest wall involvement, peripheral arthritis, dermatologic manifestations, and markedly elevated acute phase reactants.[2]

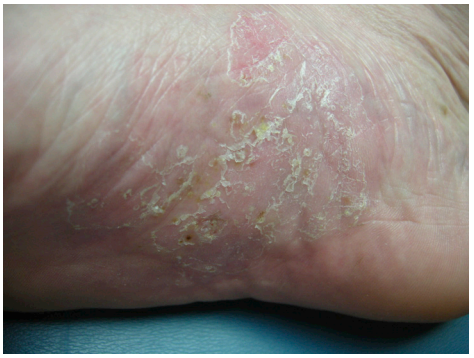

Fig 1. Palmoplantar pustulosis. Pink plaques with scattered pustules and overlying thick coarse scale on the medial aspect of the right sole.

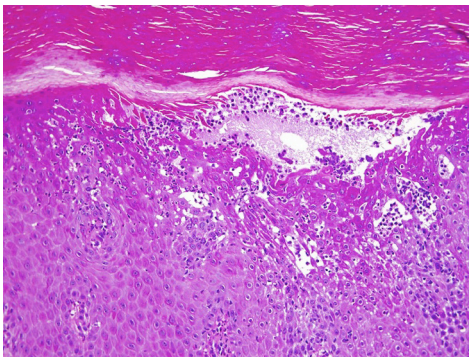

Fig 2. Histopathology of palmoplantar pustulosis. Hyperparakeratosis, acanthosis, loss of the granular layer, intraepidermal and subcorneal neutrophils, and neutrophilic pustules.