PALMOPLANTAR PUSTULOSIS ASSOCIATED WITH OSTEOARTICULAR PAIN

SIBEL ALI *,****, ANCA COJOCARU*, IRINA TUDOSE***, OLGUȚA ANCA ORZAN*,**

Summary

Palmoplantar pustulosis (PPP) is a rare chronic inflammatory disease, characterized by the recurrent appearance of sterile pustules. PPP is one of the skin manifestations that can occur in patients with SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, osteitis), a rare syndrome, characterized by the association of various forms of arthropathy and neutrophilic skin conditions. In this paper, we present the case of a patient with palmoplantar pustulosis and osteoarticular pain in the left hemithorax and left sternocostal joint that raised the suspicion of SAPHO syndrome.

Keywords: *palmoplantar pustulosis, SAPHO syndrome, neutrophilic dermatoses*

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Introduction

Palmoplantar pustulosis (PPP) is a rare chronic inflammatory disease, characterized by the recurrent appearance of sterile pustules up to 10 mm, associated with erythema, hyperkeratosis, scaling, and painful erosions, located on the palms and soles. The distribution is generally bilateral, and the symptoms include pruritus and burning sensation.

PPP is one of the skin manifestations that can occur in patients with SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, osteitis), a rare syndrome, characterized by the association of various forms of arthropathy and neutrophilic skin conditions. SAPHO syndrome arthropathy usually involves the anterior chest wall, with pain, tenderness, and edema in the sternum and regional joints.

In this paper, we present the case of a patient with palmoplantar pustulosis and osteoarticular pain in the left hemithorax and left sternocostal joint that raised the suspicion of SAPHO syndrome.

Case presentation

A 53-year-old female, an active smoker, came to our clinic for the onset of numerous small pustules on erythematous background on her palms and feet, associated with osteoarticular pain in the left hemithorax and left sternocostal joint.

The clinical examination at admission was within physiological limits. At the local cutaneous examination, bilaterally erythematous-squamous plaques, placards, papules, pustules, and painful erosions on the palms and plants detected (**Fig. 1 and 2**).

The association of palmoplantar pustulosis with osteoarticular pain in the left hemithorax and left sternocostal joint has raised suspicion of SAPHO syndrome. During the paraclinical investigations, a 4 mm punch biopsy was

* Department of Dermatology, „Elias“ University Emergency Hospital, Bucharest, Romania
** University of Medicine and Pharmacy „Carol Davila“, Bucharest, Romania
*** Department of Pathology, „Elias“ University Emergency Hospital, Bucharest
****sibel.ali@rez.umfcd.ro

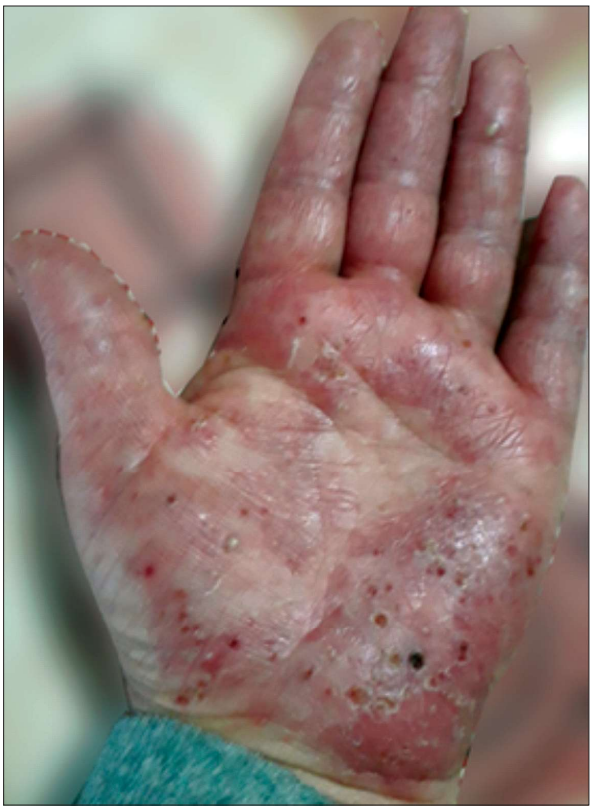

Figure 1. Pustules, papules, crusts, and scaling on an erythematous background, on the left palm.

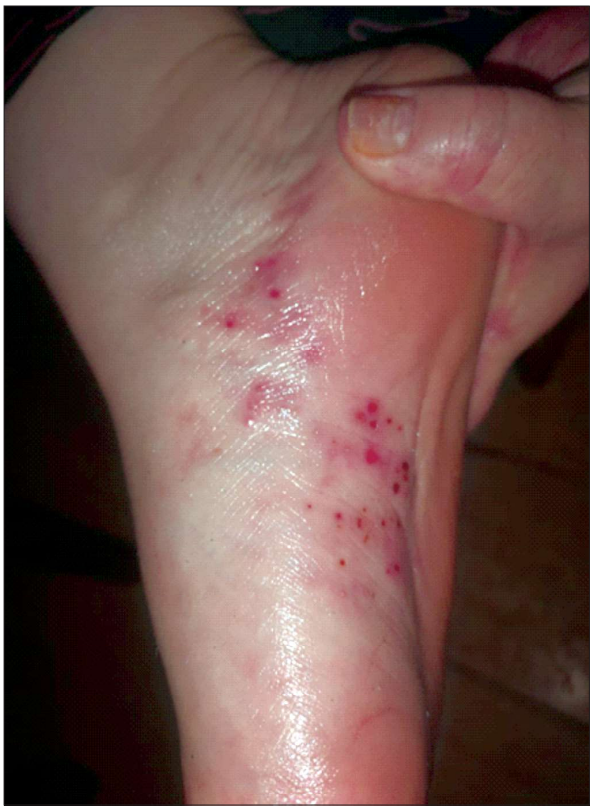

Figure 2. Painful pustules and erosions on erythematous background, on the right foot.

performed on the left palm, which confirmed the diagnosis of palmoplantar pustulosis (Fig. 4) . Ultra-sound of the left breast revealed a hypoechoic tumoral mass of 1/0.08 cm and vascularized axillary adenopathy. Soft tissue ultrasound and chest radiography showed no pathological changes. Tumor markers were within normal limits. Computed tomography revealed poorly demarcated increased density of the glandular tissue in the superior-external quadrant of the left breast. We recommended an oncological and surgical consultation, with further imaging investigations, which excluded the diagnosis of breast cancer. Imaging investigations did not reveal any changes suggestive of SAPHO syndrome, such as osteitis or hyperostosis in the anterior thorax.

Treatment

Until we received the result of the histo-pathological examination, we administered doxycycline 100 mg every 12 hours, topical treatment with dermatocorticosteroids, and 10 UVB - NB phototherapy sessions. The evolution of the skin lesions under treatment was favorable, with the persistence of osteoarticular pain. Upon discharge, we decided to start the treatment with Acitretin 20 mg/day and physical therapy for the osteoarticular pain. At the 6-month follow-up, the lesions improved, and we decided to continue treatment with Acitretin (**Figure 3**).

Discussions

The main discussion of this case is the association of the palmoplantar pustular eruption and osteoarticular symptoms that raised the suspicion of SAPHO syndrome. SAPHO syndrome describes a group of conditions with similar osteoarticular location (osteitis that mainly affects the anterior thoracic wall) associated with various dermatological manifestations [2]. The annual prevalence is estimated

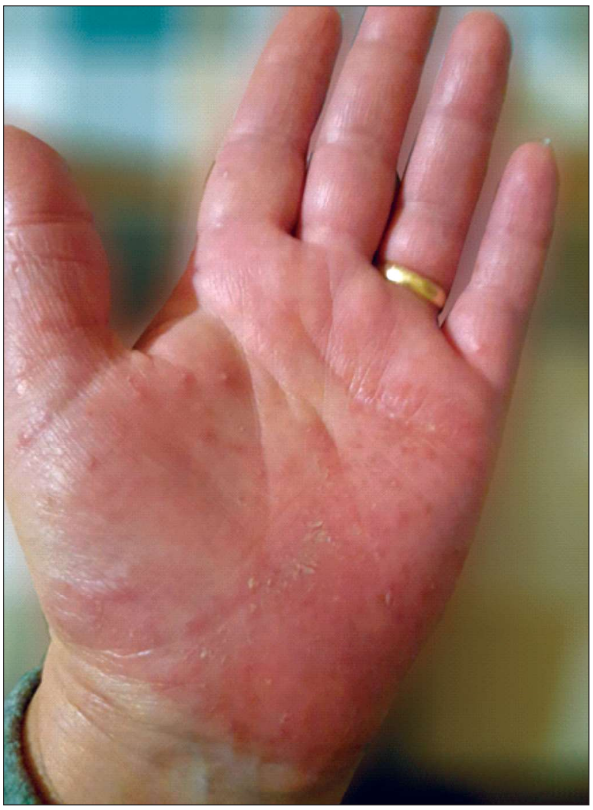

Figure 3. Lesion evolution at the 6-month follow-up, after treatment with Acitretin 20 mg/day.

at less than 1 in 10,000 in the Caucasian population and 0.00144 in 100,000 in the Japanese population [1].

The etiopathogenesis of SAPHO syndrome remains unclear, although several hypotheses have been proposed involving immunological, infectious, and genetic factors [1]. The auto-inflammatory pathogenesis of the disease was supported by the presence of elevated values of proinflammatory cytokines, such as tumor necrosis factor (TNF)-α and interleukins (IL)-1, IL-8, IL-17, and IL-18 [2]. The theory of the auto-inflammatory reaction is also supported by the response observed to the administration of biological therapies against anti-TNF-α, IL-1, and the IL-17 – IL-23 axis [2,7]. The theory of infectious etiopathogenesis is based on the identification of Cutibacterium acnes, Staphylococcus aureus, Haemophilus parainfluenzae, and actinomycetes in SAPHO osteitis lesions, C. acnes being the most frequently reported microorganism [2,8]. Although family aggregation of

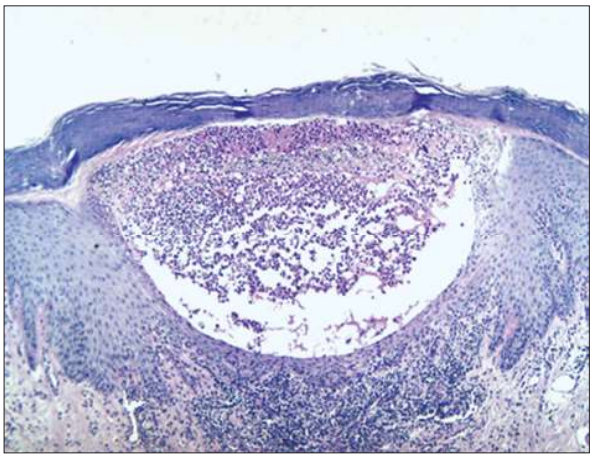

Figure 4. The histopathologic examination shows an epidermis with relatively uniform acanthosis; a subcorneal vesicle with monocytes and neutrophils. Minimal and moderate focal perivascular lymphocytic inflammatory infiltration in the superficial dermis with rare neutrophils in the perilesional dermis. This aspect is compatible with the diagnosis of palmoplantar pustulosis.

SAPHO syndrome has been reported, the genetic basis of SAPHO syndrome remains largely unknown [2].

Osteoarticular manifestations characteristic of this syndrome are synovitis, osteitis, hyperostosis usually evident later in the course of the disease, and axial spondyloarthritis-like [3]. The most common location is the anterior thoracic wall, especially the sternocostal and sternoclavicular joints as well as the costo-clavicular ligament [6]. Other locations include the mandible, the sacroiliac joint, and the spine. Peripheral joints are affected in 30% of cases [6].

Cutaneous manifestations in SAPHO syndrome include acneiform and neutrophilic dermatoses such as nodulocystic acne, hidradenitis suppurativa, pustular subcorneal dermatosis, and palmoplantar pustulosis [9].

The diagnosis of SAPHO syndrome is based on history and imaging investigations such as radiography, scintigraphy, computed tomography, magnetic resonance imaging, PET-CT, and ultrasound.

The diagnosis of SAPHO is considered in patients with:

a) Osteoarticular disease associated with palmoplantar pustulosis - inflammatory synovitis or pseudoseptic arthritis; hyperostosis/ osteitis

involving the anterior thoracic wall or another site; or sacroiliitis, spondylitis, and/or spondylodiscitis [2].

b) Osteoarticular disease associated with severe acne or suppurative hidradenitis - inflammatory synovitis, or hyperostosis/osteitis of the anterior thoracic wall; hyperostosis and osteitis; or sacroiliitis, spondylitis, and/or spondylodiscitis [2].

c) Isolated sterile hyperostosis/osteitis - sternocostoclavicular hyperostosis or other hyperostosis of the anterior thoracic wall or sterile osteitis. Skin lesions may or may not be present [2].

The therapeutic arsenal in SAPHO syndrome includes non-steroidal anti-inflammatory drugs, methotrexate, tetracyclines, anti-TNF inhibitors, oral retinoids, and other less commonly used therapies (bisphosphonates, dapsone, colchicine, sulfasalazine, anti-IL-12/23 or anti-inflammatory therapies). IL-17, systemic corticosteroid therapy, tofacitinib, etc.) [11].

The evolution of SAPHO syndrome is variable, with periods of remission and recurrence; disabling complications are rare, although peripheral arthritis can be erosive. Female gender, anterior thoracic wall involvement, peripheral arthritis, skin lesions, and high levels of inflammatory markers at baseline were associated with chronic disease progression [10].

Conclusion

The association of palmoplantar pustulosis and osteoarticular symptoms raised the suspicion of SAPHO syndrome. The result of the histopathological examination is compatible, with the diagnosis of palmo-plantar pustulosis. Imaging investigations ruled out the diagnosis of SAPHO syndrome, but the patient remains under observation for skin and joint symptoms, with regular evaluation. This case brings to our attention the possibility of neutrophilic diseases linkage with various forms of arthropathy, under the umbrella of SAPHO syndrome.

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Conflict of interest
NONE DECLARED

Correspondance address: Sibel Ali
Department of Dermatology, „Elias“ University Emergency Hospital, Bucharest, Romania
E-mail: sibel.ali@rez.umfcd.ro