SAPHO syndrome: case report

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Post Dermatol Alergol 2012; XXIX, 2: 136–138

Abstract

SAPHO syndrome is characterized by synovitis, acne, pustulosis, hyperostosis and osteitis. Dermatological manifestations include *palmoplantar pustulosis*, pustular psoriasis in other sites of the body, psoriasis vulgaris, acne (often severe, conglobata or fulminans), *hidradenitis suppurativa*. The vertebral arthritis associated with the disease belongs to a group of seronegative spondyloarthropathies. The aim was presenting a case of SAPHO syndrome which developed in a patient suffering from *palmoplantar pustulosis*. We present a patient who developed *pustulosis palmoplantaris* and low back pain at the same time. The diagnosis of SAPHO syndrome was established on clinical appearance and results of additional tests. The patient was treated with methotrexate in dose initially 15 mg, then 25 mg once a week, non-steroidal anti-inflammatory drugs and topical treatment with improvement. The pain of the chest wall or multifocal arthritis affecting the patient with *palmoplantar pustulosis* or severe acne should arouse suspicions of SAPHO syndrome.

Key words: SAPHO, palmoplantar pustulosis, pustular psoriasis, spondyloarthropathies.

Introduction

The SAPHO syndrome was first described in 1987 by Chamot *et al*. [1]. It is characterized by synovitis, acne, pustulosis, hyperostosis and osteitis, has a mild to severe course, with remissions and exacerbations. Most common dermatological manifestations include palmoplantar pustulosis, acne conglobata and *hidradenitis suppurativa*. Etiology and pathophysiology of the disease are not fully understood, but genetic, immunological and infectious factors are underlined [2, 3].

The aim of study was presenting a case of SAPHO syndrome which developed in a patient suffering from *palmoplantar pustulosis*.

Case report

A 55-year-old female patient suffering from hyperprolactinemia, glaucoma, paracetamol intolerance, thrombocytopenia (in anamnesis), is under medical care of the Psoriatic Outpatient Clinic of the Dermatological Department, Warsaw Medical University. Skin lesions on the soles in the form of pustules on erythematous and scaly background have been observed since April 2010. On dermatological examination she presented with skin lesions affecting almost 40% of the soles. There were creamy-yellow pustules occurring on inflamed, erythematous skin (Figure 1). Besides, occasional skin lesions appeared on palms, on the left knee few psoriatic papules were present. The patient's mother suffers from psoriasis vulgaris. The patient has been smoking cigarettes for 40 years. At the same time when skin lesions occurred, the lumbar spine pain appeared. Positron emission tomography (PET) revealed increased marker's cumulation in sternoclavicular joints. The patient was referred to the Institute of Rheumatology for a diagnostic procedure of arthritis and osteitis. On admission she claimed to have pains and edemas of sternoclavicular, sternocostal and left knee joints as well as diminished motility within the lumbar spine. Magnetic resonance revealed signs of inflammation of numerous bones and joints (sternoclavicular joints, patella, hip and tibial bones and within vertebral column [Th9, L2, L3, L5, S1]). Chronic inflammation of the left knee joint was shown by ultrasonography. In additional tests antigen HLA B27 was positive, erythrocyte sendimentation rate was 50 (N to 12). Antinuclear antibodies and rheumatic factor were negative. The diagnosis of SAPHO syndrome was established on clinical manifestation and additional tests. Methotrex-

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ate (15 mg per week), folic acid, tramadol, omeprazole and diclofenac were administered. Topically calcipotriolum cream and benzoyl peroxide gel were administered. Because of only partial improvement, the dose of methotrexate was increased to 25 mg/week with good response to the treatment (decreased bone and joint pains as well as clearing of skin lesions).

Discussion

The SAPHO syndrome is characterized by coexistence of osteoarticular manifestations and skin lesions. The diagnosis is made on clinical presentation, however the role of imaging studies is invaluable [4]. Diagnostic criteria of SAPHO syndrome are (according to Kahn *et al*. [5]): 1) multifocal osteitis with or without skin lesions; 2) aseptic acute or chronic joint inflammation associated with palmoplantar pustulosis, psoriasis vulgaris, acne or hidradenitis *suppurativa*; 3) aseptic osteitis associated with skin lesions as those mentioned above. One of criteria is sufficient to establish the diagnosis of SAPHO. The characteristic features of the syndrome are hyperostosis and osteitis in the upper thoracic wall. Arthritis and osteitis associated with the disease belong to a group of seronegative spondyloarthropathies. Rheumatic factor is negative, however antigen HLA B27 is often present [6]. Skin manifestations include palmoplantar pustulosis, pustular psoriasis, psoriasis vulgaris, acne (often severe, conglobata or fulminans) and *hidradenitis suppurativa* [4].

In one study, the mean age of patients was 37 ±13 years (10-71 years), without gender prevalence. The mean diagnostic delay was 4.5 ±5.6 years (0-26 years). The most common osteoarticular manifestations included anterior chest pain (73%), peripheral arthritis (32.7%) and among skin lesions (overall in 63.5%) palmoplantar pustulosis (51.5%) and acne (39.4%) predominated [7].

Current therapies for SAPHO syndrome are mainly symptomatic and comprise the use of non-steroidal anti-inflammatory drugs. Other drugs used in the treatment with different effectiveness are methotrexate, hydrochloroquine, sulfasalazine, d-penicillamine, azathioprine, and leflunomide. Moreover, glucocorticoids, acitretin, bisphosphonates and biologics are used [4]. In some studies, tumour necrosis factor α (TNF-α) antagonists (infliximab, etanercept, adalimumab) showed good efficacy [8-11]. It has to be underlined that TNF alpha antagonists themselves can exacerbate skin lesions, despite good response of the osteoarticular system. The mechanism of this phenomenon remains elusive [12]. The course of the disease varies, it is often chronic with remissions and exacerbations, with final improvement [6].

In our case, the SAPHO syndrome diagnosis was established on the presence of arthritis and osteitis, especially within anterior chest, synovitis of the knee joint and palmoplantar pustulosis. The treatment with methotrexate (dose 25 mg/week) and non-steroidal anti-inflammatory drugs was effective.

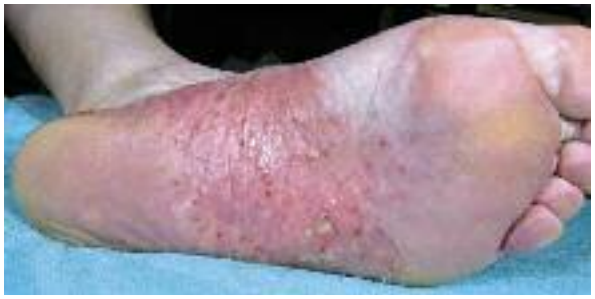

Fig. 1. Palmoplantar pustulosis skin lesions on the soles of the left foot

Because of common occurrence of skin lesions and long diagnostic delay, it is crucial to educate dermatologists in diagnosis and treatment of SAPHO syndrome [7]. Anterior chest pain or multifocal arthritis affecting the patient with palmoplantar pustulosis or severe acne should arouse suspicions of SAPHO syndrome.

Acknowledgments

The paper was awarded in the PTD competition, educational grant sponsored by Stiefel, GSK company.

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