

Pustulotic Arthro-Osteitis (Sonozaki Syndrome): A Case Report and Review of Literature

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ABSTRACT

Pustulotic arthro-osteitis (PAO) is a rare chronic inflammatory disease, which has now been classified as a seronegative spondyloarthritis. The sternoclavicular and sternocostal joints, pelvis, vertebra, hip, and long bones are affected. Skin findings of the disease are accepted as a variant of pustular psoriasis, but some authors have suggested that palmoplantar pustulosis (PPP) is a different entity. The synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome should be considered in the differential diagnosis. PAO differs from SAPHO by the absence of hyperostosis and the difference in skin manifestations. Here, we aimed to present a 34-year-old female patient with a diagnosis of PAO with typical skin findings and joint involvement.

Keywords: Sonozaki syndrome, pustular, osteitis, arthritis

Introduction

Sonozaki syndrome or pustulotic arthro-osteitis (PAO) is a rare chronic inflammatory disease characterized by sterile palmoplantar pustular lesions and anterior thoracic joint involvement [1]. The sternoclavicular and sternocostal joints, pelvis, vertebra, hip, and long bones are affected [1-3]. Palmoplantar pustulosis (PPP) is a chronic inflammatory disorder with typical pustular skin lesions symmetrically on the palmar and plantar faces of the extremities. In majority of patients, skin lesions occur concurrently with joint involvement, but the intervals may also vary between the onset of the symptoms [1-4]. Skin findings of the disease are accepted as a variant of pustular psoriasis, but some authors have suggested that PPP is a different entity [5, 6]. Although the PAO has earlier been included in psoriatic arthropathies (PsA), it also differs from PsA in the joint involvement pattern. Another disease for which differential diagnosis should be considered is the synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome. PAO is distinguished from SAPHO syndrome by the absence of hyperostosis and the difference in skin manifestations [7, 8].

Here, we aimed to present a 34-year-old female with typical clinical features of PAO, because of the rarity of the disease.

Case Report

A 34-year-old female patient was admitted to a dermatology clinic with pustular rash on both palms and soles for 4 months. The patient received treatment consisting of topical and oral steroid (0.5 mg/kg/day). On the tenth day of therapy, her lesions were resolved. However, 1 month after discontinuing the therapy, the lesions reappeared on both palms and soles. Also, 2.5 months later, she had pain in the left shoulder and pain and swelling in the left sternoclavicular joint. The patient was referred to our clinic with swelling and tenderness on the both sternoclavicular joints and multiple pustular lesions on bilateral palmoplantar regions (Figure 1, 2). The erythrocyte sedimentation rate (ESR) was 64 mm/hour, C-reactive protein (CRP) was 9.34 mg/L (0-5 mg/L), and white blood cell was 10.730. Renal-liver function tests, rheumatoid factor, and anticyclic citrulline peptide were within normal ranges. Hepatitis A, B, and C serologies; anti-nuclear antibody; and HLA-B27 were also negative. The skin punch biopsy was performed from pustular lesions, which revealed compact orthokeratosis on the surface, pustule formation in the subcorneal area, acanthosis in the epidermis, spon-



Figure 1. Multiple bilateral pustular lesions on the palmar regions



Figure 2. Multiple pustular lesions on the plantar region

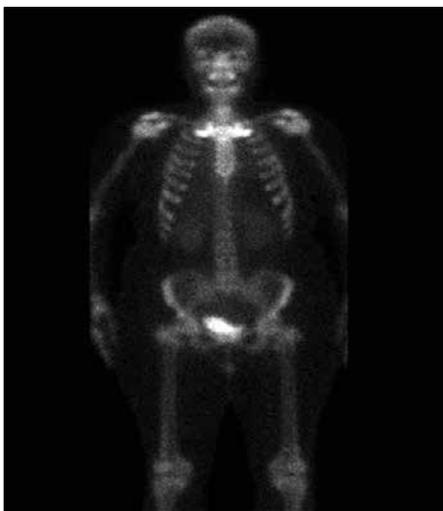


Figure 3. Bone scintigraphy (MDP Tc-99m) showed increased accumulation in both sternoclavicular joints symmetrically

giosis in the granular layer, and perivascular mononuclear cell infiltration in the superficial dermis. No pathological change was observed in the X-ray of the sternoclavicular joints. Effusion and increased Doppler activity were detected with musculoskeletal ultrasonography. Bone scintigraphy (MDP Tc-99m) showed increased accumulation in both sternoclavicular joints symmetrically (Figure 3).

Methylprednisolone (40 mg/day, per-oral [p.o]) and methotrexate (15 mg/a week, p.o) were initiated. The pustular lesions improved on the third day of treatment and were resolved almost completely after 1 week. Joint pain and swelling were completely resolved in the second week and laboratory values improved in accordance with the clinical course of the patient (ESR: 5 mm/hour and CRP: 0.76 mg/L). Written informed consent was obtained from the patient.

Discussion

The etiology is still unknown. Nonspecific chronic inflammation and neutrophil infiltration have been detected in the bone and soft tissue [1]. Sternoclavicular joint is the most commonly affected joint in Sonozaki syndrome with 77% of axial, 32% of peripheral joints, and 13% of sacroiliitis, respectively [1]. Erosion, syndesmophyte in the costochondral joints, spondylitis, suppurative spondylodiscitis in the vertebral involvement, ankylosing spondylitis-like erosions, and sclerosis in the sacroiliac joints are observed. However, the peripheral involvement of the disease is nonerosive and deformity is not expected [1]. PPP, the typical skin lesion of the disease, usually occurs concurrently with joint manifestations, but may develop before or after [1-4]. In our case, sternoclavicular involvement was present and arthro-osteitis occurred 4 months after pustular lesions.

Pustulotic arthro-osteitis is included in the group of spondyloarthropathies; however, there is no association with HLA-B27. The sternoclavicular joint involvement is uncommon in other spondyloarthropathies; thus, PAO differs from this group [9-11]. In our case, HLA-B27 was negative, sternoclavicular joint involvement was present, and there was no peripheral/sacroiliac joint involvement.

Peripheral joint involvement is a common feature of PsA. It is often distinguished from PAO with an erosive course and polyarticular joint involvement, including proximal and distal interphalangeal, metacarpophalangeal, metatarsophalangeal, ankle, and wrist joints. In addition, PPP, which is a skin lesion of PAO, has been reported to be a variation of pustular psoriasis and reported conversely that these two diseases are different clinical entities [5, 6]. Mejjad et al. [3] reported a prospective study with 23 PsA and 23 PAO patients. They found clinical and radiological differences between both groups, except for sacroiliac and spine involvement. Hence, authors have suggested that psoriasis and PPP are different dermatologic diseases [3].

Propionibacterium acnes has been suggested to play a role in the pathogenesis of SAPHO syndrome. It has been isolated from both the acne lesions and several synovial tissue biopsy materials of patients with SAPHO [10, 12]. PPP is a sterile lesion without any association with *Propionibacterium acnes*. The characteristic skeletal features are hyperostosis at the anterior thoracic joints and less often at the pelvic bones, spine, and peripheral joints [7, 13]. The characteristic “bull’s head” sign of SAPHO syndrome by bone scintigraphy (MDP Tc-99m) can also be seen in PAO. Many clinical features of SAPHO syndrome are similar to PAO. Differential diagnosis is difficult, but PAO is distinguished from SAPHO syndrome by the absence of acne and hyperostosis [7, 8]. In our patient, there was no anterior thoracic joint hyperostosis or no acne lesions characteristic of SAPHO syndrome.

Nonsteroidal anti-inflammatory drugs, corticosteroids, colchicine, methotrexate, sulphasalazine, and cyclosporine are used as therapeutic agents [14]. Infliximab and adalimumab have been reported as therapeutic agents in patients with resistant PAO [15].

The variable periods between the onset of symptoms and because it can be confused with similar joint and skin involvement diseases, lead to diagnostic and treatment difficulties. It may be difficult to diagnose Sonozaki syndrome if it is not evaluated among the differential diagnosis. Our aim was to emphasize that rheumatologists should consider PAO as a rare entity for the differential diagnosis of spondyloarthropathy, SAPHO, and PsA.

Informed Consent: Written informed consent was obtained from patients patient who participated in this study.

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