Do you know this syndrome? *

Você conhece esta síndrome?

Gerson Dellatorre[1] Caio César Silva de Castro[2]

CASE REPORT

A 47 year-old white female reported the appearance of palmoplantar pustules that progressed to scaling for 9 years. Four years ago, she started with episodes of continuous pain in the anterior chest wall, aggravated by inspiration, as well as erythema and a mass in pre-sternal area.

She had been a smoker for 32 years. Physical examination revealed palmoplantar pustules and scaling (Figures 1, 2), open comedones besides erythema, mass and pain on sternal joints. A complete blood count showed mild leukocytosis (12.380 leukocytes/μL). A chest tomography was obtained (Figure 3). The bacterial and fungal cultures from the skin were negative, and histopathologic examination showed subcorneal pustular dermatitis.

The patient was initially treated with methotrexate, with improvement of skins lesions alone. Currently, she has been using alendronate sodium PO (70mg weekly), and topical calcipotriol plus betamethasone dipropionate association, with improvement of skin lesions and reduction in frequency of bone pain.

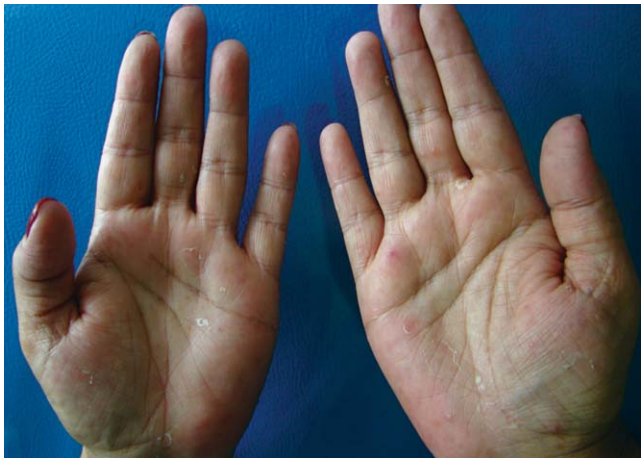

FIGURE 1: Palmar pustules and scaling of both hands

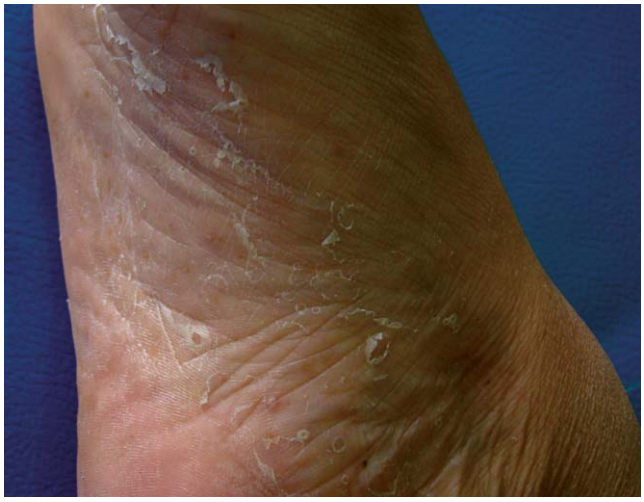

FIGURE 2: Detail of plantar pustules and scaling

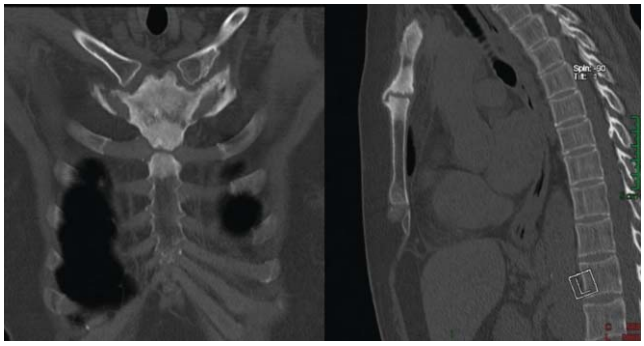

FIGURE 3: Computed tomography of the anterior chest wall (coronal and sagittal reconstructions): hypertrophy and bone sclerosis of the manubrium, associated with irregularities on borders of manubriosternal, sternoclavicular and sternocostal joints, bilaterally. Can also be observed areas of sclerosis in the upper parts of the sternal body, proximal portions of the first ribs and clavicles

Received on 31.05.2011.
Approved by the Advisory Board and accepted for publication on 20.08.2011.
* Study performed in a private clinic - Curitiba (PR) – Brazil.
Conflict of interest: None / *Conflito de interesse: Nenhum*
Financial funding: None / *Suporte financeiro: Nenhum*

[1] Medical Doctor - Medical Resident of Dermatology- Santa Casa de Misericórdia de Curitiba - Curitiba (PR), Brazil.
[2] PhD in Health Sciences - Pontifícia Universidade Católica do Paraná, Brazil. - Dermatologist and Staff Member of Dermatology Residency Program - Santa Casa de Misericórdia de Curitiba – Curitiba (PR), Brazil.

DISCUSSION

The SAPHO syndrome was first described in 1987 by Chamot et al.[1], wich have introduced this acronym to describe a disease that had five osteoarticular and dermatological manifestations, often combined: synovitis, acne, pustulosis, hyperostosis and osteitis[2]. There is predilection for involvement of anterior chest wall (presenting as pain and swelling)[3,4,5]. The etiology and its real prevalence are unknown[2,4].

The diagnosis of SAPHO syndrome is based on the exclusion of infectious arthritis or osteomyelitis and the presence of at least one of four diagnostic criteria proposed by Benhamou et al.6: 1) osteoarticular manifestations in severe acne; 2) osteoarticular manifestations in palmoplantar pustulosis; 3) hyperostosis with or without dermatosis and 4) recurrent multifocal chronic osteomyelitis involving the axial or peripheral skeleton, with or without dermatosis.

The cutaneous findings include predominantly neutrophilic skin eruptions such as palmoplantar pustulosis, hidradenitis suppurativa, acne conglobata and acne fulminans[2].

Complementary studies with radiographs, bone scintigraphy and computed tomography are useful in diagnostic workup. Laboratory tests are nonspecific and may reveal normal or elevated levels of C-reactive protein, ESR and leucometry[3]. In our case, the rheumatoid factor was negative. In another study, the analysis of 71 serum samples from patients with SAPHO was negative for rheumatoid factor[5].

The initial therapy is based on non-steroidal anti-inflammatory agents. Other treatment options include anti-rheumatic drugs such as colchicine, biphosphonates and corticosteroids, disease-modifying antirheumatic drugs such as methotrexate and sulfasalazine[2,7], and also isotretioin[8]. It has been reported the use of TNF-alpha antagonists with good efficacy for the osteoarticular symptoms and variable response to cutaneous symptomatology[2,9].

The long-term prognosis is good. In a 10 year follow-up study of 71 patients, only two developed severe osteoarticular sequelae[5].

In present study, the patient was a smoker, which could lead to the differential diagnosis of palmoplantar pustular psoriasis. However, the patient had bone symptoms, complete blood count and radiographic findings that corroborated with the diagnosis of SAPHO syndrome.

For being an underdiagnosed disease, patients are often subject to prolonged antibiotic therapy and unnecessary surgical procedures8. The dermatologic findings are present in about 60% of cases[3,4], and may be minimal and even go unnoticed by non-dermatologists. ❑

Abstract: The SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis and osteitis) includes a group of findings characterized by bone lesions usually located on the anterior chest wall, often associated with skin lesions. We report the case of a 47 years old patient, with osteochondritis at costoesternal and manubrium-sternal joints, besides of palmar-plantar pustulosis. The diagnosis is predominantly clinical and there are several treatment options described in the literature.
Keywords : Acquired hyperostosis syndrome; Acne vulgaris; Psoriasis; Synovitis

Resumo: A síndrome SAPHO (sinovite, acne, pustulose, hiperostose e osteíte) inclui um grupo de achados caracterizado por lesões ósseas localizadas geralmente na parede torácica anterior, frequentemente associadas a lesões cutâneas. Relata-se o caso de uma paciente de 47 anos, com quadro clínico composto por osteocondrite de articulação costoesternal e manúbrio-esternal, além de pustulose palmo-plantar. O diagnóstico é predominantemente clínico e há diversas opções de tratamento descritas na literatura.
Palavras-chave: Acne vulgar; Psoríase; Síndrome de hiperostose adquirida; Sinovite

REFERENCES

1. Chamot AM, Benhamou CL, Kahn MF, Beraneck L, Kaplan G, Prost A. Acne-pustulosis-hyperostosis-osteitis syndrome. Results of a national survey. 85 cases. Rev Rhum Mal Osteoartic. 1987;54:187-196.
2. Zhao Z, Li Y, Zhao H, Li H. Synovitis, acne, pustulosis, hyperostosis and osteitis (SAPHO) syndrome with review of the relevant published work. J Dermatol. 2011;38:155-159.
3. Poindexter G, Martinez S, Roubey RAS, Goldsmith LA. Synovitis-acne-pustulosis-hyperostosis-osteitis syndrome: A dermatologist's diagnostic dilemma. J Am Acad Dermatol. 2008;59:S53-S54.
4. Sallés M, Olivé A, Perez-Andres R, Holgado S, Mateo L, Riera E, et al. The SAPHO syndrome: a clinical and imaging study. Clin Rheumatol. 2011;30:245-9.
5. Colina M, Govoni M, Orzincolo C, Trotta F. Clinical and radiologic evolution of synovitis, acne, pustulosis, hyperostosis and osteitis syndrome: a single center study of a cohort of 71 subjects. Arthritis Rheum. 2009;61:813-21.
6. Benhamou C, Chamot AM, Kahn MF. Synovitis-acne-pustulosis-hyperostosis-osteitis (SAPHO) syndrome. Ann Dermatol Venereol. 1988;115:613-8.
7. Matzaroglou Ch, Velissaris D, Karageorgos A, Marangos M, Panagiotopoulos E, Karanikolas M. SAPHO syndrome diagnosis and treatment: report of five cases and review of the literature. Open Orthop J. 2009;3:100-6.
8. Galadari H, Bishop AG, Venna SS, Sultan E, Do D, Zeltser R. Synovitis, acne, pustulosis, hyperostosis, and osteitis syndrome treated with a combination of isotretinoin and pamidronate. J Am Acad Dermatol. 2009;61:123-5.
9. Massara A, Cavazzini PL, Trotta F. In SAPHO syndrome anti-TNF-alpha therapy may induce persistent amelioration of osteoarticular complaints, but may exacerbate cutaneous manifestation. Rheumatology. 2006;45:730-3.

Mailing address / Endereço para correspondência :
Caio C. Silva de Castro
Padre Anchieta Street, 1846 - Conj. 1014 - Edifício Biocentro, Curitiba (PR), Brazil.

How to cite this article/*Como citar este artigo*: Dellatorre G, Silva de Castro CC. Do you know this syndrome? SAPHO syndrome. An Bras Dermatol. 2012;87(1):155-6.