SAPHO Syndrome: Case Presentation

To the Editor: SAPHO syndrome is a group of skin and skeletal manifestations that come together during the course of a patients' life.[1-3] The term is an acronym of the most common manifestations: synovitis, acne conglobata (can even be fulminans, supurative hydrosadenitis, or cellulitis dissecans),[1] palmoplantar pustulosis, hyperostosis (usually on the anterior thoracic wall), and osteitis (sacroiliac or of the coccyx). It has an unknown etiology and most authors encase it with sponsyloarthropathies.[2] Its prevalence has not been widely studied, affecting mainly children and young adults, being rare in older patients; with a similar gender distribution. It can receive other names such as acne-associated spondyloarthropathy, sternoclavicular hyperostosis, clavicular osteitis condensans, pustular arthrosteitis, or chronic recurring multifocal osteomyelitis (pediatric form).[2,3]

We present the clinical case of a 34-year-old woman without any history of interest, diagnosed in 1994 with SAPHO syndrome (HLA B27+, pustular palmoplantar psoriasis, acromioclavicular and sternoclavicular hyperostosis and biopsied osteitis, and polyarthritis). She was resistant to high doses of phenylbuthazone and intolerant to sulphasalazine and moderate doses of methotrexate (biochemical liver affection). She did not tolerate treatment with PUVA, and high-dose corticosteroid therapy only led to transitory improvement. In December 2001 she underwent treatment with infliximab 3 mg/kg iv, with a good initial response of the bone symptoms and the acute phase reactants, but required a progressive increase in her dosage: 14 infusions. Pustulosis as well as sternoclavicular and sacroillyac joint activity worsened (gammagraphic control), opting for a change in treatment.

In May 2003 she started treatment with etanercept (50 mg sc/week) with low-dose methotrexate therapy (5 mg/week), showing improvement of the pustule lesions and the inflammatory low-back pain, with a reduction in the CRP (0.6 mg/dL). There was also improvement of the BASDAI (15) and BASFI (35.7) scores.

In October 2003 she once again presented increasing and incapacitating pain in the sternal, right acromioclavicular, and right shoulder areas (acromioclavicular space stenosis). In a new gammagram (January 2004), a subjective increase in pathologic uptake of the sternum with regards to the previous (March 2003), and sternoclavicular activity was seen.

Due to the resistance of the pathologic process, in April 2004 treatment with zolendronic acid 4 mg iv (3 doses) was decided upon. It finished on June 22, 2004 with a clear improvement in symptoms, especially sternoclavicular pain. In October 2004 she suffered a relapse of the sternal pain with important limitation of the joint balance of the right shoulder (which required local anesthesia), and worsening of the pustulosis.

The last gammagraphic control (December 2005) showed more control of the osteoblastic activity and persistence of sternal uptake (without an objective increase of the lesion). Clinically, the patient was functionally incapable and had poor pain control. The INSS evaluation team determined she had permanent total incapacity in March 2006.

The diagnosis of this syndrome is fundamentally clinical (Table),[4] there are no specific laboratory tests (HLA B27+ with a low frequency, acute phase reactant elevation). The study is completed with imaging techniques: conventional radiology (spondylitis, hyperostosis, and inflammatory lesions), computer tomography/magnetic resonance (adjacent soft-tissue lesions), bone gammagraphy (asymptomatic lesions). Imaging tests also allow for the differential diagnosis[5] with other processes (osteomyelitis, Paget's disease, bone metastasis, other spondyloarthropathies, Tietze syndrome). In some cases a bone biopsy might be warranted.[5,6]

Clinically it presents relapses and remissions and there is no curative treatment. The long-term prognosis is good and is not usually associated to severe or invalidating complications.[7]

Most of the patients are treated[8,9] in a symptomatic manner (NSAIDs, steroids), but alternative treatments have increased in number (with different degrees of efficacy), according to the scientific evidence (methotrexate, sulphasalazine, leflunomide, infliximab, etanercept, and byphosphonates). In some cases, the bone lesions (hyperostosis, osteitis) have been treated surgically,[10] but this has not proven superior to treatment with drugs.

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Diagnostic Criteria[a]

1.	Chronic multifocal relapsing osteomyelitis Generally sterile With/without coccyx affection With/without skin affection
2.	Acute/subacute/chronic arthritis in addition to Palmoplantar pustulosis Pustular psoriasis Severe acne Sterile ostheitis in any localization in addition to
3.	Palmoplantar pustulosis Pustular psoriasis Vulgar psoriasis Severe acne

[a]One of the 3 presentations is enough for diagnosis.

References

1. Martín Sánchez MC, Ruiz Villaverde R, Fernández Ángel I, Blasco Melguizo J, Jiménez Burgos F, Salvatierra Ossorio J, et al. Síndrome SAPHO Revista Internacional de Dermatología y Dermocosmética Clínica. 2002;5:106-8.
2. Crespi Villarías N, Rodríguez de Frutos J. Enfermedades poco prevalentes que debemos conocer: Síndrome de Sapho. Revista de la Sociedad Madrileña de Medicina de Familia y Comunitaria. 2003;1:37-8.
3. Olivé A, Pérez Andrés R, Rivas A, Holgado S, Casado E, Gumá M, et al. Síndrome SAPHO: estudio de 16 casos. Med Clin (Barc). 1999;112: 61-3.
4. Kohlfuerst S, Igerc I, Lind P. FDG PET heplful for diagnosing SAPHO syndrome. Clinical Nuclear Medicine. 2003;28:838-9.
5. Valverde I García J, Ordóñez S, Poca V. Hombro doloroso. Jano. 2000;1368: 43-83.
6. Hayem G, Bouchaud-Chabot A, Benali K, et al. SAPHO syndrome: a long-term follow-up study of 120 cases. Seminars in Arthritis and Rheumatism. 2000;29:332-4.
7. Solau-Gervais E, Soubrier M, Gerot I, et al. The usefulness of bone remodelling markers in predicting the efficacy of pamidronate treatment in SAPHO syndrome. Rheumatology (Oxford). 2006;45:339-42.
8. Scarpato S, Tirri E. Successful treatment of SAPHO syndrome with leflunomide. Report of two cases. Clin Exp Rheumatol. 2005;23:731.
9. Kopterides P, Pikazis D, Koufos C. Successful treatment of SAPHO síndrome with zoledronic acid. Arthritis Rheum. 2004;50:2970-3.
10. Bianchi G, Marinelli A, Frizziero A, Mercuri M. Hyperostosis and osteítis in SAPHO syndrome: coservative o surgical treatment. La Chirurgia degli Organi di Movimento. 2004;89:45-9.

Erratum

In the Letter to the Editor "Precisions on the history of quinine," published in this journal (Reumatol Clin. 2007;3[4]:194-6) it has been detected that only 1 author appears when, in truth, the authors are:

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