Imaging for Synovitis, Acne, Pustulosis, Hyperostosis, and Osteitis (SAPHO) Syndrome

Steven Schaub, MD[a], Hartley M. Sirkis, MB BCh[b], Jonathan Kay, MD[a,*]

KEYWORDS

• SAPHO syndrome • CRMO • Acne • Palmoplantar pustulosis • 'Bull's head' sign • Vertebral corner lesion • Hyperostosis • Osteitis

KEY POINTS

- Synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome, is a spectrum of disease that includes chronic recurrent multifocal osteomyelitis and is characterized by inflammatory bone lesions.
- SAPHO syndrome resembles the spondyloarthropathies, predominantly involving the anterior chest wall, spine, sacroiliac joints, and peripheral joints with enthesitis.
- The pattern of increased technetium-99m uptake in the sternoclavicular region can resemble a bull's head, which is a finding considered to be specific for SAPHO syndrome.
- Spinal involvement on MRI may appear as T2-weighted signal hyperintensity of the anterior corners of vertebral bodies, deemed corner lesions.
- Early lesions of SAPHO syndrome tend to be osteodestructive with osteolysis, whereas later lesions are osteoproliferative with hyperostosis and sclerosis.

INTRODUCTION

The entity of chronic recurrent multifocal osteomyelitis (CRMO) was introduced in 1978, with the description of 9 patients between the ages of 4 and 26 who had inflammatory bone lesions predominantly in the clavicular region and in the metaphyses of tubular bones with associated pustulosis palmoplantaris.[1,2] Deemed a sterile or

Disclosure Statement: Dr S. Schaub, Dr H. M. Sirkis, and Dr J. Kay have no relevant disclosures.

[a] Division of Rheumatology, Department of Medicine, UMass Memorial Medical Center and University of Massachusetts Medical School, 119 Belmont Street, Worcester, MA 01605, USA; [b] Division of Musculoskeletal Imaging, Department of Radiology, UMass Memorial Medical Center and University of Massachusetts Medical School, 55 Lake Avenue North, Worcester, MA 01655, USA

* Corresponding author. Rheumatology Center, Memorial Campus, UMass Memorial Medical Center and University of Massachusetts Medical School, 119 Belmont Street, Worcester, MA 01605.

E-mail address: jonathan.kay@umassmemorial.org

Rheum Dis Clin N Am 42 (2016) 695–710
http://dx.doi.org/10.1016/j.rdc.2016.07.011
rheumatic.theclinics.com

aseptic autoinflammatory disorder, based on frequently negative cultures of involved sites, the disease is characterized by multiple foci of osteomyelitis with episodic bone pain. However, CRMO is now considered to be part of the spectrum of disease labeled as synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome.

The term "SAPHO syndrome" was coined in 1987 by Chamot and colleagues,[3] who collected 85 cases with dermatologic findings, including severe acne and palmoplantar pustulosis, coupled with radiographic features of hyperostosis and osteitis.[4] Initially named "syndrome acne-pustulosis-hyperostosis-osteitis," the "S" was later used to signify "synovitis" when the association with inflammatory arthopathy was identified, placing the syndrome within the spectrum of spondyloarthropathies.[5] However, this categorization remains controversial.

The same French group proposed inclusion and exclusion criteria for the SAPHO syndrome that remain widely used, but no diagnostic criteria are accepted universally (**Box 1**).[4,5] Thus, the SAPHO syndrome often is diagnosed by excluding other disease processes, such as infection and malignancy, which may delay the initiation of treatment.

Because the diagnostic label of CRMO focuses only on the osseous manifestations of this disorder, its use no longer is favored. Instead, the acronym SAPHO now is used to describe a broad variety of disorders, including CRMO, that have both osteoarticular and dermatologic manifestations. More than 50 terms have been used to describe these combinations of cutaneous and osseous findings, including sternocostoclavicular hyperostosis, acquired hyperostosis, pustulotic arthroosteitis, and acne-associated spondyloarthropathy.[6,7] More recently, some have suggested that CRMO and SAPHO syndrome should be classified within the spectrum of

Box 1
Diagnosis of SAPHO syndrome

SAPHO criteria

- Inclusion (presence of one criterion sufficient for inclusion as SAPHO syndrome)
  - Osteoarticular manifestations of acne congoblata, acne fulminans, or hidradenitis suppurativa
  - Osteoarticular manifestations of PPP
  - Hyperostosis (of the anterior chest wall, limbs, or spine) with or without dermatosis
  - CRMO involving the axial or peripheral skeleton with or without dermatosis
- Sometimes reported
  - Possible association with psoriasis vulgaris or inflammatory enterocolopathy
  - Features of ankylosing spondylitis
  - Presence of low-virulence bacterial infections
- Exclusion
  - Septic osteomyelitis
  - Infectious chest wall arthritis
  - Infectious PPP
  - Palmoplantar keratoderma
  - Diffuse idiopathic skeletal hyperostosis
  - Osteoarticular manifestations of retinoid therapy

Abbreviations: CRMO, chronic recurrent multifocal osteomyelitis; PPP, palmoplantar pustulosis; SAPHO, synovitis, acne, pustulosis, hyperostosis, and osteitis.

Data from Benhamou CL, Chamot AM, Kahn MF. Synovitis-acne-pustulosis-hyperostosis-osteomyelitis syndrome (SAPHO): a new syndrome among the spondyloarthropathies? Clin Exp Rheum 1988;6(2):109–12. *Adapted from* Carneiro S, Sampaio-Barros PD. SAPHO syndrome. Rheum Dis Clin North Am 2013;39(2):401–18.