

## Caso Clínico / Radiological Case Report

**SINAL DE BULLHEAD NA SÍNDROME SAPHO***BULLHEAD SIGN IN SAPHO SYNDROME*

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Recebido a 11/05/2016  
Aceite a 12/07/2016

**Resumo**

O síndrome SAPHO – sinovite, acne, pustulose, hiperostose e osteíte – é uma entidade rara caracterizada por alterações osteoarticulares, frequentemente na parede torácica anterior, usualmente associadas a lesões dermatológicas. A variabilidade das manifestações clínicas dificulta o diagnóstico precoce, que é essencial para evitar procedimentos diagnósticos invasivos e terapêuticas desnecessárias. Apresentamos o caso de uma mulher de 41 anos de idade com queixas de dores osteoarticulares generalizadas, sobretudo na parede torácica ântero-superior, e lesões dermatológicas intermitentes. Uma TC torácica revelou alterações esternoclaviculares sugestivas de sequelas de osteomielite. Clinicamente não havia história de infecção. A dor osteoarticular constante e exames não conclusivos desencadearam a requisição de cintigrafia óssea, que revelou o sinal de bullhead, compatível com osteíte e hiperostose nas regiões esternoclaviculares. O sinal de bullhead na cintigrafia óssea é altamente específico de síndrome de SAPHO e foi essencial para estabelecer o diagnóstico final neste caso.

**Palavras-chave**

Síndrome SAPHO; Sinal de Bullhead.

**Abstract**

SAPHO – synovitis, acne, pustulosis, hyperostosis and osteitis – syndrome is a rare entity characterized by osteoarticular alterations, frequently in the anterior thoracic wall, usually associated with skin lesions. The variability of clinical manifestations hinders early diagnosis, which is essential to avoid unnecessary invasive diagnostic procedures and therapeutics. We present the case of a 41-year-old woman with complaints of generalized osteoarticular pain mainly in the superior anterior chest, and intermittent skin lesions. A thoracic CT revealed sternoclavicular abnormalities suggestive of osteomyelitis sequelae. There were no clinical clues for past infection. The constant osteoarticular pain and non-conclusive exams triggered the request of bone scintigraphy which showed the bullhead sign, compatible with osteitis and hyperostosis in the sternoclavicular regions. Bone scintigraphy's bullhead sign is highly specific of SAPHO syndrome and was essential to establish the final diagnosis in this case.

**Key-words**

SAPHO Syndrome; Bullhead Sign.

SAPHO syndrome – synovitis, acne, pustulosis, hyperostosis and osteitis – is a rare entity of unknown etiology characterized by an association of osteoarticular alterations and skin lesions<sup>1,2</sup>. This condition affects mainly children and young adults, developing in outbreak episodes, chronic, non-malignant and non-infectious<sup>3,4</sup>. Diagnostic criteria of SAPHO syndrome include multifocal osteitis, sterile acute or chronic joint inflammation, and sterile osteitis, in association with skin lesions of psoriasis (pustular psoriasis and palmo-plantar pustulosis) or acne (conglobata, fulminans or follicular occlusion triad), but there are no specific diagnostic tests<sup>3,4</sup>. Anterior chest wall pain is the most typical osteoarticular symptom and palmo-plantar pustulosis the most frequent skin manifestation<sup>3,5</sup>. Despite this, the wide spectrum of clinical manifestations mimetizes several diseases, hindering early diagnosis, which is essential to avoid unnecessary invasive diagnostic procedures and therapeutics<sup>3,5</sup>. Bone scintigraphy is a Nuclear Medicine imaging exam with high sensitivity to detect pathophysiological changes in bone metabolism<sup>7,8</sup>. Diphosphonates labeled with technetium-99m are the most commonly used radiopharmaceuticals in this technique<sup>7,8</sup>. These agents concentrate predominantly in the mineral phase of the bone and their uptake reflects osteoblastic activity, as well as the bone's blood supply (an increased flow is

matched with an increased uptake)<sup>7,8</sup>. Technetium-99m is a low-energy gamma-ray emitter with a half-life of 6 hours, allowing good quality images exposing the patient to a relatively low radiation dose<sup>6</sup>. The administration of a single dose of 99m-Tc-diphosphonates allows the acquisition of two and three-dimensional whole-body images without exposing the patient to extra-radiation<sup>7,8</sup>. Several factors affect the 99m-Tc-diphosphonates' uptake, namely vitamin D, parathyroid hormone, the use of corticosteroids and acid-base balance, allowing the evaluation of trauma, tumours, infections, metabolic diseases and others<sup>7</sup>. We present the case of a 41-year-old woman with a 6-year-long complaint of generalized osteoarticular pain, mainly located in superior anterior chest, and intermittent pustulosis. Sequential laboratory findings revealed elevated Sedimentation Rate and C-Reactive Protein values. Thoracic CT revealed sternoclavicular abnormalities, with apparent osteoblastic/osteolytic lesions (Fig. 1) suggestive of osteomyelitis sequelae. The constant osteoarticular pain, which led to chronic use of non-steroidal anti-inflammatory drugs and the non-conclusive diagnostic tests triggered the request of bone scintigraphy, in order to evaluate possible osteoarticular inflammatory disease.



**Figure 1** - CT abnormalities interpreted as suggestive of osteomyelitis sequelae in the sternoclavicular regions.

Bone scintigraphy whole-body images revealed the “bullhead sign” (Fig. 2), considered pathognomonic of SAPHO’s syndrome. The “bullhead sign” is compatible with osteitis and hyperostosis in the sternoclavicular regions, which lead to a higher uptake of the radiopharmaceutical in this area<sup>4,6</sup>. The analysis of the complete exam revealed high uptake in the right femoral diaphysis, suggestive of hyperostosis, as well as in the shoulders and hips joints, suggestive of arthritis. In this case, the “bullhead sign” was essential to establish the final diagnosis. Due to hypersensitivity to salazopyrin, the patient received pamidronate therapy and remained asymptomatic for 2 years. After this period, she presented new pain symptoms and was sequentially submitted to desensitization and treatment with salazopyrin, having remained asymptomatic since then (25 months ago).

The osteoarticular alterations in SAPHO’s syndrome are not always seen or are non-specific in conventional radiological exams, namely thoracic CT and radiograph. With bone scintigraphy it is possible to visualize areas of unincreased uptake in affected bone and the possibility of acquiring whole-body images without exposing the patient to additional radiation is very helpful to detect multiple involved locations<sup>3,4</sup>. Its high sensitivity for detection of morphofunctional changes in bone metabolism when compared to other imaging techniques allows detection of bone alterations even during asymptomatic phases, being of high value in early diagnosis<sup>3,7,8</sup>.

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**Figure 2** – Bone scintigraphy’s bullhead sign in the sternoclavicular regions.

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