Case Report

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# A Bull's Head Sign in SAPHO

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## ABSTRACT

A syndrome of synovitis, acne, pustulosis, hyperostosis and osteitis (SAPHO) is a rare autoimmune disease. It occurs in genetically predisposed individuals by immune activation triggered by possible infectious agents. A 60-year-old man was presented with musculoskeletal and cutaneous manifestations. After exclusion of differential diagnoses, he was diagnosed with SAPHO and successfully treated with adalimumab. He had a typical image defined as a bull's head sign in bone scintigraphy with 99mTc-methylene diphosphonate. Positron emission tomography also revealed increased inflammatory activity in related anterior chest joints and soft tissue. We want to emphasize diagnostic radiological images in patients with SAPHO to increase the awareness of clinicians.

## Introduction

A syndrome of synovitis, acne, pustulosis, hyperostosis and osteitis (SAPHO) is a rare autoimmune disease characterized by articular and cutaneous presentations [1]. Although it is not clarified, immune activation by infectious triggers such as propionibacterium acnes is considered as one of the main pathogenetic mechanisms in genetically predisposed individuals [2]. Patients generally present with local anterior chest pain and/or inflammatory back pain and dermatological manifestations [3]. A detailed medical history and comprehensive physical examination are quite important in these patients. A vast majority of patients were suffering from psoriasis, palmoplantar pustulosis and acne in a Japanese cohort [4]. The authors define hyperostosis and osteitis as the characteristics of the SAPHO syndrome after exclusion of other possible diagnoses. Hyperostosis of the anterior chest wall can be confused with sclerotic bone metastases. Differential diagnosis of SAPHO includes malignant, infectious, and/or metabolic bone diseases such as bone metastases, tuberculosis, and Paget's disease [5].

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Different imaging modalities including X-ray, computed tomography (CT) scans, bone scintigraphy, magnetic resonance imaging, and F-18 fluorodeoxyglucose-positron emission tomography (PET) may be supportive for the diagnosis of the disease. Being familiar with each radiographic image may prevent unnecessary invasive procedures and facilitates the correct and early diagnosis [6].

## Case presentation

A 60-year-old male presented with a 3-year history of anterior chest pain, inflammatory back pain and palmoplantar pustulosis of hands. The sternoclavicular and manubriosternal joints were tender on physical examination. There were not any additional abnormal findings on systemic examination. A complete blood count was compatible with chronic disease anemia, comprehensive biochemical tests including alkaline phosphatase, plasma parathormone, and serum calcium levels were normal. The erythrocyte sedimentation rate and serum C-reactive protein were 128 mm/hour and 226 mg/L, respectively. Bone scintigraphy with 99mTc-methylene diphosphonate revealed increased radionuclide uptake in the bilateral sternocostoclavicular and manubriosternal joints (bull's head sign) (Fig. 1). Positron emission tomography (PET) revealed moderately increased F-18 fluorodeoxyglucose uptake in the manubrium sterni, 3rd and 4th ribs, and adjacent soft tissue components (Fig. 2, 3) in addition to a three-dimensional image (Fig. 4). After ruling out infectious diseases (tuberculosis, brucellosis) and malignancies, he was diagnosed with SAPHO. He was initially treated with csDMARDs, including methotrexate and sulfasalazine. Subsequently, adalimumab resulted in significant clinical improvements of arthritis and cutaneous symptoms.

## Discussion

Anterior chest area involves sternum, sternoclavicular, manubriosternal, costosternal and costochondral junctions. Any of them may be affected by the disease [7]. Fu Z et al. noted that the bull's head

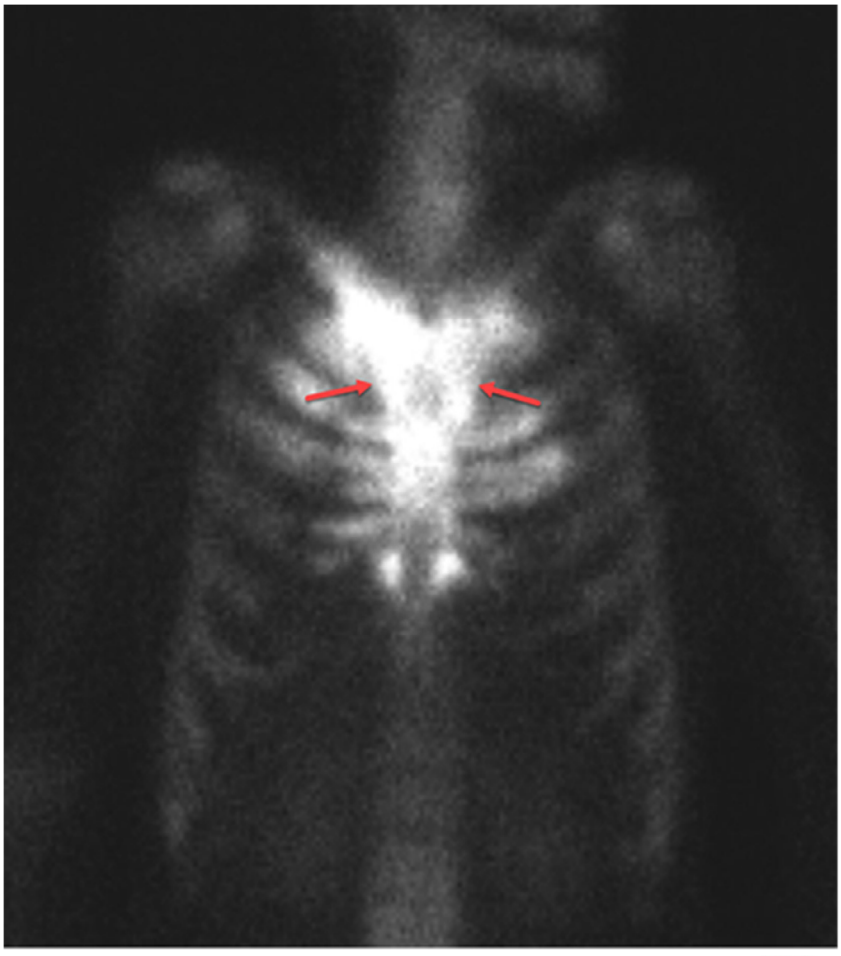

**Fig. 1.** Bone scintigraphy (99mTc-methylene diphosphonate); radionuclide uptake in the bilateral sternocostoclavicular and manubriosternal joints (bull's head sign).

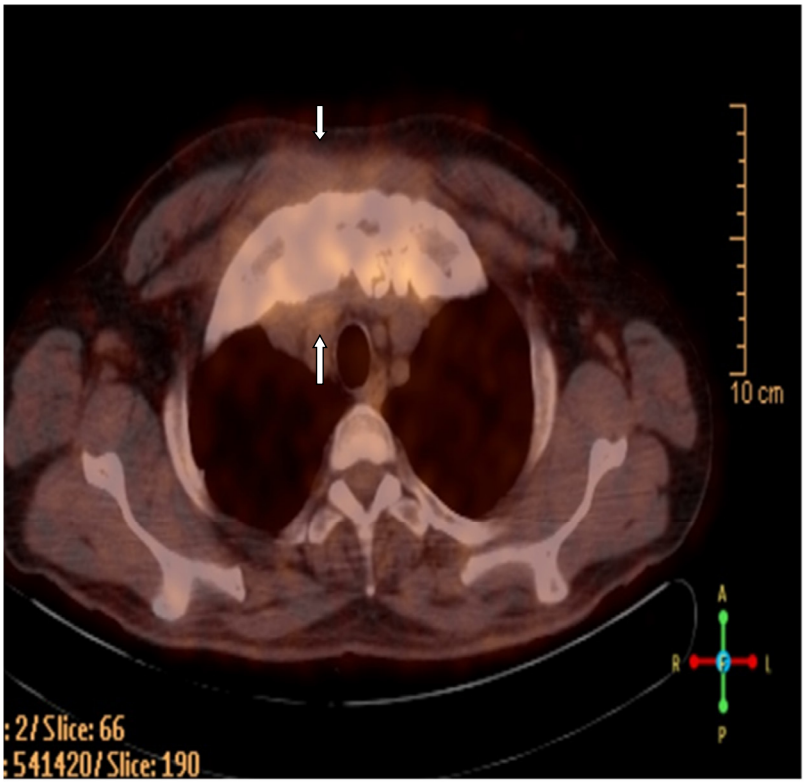


**Fig. 2.** Positron emission tomography (PET-CT); moderately increased F-18 fluorodeoxyglucose uptake in the manubrium sterni, 3rd and 4th ribs, and adjacent soft tissue components.

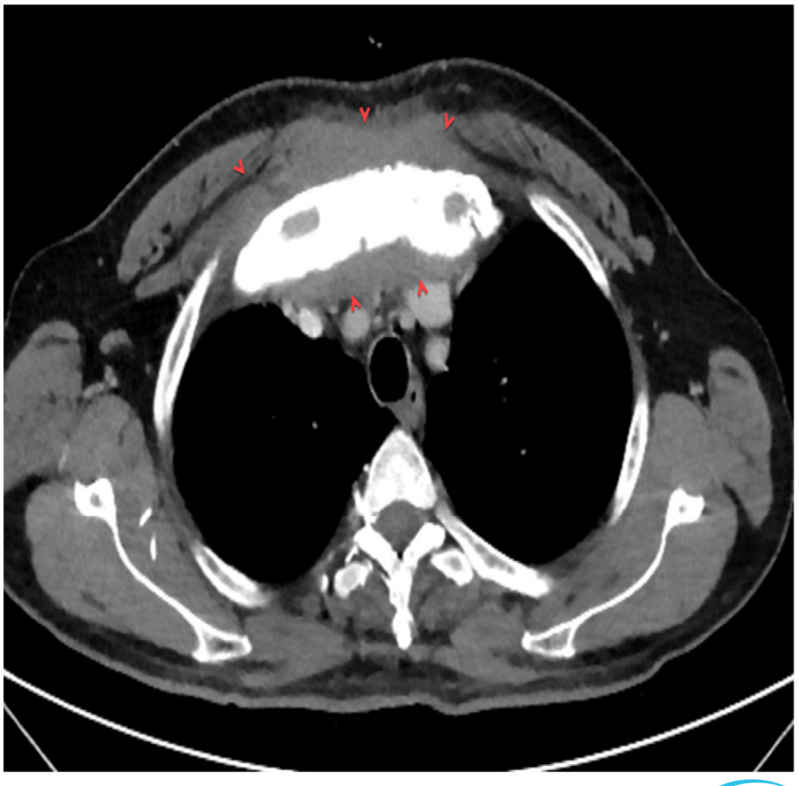

**Fig. 3.** Computed tomography; three-dimensional image of anterior chest wall and ribs.

sign is not completely but highly sensitive and quite characteristic for SAPHO, and this typical scintigraphy image can be in stable appearance even years later [8]. Moreover, early detection ability of bone scintigraphy was highlighted in eight patients [9]. More recent publications are related to PET. Multiple bone lesions and low to moderate FDG uptake are commonly seen in PET scans in patients with SAPHO. Recently, the coexistence of osteosclerosis and osteolysis is also emphasized as a typical feature. In the same study, PET/CT and bone scintigraphy had comparable effectiveness in revealing skeletal lesions [10].

Having a wide list of differential diagnoses including malignant, infectious, and/or metabolic bone diseases is quite challenging for patients with SAPHO. Besides, early diagnosis gives a chance to patients for early and appropriate treatment and prevents possible further musculoskeletal damage [11]. Although there is no standard treatment, there are many successfully treated case reports and case series with different treatment modalities. Current literature involves many conventional synthetic disease-modifying antirheumatic drugs (DMARDs) such as methotrexate and sulfasalasine, and biological DMARDs, including tumor necrosis factor-alpha inhibitors and anti-interleukin-1 (anti-IL-1) agents that are effectively used in the treatment of the disease [12]. Adalimumab was successfully used in our patient.

As a result, bone scintigraphy is an important and highly sensitive imaging method that prevents unnecessary invasive procedures and facilitates the correct diagnosis with a specific image such as a Bull's head sign.Furthermore, all physicians dealing with SAPHO think that it has underestimated prevalence due to lack of recognition. We believe that this paper will provide a great awareness.

## Ethical Considerations

### Consent

Written informed consent form was obtained from the patient for publication

### Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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### Conflict of Interests

All authors declare that they have no conflict of interest or financial support.

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