Case 17894

Eurorad

Anterior chest wall involvement in SAPHO syndrome

Published on 14.10.2022

DOI: 10.35100/eurorad/case.17894
ISSN: 1563-4086
Section: Musculoskeletal system
Area of Interest: Bones Musculoskeletal joint
Imaging Technique: Conventional radiography
Imaging Technique: CT
Imaging Technique: MR
Special Focus: Inflammation Case Type: Clinical Cases
Authors: Olivier Leroij1,2, Filip Vanhoenacker1-3
Patient: 55 years, female

Clinical History:

A 55-year-old woman presented with general arthralgia and axial pain since many years. Clinical inspection revealed palmoplantar pustulosis.

Imaging Findings:

Chest radiography showed sclerosis of the medial aspect of the left clavicle (Figure 2).

CT confirmed osteosclerosis and periosteal reaction at the left clavicle. Erosions were seen at the sternoclavicular and costosternal joints with ligamentous ossification of the left costoclavicular ligament (Figure 3a-b).

MRI of the sternoclavicular joints revealed a mixture of sclerosis and bone marrow oedema (Figure 4a-c). After administration of gadolinium contrast, there was also enhancement of the left clavicle (Figure 4d).

Discussion:

SAPHO is an acronym that stands for synovitis, acne, pustulosis, hyperostosis and osteitis.

The hallmark of the skin manifestations is palmoplantar pustulosis and acne [1].

SAPHO syndrome tends to involve the axial skeleton and the anterior chest wall, especially the sternoclavicular joint, with clavicular involvement typically starting at the medial aspect [2]. Spinal involvement is usually monovertebral, with the thoracic spine being most frequently affected [3]. Sacroiliitis is also sometimes seen. Peripheral joints in the form of a mono- or polyarthritis can also be affected, but to a lesser extent [4].

Its pathogenesis is still not completely understood. There is an association with HLA-B27 and different pathogens were isolated from different bone sites and pustules in the skin. The most probable hypothesis is that it is caused by autoimmune reactions in genetically predisposed individuals triggered by some infectious agent [5].

Patients present with pain, swelling, and limitation of movement, most often in the anterior chest wall. In many patients, there is a considerable delay in the correct diagnosis [1].

In children and adolescents, a similar condition called chronic recurrent multifocal osteomyelitis (CRMO), more commonly involves the long bone metaphyses and clavicles [1].

The imaging features are dominated by hyperostosis, osteitis and synovitis, arthritis, and chronic inflammatory bone disease. Osteitis consists of inflammation of the medullary bone, which manifests as osteosclerosis on conventional radiography. The term hyperostosis is used to designate chronic periosteal reaction and cortical thickening leading to bone hypertrophy. Radiography is usually the initial modality and can reveal sclerosis and periosteal reaction in advanced disease, but it is less sensitive for early detection. CT scan is the preferred modality to demonstrate osteosclerosis and/or hyperostosis, erosions, and ligamentous ossifications. MR imaging on the other hand, is best suited for demonstration of inflammatory changes and bone marrow oedema using fluid-sensitive sequences. Whole-body MRI is recommended for the evaluation of multifocality and asymptomatic lesions [6, 7].

The treatment remains a challenge. It includes administration of nonsteroidal anti-inflammatory drugs, corticosteroids, methotrexate, and tumour necrosis factor alfa inhibitors. Except for a minority of patients who have a self-limited course, most of them have either a relapsing-remitting course or a chronic indolent disease pattern [5].

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: SAPHO syndrome with anterior chest wall involvement, Septic arthritis, Spondylarthritis, Sternoclavicular osteoarthritis, Friedrich's disease (avascular necrosis of the medial clavicle), Osteitis condensans of the clavicle

Final Diagnosis: SAPHO syndrome with anterior chest wall involvement

References:

Lew PP, Ngai SS, Hamidi R, Cho JK, Birnbaum RA, Peng DH, and Varma RK (2014) Imaging of Disorders Affecting the Bone and Skin. RadioGraphics 34:1, 197-216 (PMID: 24428291)
Nguyen MT, Borchers A, Selmi C, Naguwa SM, Cheema G, Gershwin ME (2012) The SAPHO syndrome. Semin Arthritis Rheum 42(3):254-65 (PMID: 23153960)
Sweeney SA, Kumar VA, Tayar J, Weber DM, Safdar A, Alonso C, and Hymes S (2012) Case 181: Synovitis Acne Pustulosis Hyperostosis Osteitis (SAPHO) Syndrome. Radiology 263:2, 613-617 (PMID: 22517965)
Cotten A, Flipo RM, Mentre A, Delaporte E, Duquesnoy B, and Chastanet P (1995) SAPHO syndrome. RadioGraphics 15:5, 1147-1154 (PMID: 7501856)
Rukavina I (2015) SAPHO syndrome: a review. J Child Orthop. 9(1):19-27 (PMID: 25585872)
Earwaker, JWS, Cotten, A (2003) SAPHO: syndrome or concept? Imaging findings. Skeletal Radiol 32, 311–327 (PMID: 12719925).
Himuro, H., Kurata, S., Nagata, S. et al. (2020) Imaging features in patients with SAPHO/CRMO: a pictorial review. Jpn J Radiol 38, 622–629 (PMID: 32356235).

Figure 1

a

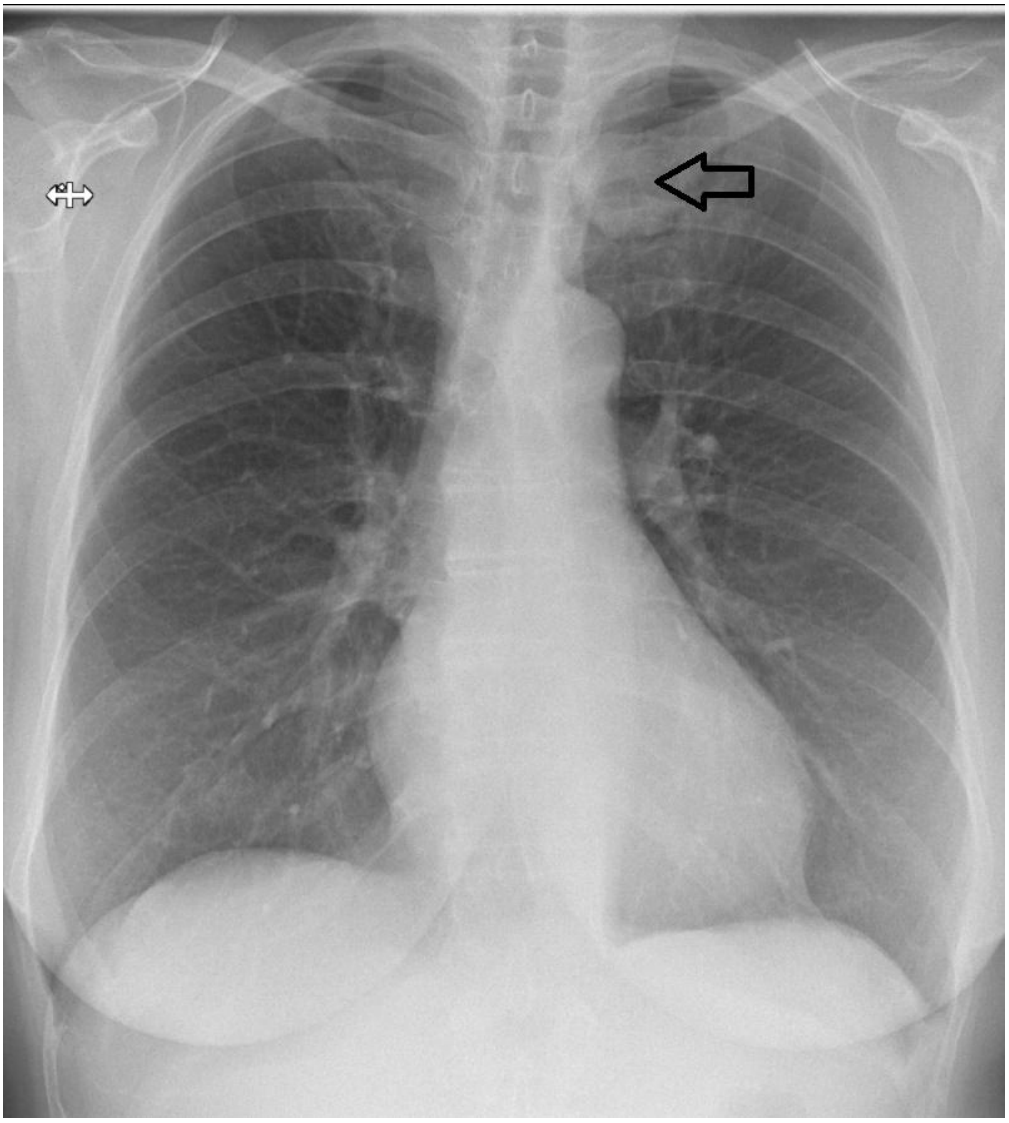
Description: Palmoplantar pustulosis. **Origin:** © Filip Vanhoenacker, General Hospital Sint-Maarten, Mechelen, Belgium 2022.

b

Description: Palmoplantar pustulosis. **Origin:** © Filip Vanhoenacker, General Hospital Sint-Maarten, Mechelen, Belgium 2022.

Figure 2

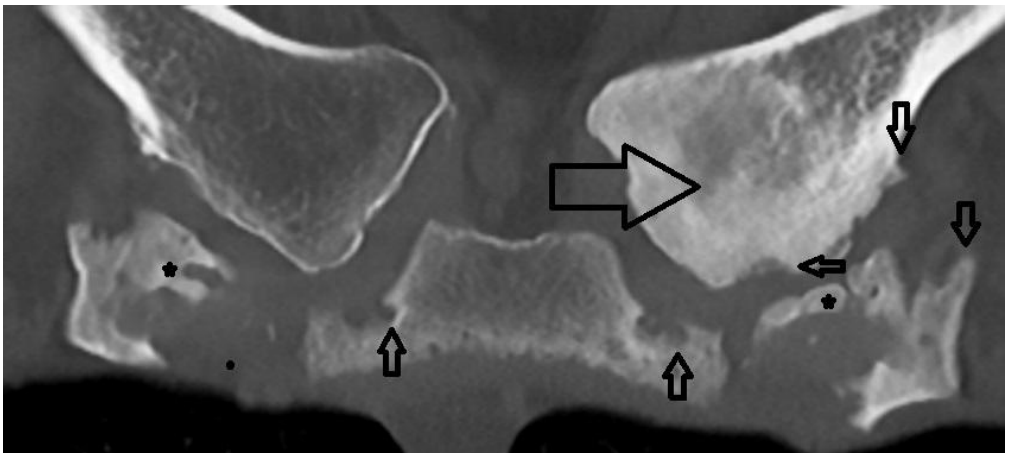
a



Description: Conventional radiograph of the Chest. Sclerosis at the medial end of the left clavicle (open arrow). **Origin:** © Filip Vanhoenacker, General Hospital Sint-Maarten, Mechelen, Belgium 2022.

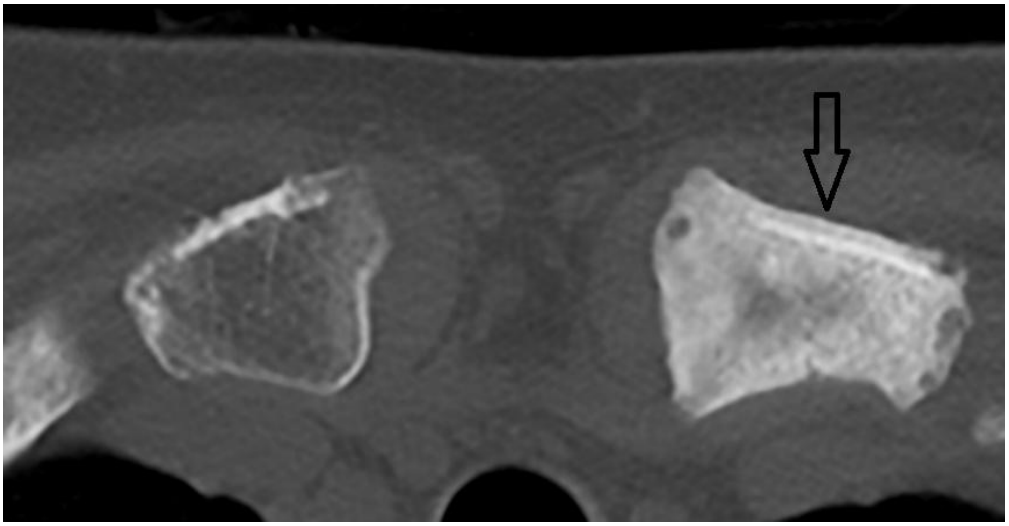
Figure 3

a



Description: Coronal CT reformatted images. Note the osteosclerosis of the left medial clavicle (large arrow) indicating osteitis. Bilateral erosions at the manubrium sterni (2 upward pointing arrows) and at the inferior border of the left clavicle (leftward pointing arrow). Subtle ligamentous ossification at the attachment of the costoclavicular ligament (2 downward pointing arrows). Also note the focal ossification at the costochondral junction of the first ribs (asterisk). **Origin:** © Filip Vanhoenacker, General Hospital Sint-Maarten, Mechelen, Belgium 2022.

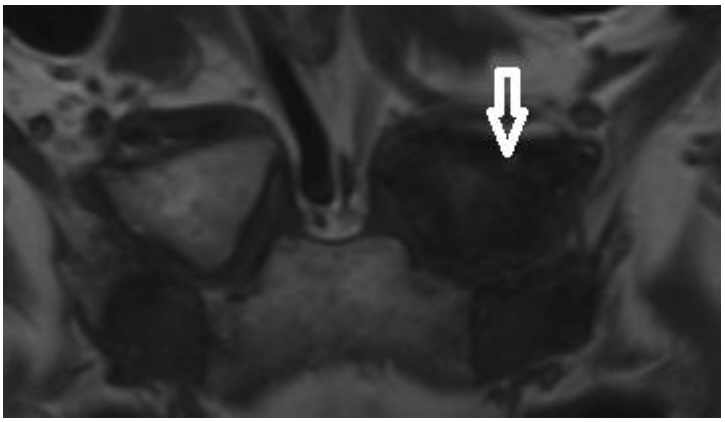
b



Description: Axial CT. Note the periosteal reaction of the clavicle with bone hypertrophy in keeping with hyperostosis. **Origin:** © Filip Vanhoenacker, General Hospital Sint-Maarten, Mechelen, Belgium 2022.

Figure 4

a

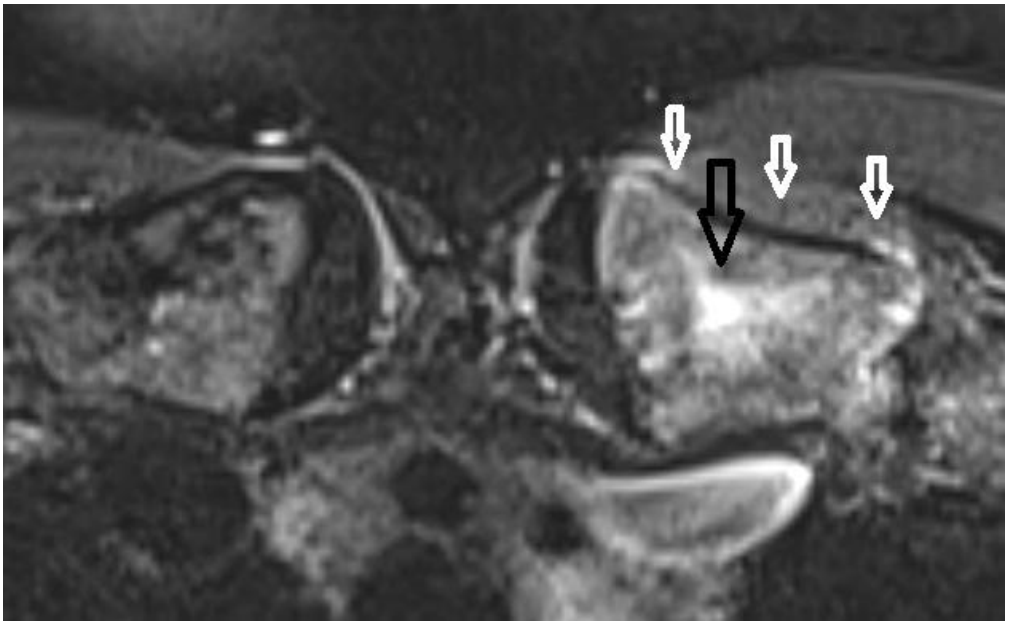


Description: Coronal T1-WI and FS T2-WI. The left clavicle is hypointense due to sclerosis (open arrow in a). FS T2-WI revealed also subtle bone marrow edema at the medial left clavicle (open arrow in b). **Origin:** © Filip Vanhoenacker, General Hospital Sint-Maarten, Mechelen, Belgium 2022.

b

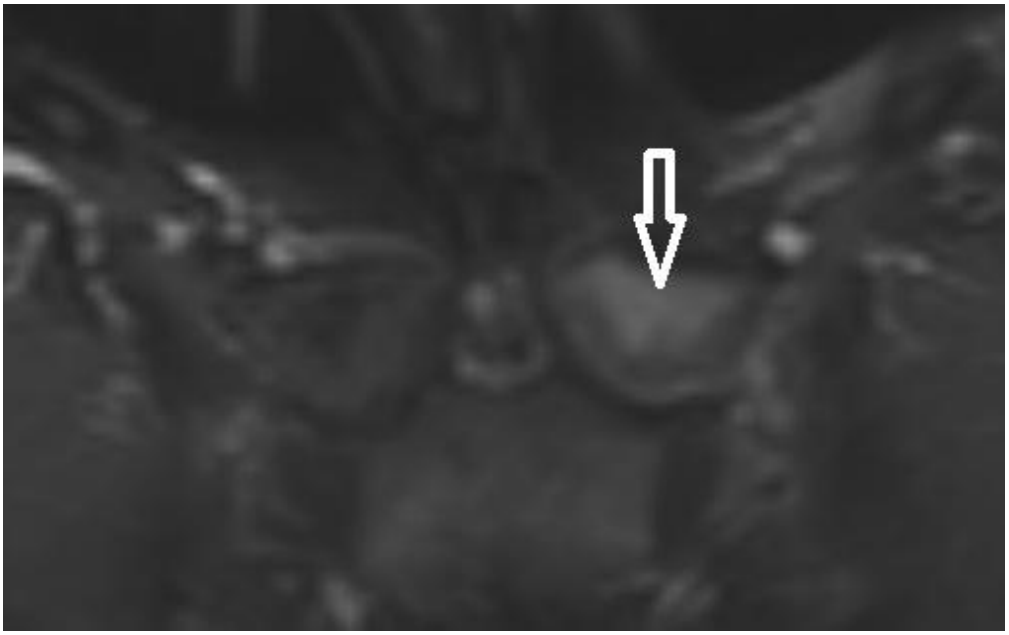
Description: Coronal T1-WI and FS T2-WI. The left clavicle is hypointense due to sclerosis (open arrow in a). FS T2-WI revealed also subtle bone marrow edema at the medial left clavicle (open arrow in b). **Origin:** © Filip Vanhoenacker, General Hospital Sint-Maarten, Mechelen, Belgium 2022.

c



Description: Axial FS T2-WI. Bone marrow edema in the left medial clavicle (black open arrow). Note also subtle soft tissue edema (white arrows). **Origin:** © Filip Vanhoenacker, General Hospital Sint-Maarten, Mechelen, Belgium 2022.

d



Description: Coronal FS TI-WI after gadolinium contrast administration. subtle enhancement of the medial left clavicle (open arrow). **Origin:** © Filip Vanhoenacker, General Hospital Sint-Maarten, Mechelen, Belgium 2022.