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Imaging of Osteoarticular Lesions in SAPHO Syndrome: A Case Report

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Abstract **Case Report**

***Introduction*:** Synovitis, acne, pustulosis, hyperostosis, osteitis (SAPHO) syndrome is an acronym for a variety of osteoarticular and dermatologic manifestations. Although the incidence and prevalence are probably underestimated, the syndrome is considered rare. ***Observation*:** A 45-year-old female patient, without any particular pathological history, presented with anterior chest pain, spinal pain and palmar pustulosis. The symptoms had been evolving for six months, in a context of apyrexia and conservation of the general state. The diagnosis of SAPHO syndrome was retained in view of the young age, the clinical presentation and the axial osteoarticular involvement on imaging. In addition, bone scintigraphy objectified the "bull's head sign". ***Discussion*:** The osteoarticular manifestations of SAPHO syndrome are characterized by osteosclerosis, hyperostosis, erosions, synovitis and synostosis. Some locations, especially the anterior chest wall, and some topographical associations are very suggestive. Clinical symptoms are non-specific, hence the interest of imaging. CT is the modality of choice to determine the extent of osteoarticular lesions. Magnetic resonance imaging has an important place. It allows to establish a correlation between the stage of the disease and the clinical symptoms. ***Conclusion*:** Knowledge of clinical manifestations and radiological features allows for early diagnosis and prevention of osteoarticular progression.

Keywords: Sapho syndrome, magnetic resonance imaging, osteosclerosis, hyperostosis, case report.

1. INTRODUCTION

SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, osteitis) is a combination of dermatologic and osteoarticular manifestations. It is a non-specific aseptic osteitis. The prevalence of SAPHO syndrome does not exceed 1/10,000 worldwide. It is probably underestimated due to its relatively recent occurrence [1]. SAPHO syndrome can occur at any age. It poses a problem of differential diagnosis mainly with infectious osteomyelitis, osteosarcoma and Paget's disease [2]. Through an observation and a review of the literature, we report the different radiological aspects of the osteoarticular involvement of SAPHO syndrome.

2. OBSERVATION

A 45-year-old female patient with no previous history of disease presented with anterior sternoclavicular and chondrocostal chest pain. In addition, she had localized ostealgia of the right clavicle, as well as cervical and dorsal spinal pain of a mixed predominantly mechanical nature. The patient presented with a palmar pustular rash as an extra-articular manifestation. The symptoms had been evolving for six months, in a context of apyrexia and conservation of the general state. Biological examination showed a minimal inflammatory syndrome with a normal blood count.

A frontal chest X-ray showed osteocondensation, hyperostosis of the sternal end of the first ribs and of the right clavicle (fig. 1a). X-ray of the pelvis showed an irregularity of the cortical bone of the ischium in favour of enthesitis. In the right sacroiliac joint, a fusion with disappearance of the joint space was observed. On the left side, a condensation of the joint margins was found in favour of sacroiliitis (fig.1b). Cervical spine profile radiography showed erosion of the anterior corner of C5 with C5-C6 syndesmophyte (fig.1c). Dorsolumbar spine X-ray showed a left convex scoliosis with D12-L1 syndesmophyte.

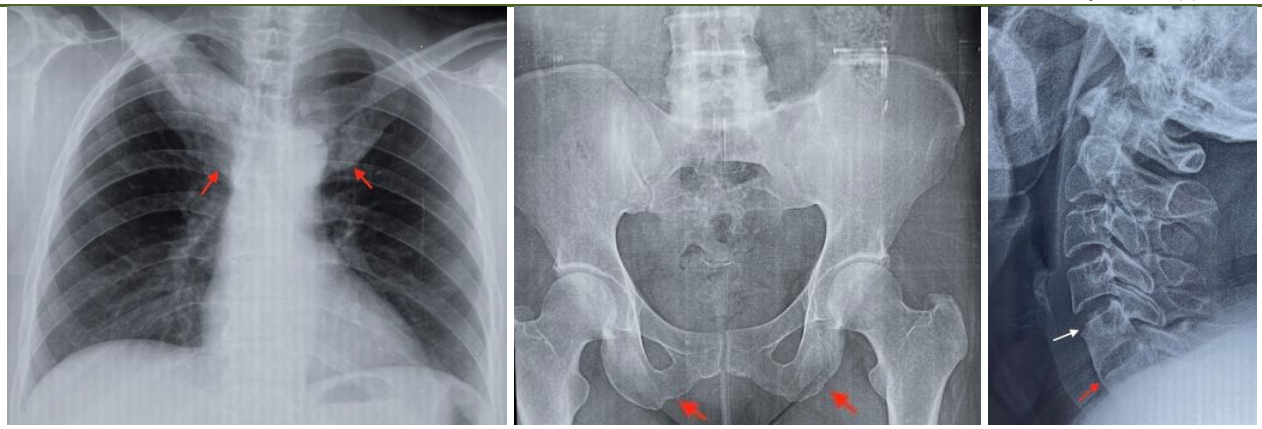

Figure 1: a) Frontal chest X-ray showing osteocondensation and hyperostosis of the sternal end of the bilateral first ribs and the right clavicle (arrows). b) X-ray of the pelvis showing irregularity of the cortical bone of the ischium in favour of enthesitis (arrows), fusion with disappearance of the interline of the right sacroiliac joint and condensation of the margins of the left sacroiliac joint in favour of sacroiliitis. c) Radiograph of the cervical spine in profile showing erosion of the anterior corner of C5 (white arrow) with C5-C6 syndesmophyte (red arrow).

Cervical MRI was requested in view of the painful right supraclavicular tumefaction. It showed a swollen appearance of the right clavicle, with loss of T1 hypersignal of the cancellous bone, without contrast after injection of gadolinium. There was an associated swollen appearance of the sternal manubrium and sternoclavicular joints without significant signal abnormality or contrast enhancement (Fig. 2a).

Complementary CT scan showed an enlarged and osteocondensed appearance of the right clavicle, sternal manubrium, sternoclavicular joints and the first sternocostal joints (Fig. 2b). Sternoclavicular ultrasound showed ankylosis of the right sternoclavicular joint and congestion on the left. Bone scintigraphy showed intense and diffuse bilateral hyperfixation of the clavicles suggestive of inflammatory involvement (Fig.3).

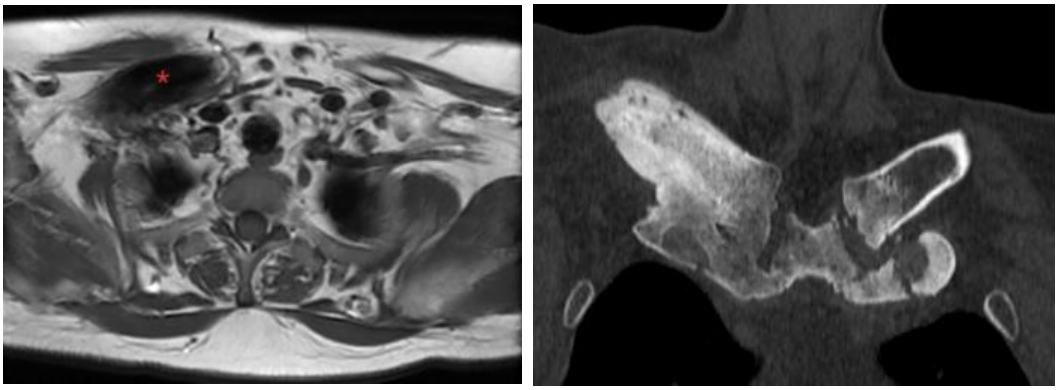

Figure 2: a) Cervical MRI axial section T1 sequence showing a swollen appearance of the right clavicle with loss of T1 hypersignal of the cancellous bone (star). b) CT scan of the thoracic bone window in coronal section, showing a hypertrophied aspect with osteocondensation of the right clavicle, the sternal manubrium, the sternoclavicular joints and the first sternocostal joints, with some lytic areas.

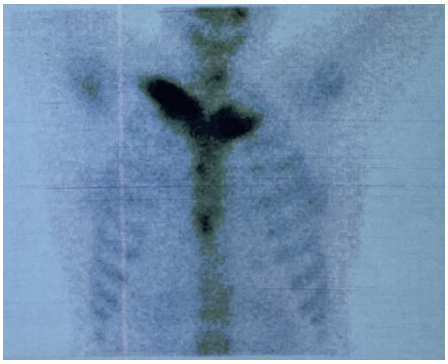

Figure 3: Bone scintigraphy showing intense and diffuse bilateral hyperfixation of the clavicles, predominantly on the right, giving a "bull's head sign"

The patient was put on a non-steroidal anti-inflammatory drug (NSAIDs) based on naproxen 500mg twice a day. The evolution was good under treatment with regression of the pain

3. DISCUSSION

Osteoarticular involvement is very characteristic of SAPHO syndrome. Osteolysis and hyperostosis are the mainstays of the diagnosis. In adults, the sternocostoclavicular region is the most frequently affected site. SAPHO syndrome shares clinical and radiological features with ankylosing spondylitis (AS), including sacroiliitis, enthesitis, paravertebral ossifications and ankylosis. Its consideration as a variant of AS is controversial [3].

Imaging of SAPHO syndrome combines anterior chest wall, spinal, sacroiliac and peripheral osteoarticular images. Conventional radiology can demonstrate lesions consistent with SAPHO syndrome with a low sensitivity of 13%. Sternocostoclavicular involvement is characterised by enthesitis, hyperostosis of more than 5 mm in diameter, and osteitis [1]. Spinal involvement is highly variable. It includes vertebral hyperostosis, osteolytic lesions with vertebral compression, spondyloarthritis with or without discitis, paravertebral ossifications and other syndesmophytes [3]. Sacroiliitis is usually unilateral with erosions and osteosclerosis along the iliac side of the joint [4].

CT is the modality of choice to determine the extent of osteoarticular damage to the anterior chest wall. Minor bony changes may be difficult to detect by magnetic resonance imaging (MRI) [5].

MRI is of great diagnostic value. Because of its performance in the exploration of entheses, this examination allows a correlation between the stage of the disease and the clinical symptoms. Furthermore, it can differentiate inflammatory spondylodiscitis from infectious pyogenic disease by showing the absence of micro-abscesses or epiduritis. The limited appearance of erosions is also in favour of a non-infectious cause. MRI demonstrates the effectiveness of the treatment. Remission is accompanied by the restoration of a normal fat signal in the bone marrow [1].

Indeed, MRI can detect oedema of active lesions in the bone marrow and soft tissue. The oedema appears as hypointense in T1 and hyperintense in STIR and T2. Chronic sclerotic bone lesions may appear hypointense on T1, T2 and STIR sequences. Sequences after gadolinium injection are used to differentiate SAPHO syndrome from malignancies and infections. Due to the multifocal nature of the disease, whole-body MRI is increasingly used to detect multifocal bone lesions, both at baseline and at follow-up [5].

Bone scintigraphy can also identify subclinical locations. Symmetrical hyperfixation at the sternocostoclavicular joints giving the bull's head sign is highly suggestive of SAPHO syndrome [4].

Bone biopsy is only performed in atypical forms. It does not provide any specific information, but in doubtful cases it can be used to exclude an infectious or tumorous pathology [1].

The treatment of SAPHO syndrome is essentially symptomatic with NSAIDs, but their effectiveness is low. Bisphosphonates are one of the new therapeutic modalities available. Osteoarticular involvement in SAPHO syndrome generally has a good prognosis. However, it may be complicated by superior vena cava syndrome due to extrinsic compression by hyperostosis [2].

4. CONCLUSION

The detection of SAPHO syndrome has been greatly facilitated by the advent of new imaging methods, dominated by nuclear magnetic resonance. Cooperation between rheumatologists, dermatologists and radiologists will improve the quality of life of SAPHO patients.

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