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Contribution of Bone Scintigraphy in the Diagnosis of a Case of SAPHO in the Nuclear Medicine Department of Idrissa Pouye General Hospital (Dakar, Senegal)

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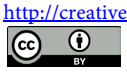
Abstract

Introduction: The acronym SAPHO (Synovitis, Acne, Pustulosis, Hyperostosis and Osteitis) is a syndrome combining osteoarticular and cutaneous manifestations. It occurs mainly between the ages of 30 and 50. Sternocostoclavicular hyperostosis is one of the main distinguishing features. We report a case of SAPHO in Dakar diagnosed by bone scintigraphy. **Observation:** 28-year-old Senegalese women presented with left shoulder pain and relative functional impotence for over 2 years. Examination revealed right sternoclavicular hyperostosis and left shoulder pain on palpation. Questioning revealed a history of acne and hyperostosis of the right first toe. Bone scintigraphy, performed after injection of 630 MBq of ^{99m}Tc-HMDP, revealed: hyperfixation of the bilateral (right++) manubrio-sternal and sternoclavicular junction, producing the classic bull's horn image; hyperfixation of the left shoulder with an inflammatory appearance; hyperfixation of the sacroiliac joints suggestive of bilateral sacroiliitis; hyperfixation of the right first toe; two mandibular hyper fixations probably related to dental damage. This scintigraphic appearance in one was strongly suggestive of SAPHO syndrome. **Conclusion:** SAPHO syndrome, related to spondyloarthropathy, associates



cutaneous and osteoarticular signs. It is characterized by frequent delays in diagnosis due to poor recognition. Soy is an invaluable diagnostic tool, enabling us to assess the extent of the disease and its evolution.

Keywords

SAPHO, Bone Scintigraphy, Young Woman, Senegalese

1. Introduction

SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) syndrome is a rare entity that combines a heterogeneous set of cutaneous and osteoarticular manifestations, with a common aseptic inflammatory process. It associates osteoarticular disorders (synovitis, hyperostosis, osteitis), essentially sternocostoclavicular, with skin abnormalities (acne, pustulosis) [1] [2] [3].

SAPHO syndrome is known as "Skibo-disease" (short for skin-bone), *i.e.* a combination of bone and joint manifestations linked to dermatological signs [2].

It is a painful and disabling disease that affects women more frequently than men [2].

The etiopathogenesis of this syndrome remains debated. While some consider it to be a spondyloarthropathy, others consider it to be of infectious origin. Diagnosis is based on the presence of at least one of the three diagnostic criteria proposed by Kahn [1].

This syndrome is considered rare, occurring mainly in young adults between the ages of 30 and 50. In adults, sternocostoclavicular hyperostosis is one of the main distinguishing features.

Diagnosis is based on a combination of clinicobiological and radiological criteria, and may be pathognomonic on bone scintigraphy [4]. Bone scintigraphy (BS) with ^{99m}Tc-HMDP is an invaluable diagnostic tool, as radiotracer fixation can precede clinical and even radiological manifestations, particularly in the anterior thorax [5].

We report a case of SAPHO diagnosed on bone scintigraphy at the nuclear medicine department of Idrissa Pouye General Hospital in Dakar.

2. Observation

This was a 28-year-old Senegalese woman presenting with left shoulder pain and relative functional impotence for over 2 years. This condition had prompted several consultations, notably in neurology, orthopaedics and rheumatology, with transient remissions.

Examination revealed hyperostosis of the right sternoclavicular joint (**Figure 1**) and left shoulder (**Figure 2**), painful on palpation and mobilization. Questioning revealed a history of acne and hyperostosis of the right first toe.

The biological workup, carried out two months in advance, showed a discrete inflammatory syndrome with a C-reactive protein elevated to 7 mg/L (normal < 6 mg/L), a sedimentation rate of 15 mm at the first hour, and an absence of in-

fectious syndrome with a white blood cell count equal to 6.1.103/mm^3 (norms: 4.103 to 10.10/mm^3). Calcemia was normal at 93 mg/l (84 - 102 mg/l). The blood culture was negative and no bacteria were found on bacteriological examination.

Shoulder ultrasound and spinal cord MRI (absence of bone erosion and synovitis) were normal.

Hand and chest X-rays (absence of hyperostosis, condensing lesions and osteolytic lesions) were also unremarkable.

In this context, whole-body bone scintigraphy (anterior and posterior) was performed 3 hours after injection of 630 MBq of ^{99m}Tc-HMDP, using a SPECT gamma camera (dual-head, Mediso Nucline TM Spirit DH-V type). It revealed:

- Hyperfixation of the bilateral (right +++) manubrio-sternal and sternoclavicular junction, producing the classic bull's horn image (**Figure 3**);
- Inflammatory hyperfixation of the left shoulder;
- Hyperfixation of the sacroiliac joints, suggestive of bilateral sacroiliitis (**Figure 4**);
- Hyperfixation of the right first toe (**Figure 5**);
- Strengthening the fixation of the pubic symphysis and dorsolumbar spine;
- Two mandibular hyperfixations (**Figure 5**).

Figure 1. Sternoclavicular hyperostosis.

Figure 2. Left shoulder hyperostosis.

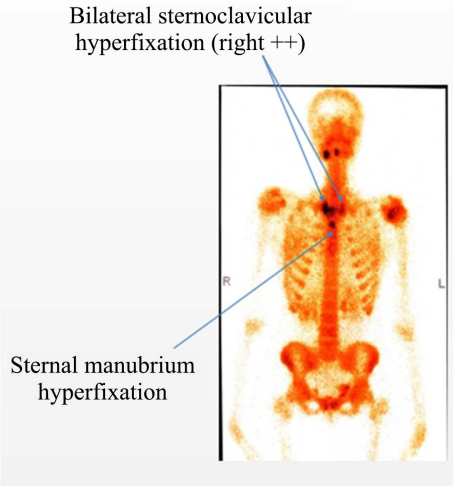


Figure 3. Bone scintigraphy (anterior side): Bull's horn appearance.

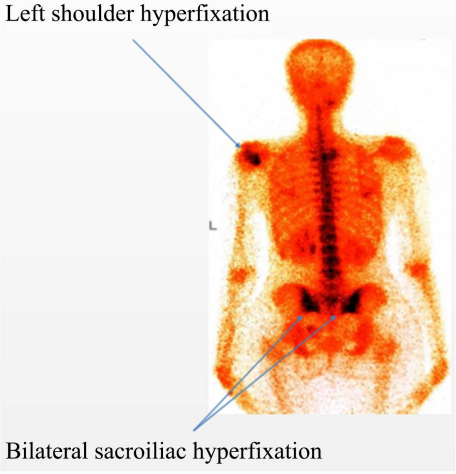


Figure 4. Bone scintigraphy (posterior side): Sacroiliacs and left shoulder inflammatory disorders.

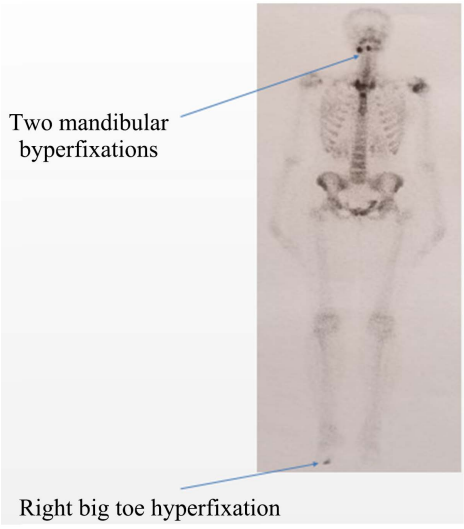


Figure 5. Bone scintigraphy (anterior side): mandibular and right big toe hyperfixations.

This bull's horn scintigraphic appearance, in a context of hyperostosis and a history of acne, was strongly suggestive of SAPHO syndrome.

The thoracic CT scan, in comparison with the bone scan, was performed 24 hours later, and found no abnormalities (absence of hyperostosis and signs of osteitis).

His treatment included analgesics, anti-inflammatories and physiotherapy, with significant improvement in functional impotence of the left shoulder.

3. Discussion

The origin of SAPHO syndrome is unknown, but it is generally considered to be an auto-inflammatory disease, *i.e.* due to a deregulation of innate immunity, as opposed to autoimmune diseases in which it is the acquired/adaptive immunity system that is deregulated [6].

The frequency of this pathology is very low, which is why studies on it are rare, and randomized controlled trials on the subject are totally non-existent. As a result, the pathophysiology and efficacy of various treatments are poorly understood. This is particularly problematic when it comes to drawing up recommendations for diagnosis and therapeutic strategies. In fact, there are no official recommendations [6].

Our patient was a 28-year-old Senegalese woman. It is reported in the literature that SAPHO syndrome affects more young adults between 30 and 50 years of age [1] [3] [7] [8].

The average age of onset of the first symptoms is between 35.8 and 38.5 years; they almost never appear after the age of 60, and the average diagnosis is made at around 45.5 years [6] [9] [10]. Our patient was diagnosed slightly earlier, at the age of 28. However, this syndrome does not spare the elderly, according to Chamot A. M. *et al.* [11].

C. Ghomari reported a case of SAPHO in a 56-year-old patient [4]. Occurrence in children is rare, but possible [1] [3].

Female predilection is also described in the literature [3] [4] [11].

However, according to I. Gharsallah [1] and Assia Haddouche [7], it affects both sexes equally, with a sex ratio close to 1.

This is the second case of SAPHO in Senegal diagnosed by bone scintigraphy, following that of NDong Boucar *et al.* in 2010 [8].

Globally, the prevalence of SAPHO syndrome is poorly known, with probable variations from one continent to another [12]. According to Sophie Przybylowski [6], the prevalence does not exceed 1/10,000 worldwide and in the global Caucasian population. It is probably underestimated due to the relatively recent individualization of the disease and a lack of knowledge of all its components [1] [7]. In France, it is estimated to affect 1 in 50,000 people [6]. The syndrome appears to be more prevalent in the Caucasian population, although some studies have focused on other populations, notably Japanese and North African [6]. According to Assia Haddouche, SAPHO syndrome has mainly been described in Japan and Northern Europe, but rarely in Anglo-Saxon countries [7].

In our case, the clinical picture was dominated, as widely described in the literature, by osteoarticular manifestations (painful hyperostosis of the right sternoclavicular joint, left shoulder and big toe) and dermatological manifestations with a history of acne. In fact, the manifestations are primarily cutaneous and osteoarticular [7] [12].

The diagnosis was made on the basis of the following arguments:

- Clinical: anterior thoracic swelling tender to palpation, localized in the right sternoclavicular region (most frequent localization) and the notion of acne one year ago (skin involvement in SAPHO syndrome may precede bone involvement with a delay of one to two years, up to 20 years);
- Para-clinical findings in favour of chronic aseptic inflammatory osteitis based on an inflammatory syndrome, with absence of hyperleukocytosis on biology, negative blood culture on bacteriological examination and the very characteristic bull's horn appearance on bone scintigraphy [13] [14].

Anterior thoracic involvement is one of the main distinguishing features of SAPHO syndrome. In the mono-centric series from Bichat hospital, its prevalence was measured at 63%. Earlier, several publications had already drawn attention to the elective involvement of the sternocostoclavicular region, with prevalence as high as 100% for Sonozaki *et al.* [12] [15].

I Gharsallah [1] estimates the frequency of articular damage to the anterior thoracic wall at 65% - 90%, affecting all anatomical components, but in particular the medial fragment of the clavicle. It manifests as inflammatory pain in the anterior-upper part of the thorax, which may radiate to the neck and shoulders [1].

Clinically, the initial signs consist of an inflammatory, painful swelling involving one or two anterior chest wall joints, primarily sternoclavicular, less frequently manubrio-sternal or chondrosternal. Patients frequently describe irradiation to the shoulder or trapezius, sometimes exclusively in the case of sternoclavicular involvement. More rarely, the initial area of pain is centred on one of the bony parts (clavicle, sternum), with later swelling resulting from the spread of osteitis to the adjacent soft tissues. Secondly, the almost systematic transition to chronicity leads to a progressive extension of inflammatory phenomena, with the dual characteristic of inducing erosive joint damage and, above all, bone hypertrophy, sometimes considerable. The latter primarily affects the clavicles, with the formation of hyperostosis and periostosis, sometimes of major proportions, which may themselves be complicated by compression and then subclavian phlebothrombosis, with possible collateral circulation [11] [12] [16] [17].

Along with sacroiliac involvement, this type of manifestation obviously supports the diagnosis of spondyloarthropathy, and usually indicates greater severity. However, this axial involvement may also remain clinically silent, only to be detected by various imaging techniques [12].

In 1994, Kahn *et al.* defined three criteria for diagnosis:

- Aseptic multifocal osteomyelitis, with or without skin lesions;

- Acute or chronic joint involvement associated with palmoplantar pustulosis, palmoplantar pustular psoriasis or severe acne;
- Aseptic mono- or polyostotic osteitis associated with palmoplantar pustulosis, palmoplantar pustular psoriasis, and severe acne.

The presence of just one of these criteria is sufficient to establish the diagnosis. However, the appearance of dermatological signs makes it easier to establish the diagnosis.

Biological blood tests are especially useful for diagnosing pathologies associated with the syndrome, such as chronic inflammatory bowel disease [3].

Tc99m bone scintigraphy is an invaluable diagnostic tool, as isotope fixation can precede clinical and even radiological manifestations, particularly in the anterior thorax [5].

On scintigraphy, all these bone foci, which correspond to foci of aseptic osteitis, strongly fix the tracer. Scintigraphy can guide the diagnosis when it reveals foci on the clavicle and sternum, with the classic bull's horn appearance [8]. So is also very useful for assessing the extent of the disease and its evolution [15].

Skin biopsies are rarely performed, but can be useful in eliminating infectious causes, particularly staphylococcal infections. If the biopsy reveals the presence of *P. acnes* in the skin lesions, the doctor will preferentially prescribe the antibiotics classically used in acne [6].

The etiopathogenesis of SAPHO syndrome remains mysterious or poorly understood [1] [7]. However, certain mechanisms seem to be becoming clearer, in particular, the terrain of onset and the role of certain infectious agents. Propionibacterium acnes has been incriminated in SAPHO syndrome, since it has been isolated within bone lesions. In the series by Assman *et al.*, it was isolated from 67% of patients undergoing bone biopsy. However, this infectious theory remains controversial, partly because P. acnes is often found to be negative by polymerase chain reaction (PCR), and partly because of the inconsistent efficacy of antibiotics and the non-aggravation of lesions after corticosteroid infiltration. A German team has demonstrated strong expression of tumour necrosis factor (TNF) within a mandibular osteitis lesion observed in SAPHO syndrome, suggesting TNF involvement in the genesis or maintenance of osteoarticular damage. Familial forms have been described. The predisposing genetic background appears to be similar to that of spondyloarthropathy, since the HLA B27 antigen is found in 4% to 18% of cases of SAPHO syndrome. However, results in different series remain discordant, and the role of this antigen remains controversial. The frequent association of Crohn's disease (CD) with SAPHO syndrome reinforces the hypothesis of a genetic mechanism via mutation of the NOD2/CARD15 gene [1].

There is no well-coded treatment for SAPHO syndrome. It is essentially based on NSAIDs (non-steroidal anti-inflammatory drugs) or sulphonamides such as sulfasalazine. Dosage must be adapted to each individual patient, taking side effects into account [2].

Their efficacy is often inadequate, which explains why they are generally com-

bined with other treatments (sulfasalazine or methotrexate). A drug in the bisphosphonate class (pamidronate) has shown favourable action not only on pain, but also on pustular lesions [8].

Corticosteroids should only be prescribed very rarely, and are only indicated in emergencies and for a very limited duration. There have been a few trials with various immunomodulators, including anti-TNF-alpha. In most cases, treatment with methotrexate is initiated [2].

The disease never progresses to carcinogenesis. In some people, symptoms may even disappear spontaneously, but in all cases the evolution is unpredictable [8].

Therapeutic evaluation is difficult, in the absence of strict controlled studies. To date, treatment of SAPHO remains symptomatic. However, the management of SAPHO needs to be multidisciplinary [2].

The long-term prognosis of SAPHO syndrome is favourable, with remissions lasting several years followed by new exacerbations [6].

4. Conclusion

SAPHO syndrome, related to spondyloarthropathy, associates to varying degrees cutaneous and osteoarticular signs. Diagnosis is frequently delayed due to a lack of awareness of the syndrome. ^{99m}Tc-HMDP bone scintigraphy is an invaluable diagnostic tool, as isotope fixation can precede clinical and even radiological manifestations, particularly in the anterior thorax. Bone scintigraphy can also be useful in assessing the extent of damage and its progression.

Conflicts of Interest

None.

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