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SAPHO Syndrome: About an Observation and Review of the Literature

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Abstract | **Case Report**

Introduction: SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) is autoinflammatory rheumatism of the spondyloarthritis group. It is rare, it prevalence is 1/10000 inhabitants. The average age at diagnosis is 38 years, with no gender predominance, first described by Chamot and colleagues in 1987. ***Objective:*** To present observation and review of the literature on SAPHO syndrome which is rarely diagnosed in our daily practice. ***Observation:*** This was a 28 year old female patient, Senegalese, living in Dakar, followed in obstetrics gynecology for the desire of maternity, the family context of sickle cell disease who consulted for diffuse mechanical and articular inflammatory pains involving the proximal interphalangeal (IPP) of the left 5th toe, the left shoulder, the sternocostal and dorsolumbar joints evolving for 3 years. The clinical examination found: A painful swelling with a visual analog scale (VAS) of 6/10 of the right sternocostal joint and of the interphalangeal joint (IP) of the right hallux, diffuse acne, no palmoplantar pustulosis, no psoriasis, no gluteal or talalgia. The biology shows an inflammatory syndrome (VS at 37 mn at the 1st hour and CRP at 41mg/l), a decrease of 25-OH-Vitamin D3 at 22 ng/ml (N: 25-30 ng/ml), the positive latex test, Ac anti-CCP: 43 IU/ml (N < 5 IU/ml), positive Ac anti-HBc , Ac anti HBs > 1000 IU/l on the other hand blood calcium, blood phosphorus, G6PD, PTH, TSH us, free T4, anti ECT Ac, HLA-B27 Ag with no particularities. The scintigraphy showed hyper fixation of the right 1st toe, sacroiliac joints, dorsolumbar spine, left shoulder, manubrium sternal junction, and bilateral sternoclavicular junction realizing a classical bull horn image in SAPHO syndrome. Because of this clinical and paraclinical manifestation helped by the 1994 diagnostic criteria of Kahn at 1/3 (chronic joint involvement associated with severe acne), the SAPHO syndrome was retained and the treatment instituted after the normal pre-therapeutic analysis and systematic deworming was: Salazopyrine 2000mg/day (dosage reached by increments of 500 mg/week); Uvedose (vitamin D3) 100000 IU / month; bisphosphonates 70 mg /week and paracetamol + codeine 1g x 2/day as needed for 3 months with a favorable follow-up by the regression of signs (pain VAS at 2/10). ***Conclusion:*** The SAPHO syndrome is the prerogative of the young adult subject, bone scintigraphy is very valuable for its diagnosis, its better knowledge would avoid diagnostic and therapeutic erratic.

Keywords: SAPHO syndrome, bone scan, Dakar, Senegal.

INTRODUCTION

SAPHO (synovitis, acne, pustulosis, hyperostosis and osteitis) is autoinflammatory rheumatism of the spondyloarthritis group with a genetic component that affects bones, joints and skin, It is rare, its prevalence is 1/10000 inhabitants [1]. It affects children and adults, the average age at diagnosis is 38 years, and the sex ratio male/female is 1 [2]. Proposed in 1987 by the French team [3]. rarely reported in Africa. We report an observation with a review of the literature.

OBSERVATION

This was a 28-year-old patient with the initials L S, Senegalese, residing in Dakar, followed in obstetrics gynecology for the desire of maternity, having a benign nodule in the left breast, tonsillectomy performed in 2012, the family context of sickle cell disease who consults for diffuse mechanical and articular inflammatory pain involving the proximal interphalangeal (IPP) of the left 5th toe, the left

shoulder, the sternocostal and dorsolumbar joints evolving for 3 years.

The clinical examination finds : a good general state, anxiety, a painful swelling with the visual analog scale (VAS) at 6/10 of the right sternocostal joint (figure 1) and of the interphalangeal (IP) of the right hallux, diffuse acne, no palmoplantar pustulosis, no psoriasis, no gluteal pain or talalgia.

The biology shows an inflammatory syndrome (VS at 37 mn at the 1st hour and CRP at 41mg/l), a decrease of 25-OH-Vitamin D3 at 22 ng/ml (N : 25-30 ng/ml), the positive latex test, Ac anti-CCP : 43 IU/ml (N < 5 IU/ml), positive Ac anti-HBc, Ac anti HBs > 1000 IU/l on the other hand blood calcium, blood phosphorus, G6PD, PTH, TSH us, free T4, anti ECT Ac, HLA-B27 Ag without any particularity.

The scintigraphy showed hyper fixation of the right 1st toe, sacroiliac joints, dorsolumbar spine, left shoulder, manubrium-sternal junction and bilateral sternoclavicular junction realizing a classical bull horn image in SAPHO syndrome.

Because of this clinical and paraclinical manifestation helped by the 1994 diagnostic criteria of Kahn at 1/3 (chronic joint involvement associated with severe acne), the diagnosis of SAPHO syndrome was retained and the treatment instituted after the normal pre-therapeutic analysis and systematic deworming was: Salazopyrine 2000mg/day (dosage reached in steps of 500 mg/week); Uvedose (vitamin D3) 100000 IU/month; bisphosphonates 70 mg/week and paracetamol + codeine 1g x 2/day as needed for 3 months which gave a favorable outcome by the regression of signs (pain VAS 2/10).

Figure 1: Painful swellings of the anterior chest wall in a 28-year-old female patient with SAPHO syndrome in the rheumatology department of Aristide Ledantec Hospital in Dakar, Senegal

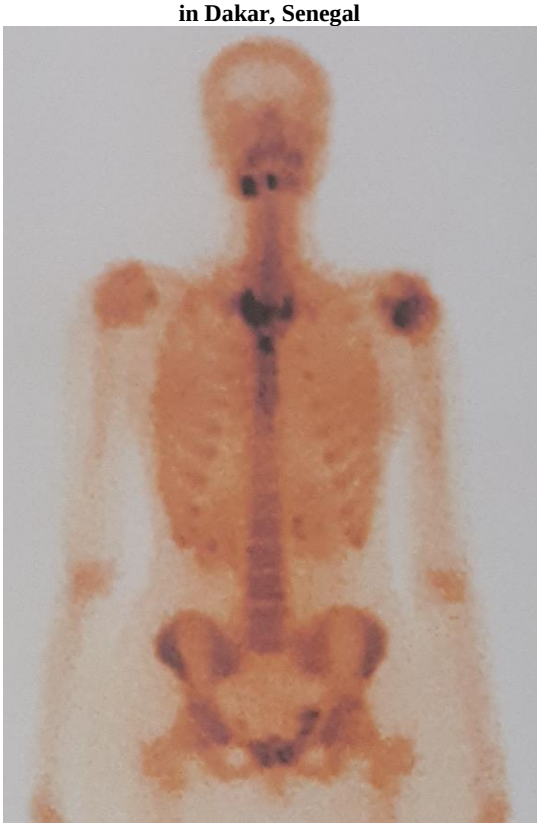

Figure 2: Scintigraphy of the 28-year-old patient showing hyper fixation of the sacroiliac, dorsolumbar spine, left shoulder, manubrium-sternal junction and bilateral sternoclavicular making a classic bull horn image in SAPHO syndrome at the rheumatology department of the Aristide Ledantec Hospital in Dakar, Senegal

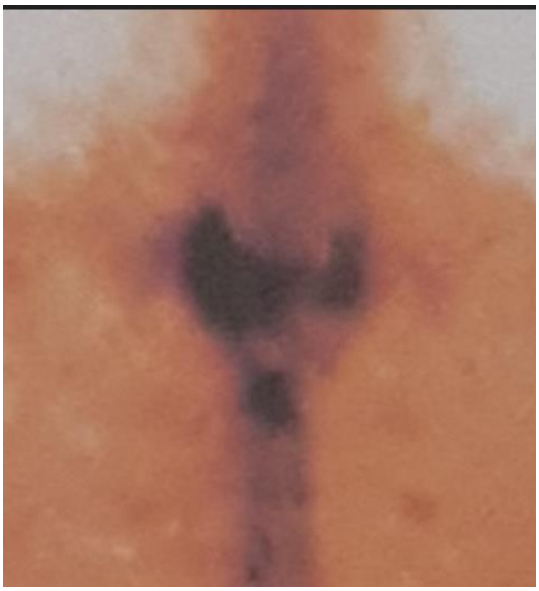

Figure 3: Bilateral sternoclavicular hyper fixation realizing a classic bull horn image in SAPHO syndrome in a 28-year-old female patient in the rheumatology department of Aristide Ledantec Hospital in Dakar, Senegal.

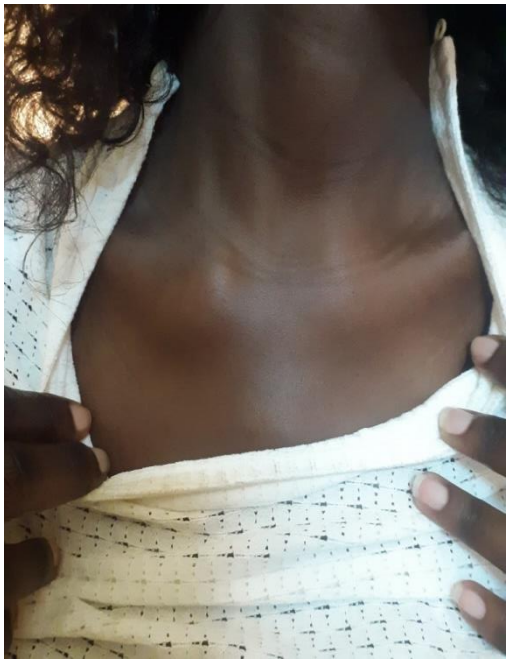

Table 1: Khan criteria for inclusion in SAPHO syndrome. The presence of only one of the 3 criteria is sufficient [15]

1. Chronic multifocal osteitis	without skin lesions reaching thorax, spine, sterile pelvis
2. Acute or chronic joint involvement associated with	palmoplantar pustulosis palmoplantar pustular psoriasis severe acne hidrosadenitis (Verneuil)
3. Sterile mono- or polyostotic osteitis associated with	palmoplantar pustulosis palmoplantar pustular psoriasis severe acne hidrosadenitis (Verneuil)

DISCUSSION AND COMMENT

Our observation is that of a SAPHO syndrome which is of epidemiological, diagnostic, therapeutic and prognostic interest.

The SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis and osteitis) is rare, its prevalence is 1/10000 inhabitants [1]. It affects children and adults, the elderly are not spared, the average age at diagnosis is 38 years, and the sex ratio male/female is 1[2]. Chamot and colleagues were the first to describe the disease in 1987 [3]. Few cases have been reported in the United States [4]. In Africa, it is less reported. In the Maghreb, only one Tunisian case has been published [5].

Our patient presented with bone and joint pain, which is in agreement with the data in the literature according to which pain in superficial bone structures varies according to the topography of the structures affected ; asymptomatic forms have also been described. The main location is the anterior chest wall (APW) in 65-90%, it is the prerogative of the young adult and all its anatomical structures can be involved : the clavicles (inner third), the sternum, the sterno-costo-clavicular joints and the ribs [6]. The spine is involved in 32-52% of cases in the form of spondylodiscitis or posterior inter-apophyseal arthritis [6].

Tc99m bone scintigraphy is very valuable for the diagnosis as the fixation of the isotope may precede the clinical and even radiological manifestations, especially in the anterior thorax [7]. This is a comfort to our result where the scintigraphy showed hyper fixation of the bilateral manubrium-sternal and sternoclavicular junction realizing a classical bull horn image in SAPHO syndrome. The skin involvement (diffuse acne) of our patient is consistent with the literature in which it is stated that it is present in 89% of cases and is not mandatory to evoke the diagnosis [8, 9]. It can be acne conglobata, acne fulminans, palmoplantar pustulosis or pustular psoriasis. It may precede, accompany or occur long after the osteoarticular involvement [10, 11].

SAPHO syndrome remains a non-disabling condition with a good prognosis [12]. Its treatment is highly variable and aims to put the disease into remission ; therapies are chosen according to the type of involvement (bone or joint). The reference medical treatment involves non-steroidal anti-inflammatory drugs (NSAIDs), which have shown modest effectiveness [12, 13]. Our patient received salazopyrine and bisphosphonates, which is in line with the literature according to which bisphosphonates act on bone remodeling by inhibiting bone resorption, they control the flare-ups of the disease thanks to their anti-inflammatory faculty [14].

CONCLUSION

SAPHO syndrome is auto-inflammatory rheumatism of the spondyloarthritis group, it is the prerogative of the young adult, with no predominance.

The diagnosis is based on clinical signs (bone and joint pain) and paraclinical signs (hyper fixation of the painful parts through the very valuable bone scan) following the 1994 Kahn diagnostic criteria.

Its management is based on NSAIDs as a first line treatment which improves the functional and vital prognosis.

It is important to recognize this syndrome to avoid invasive and repetitive diagnostic procedures and the prescription of therapies.

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DECLARATION OF INTEREST: No interest.

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