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The lobster sign in SAPHO syndrome: unusually extensive osteitis of the anterior chest wall partially responsive to infliximab

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To the Editor,

The SAPHO (synovitis, acne, pustulosis, hyperostosis, osteitis) syndrome is an uncommon entity comprising several osteoarticular and cutaneous features [1]. Osteitis and hyperostosis remain key diagnostic features, since the proposed clinical criteria [2] have never been validated, especially regarding the distinction between the SAPHO syndrome and psoriatic arthritis [3]. Tumor necrosis factor-alpha (TNF- α) antagonists are starting to play an important role in the treatment of patients inadequately responsive to conventional treatment [4].

We present a 48-year-old female patient diagnosed with SAPHO syndrome five years ago. She has been treated with nonsteroidal anti-rheumatics, sulfasalazine and methylprednisolone, with partial and unsatisfactory response in terms of clinical and laboratory features, as well as radiological and bone scan findings [5, 6]. Four years after initiating conventional treatment she underwent a reevaluation to assess disease extent and activity. Physical examination revealed palmoplantar pustulosis and multiple joint tenderness, including sternoclavicular, costochondral, sacroiliac and peripheral joints. Laboratory investigation revealed elevated inflammatory markers, also suggesting disease activity.

A technetium 99-m bone scan was subsequently performed [5]. Increased tracer uptake was observed in both sternoclavicular joints, the sternum, first ribs bilaterally, fifth and sixth ribs near the costosternal junctions and the anterior portion of the eighth left rib, resembling a lobster. It was also revealed in the right hip and pubic bone, as well as in the pubic symphysis (Figure 1). Less pronounced accumulation was noticed in the L4 and L5 vertebrae.

Infliximab was added to the treatment, leading to an almost complete regression of osteoarticular complaints and normalization of laboratory findings. However, a follow-up bone scan performed after the fourth application of infliximab revealed a pattern of tracer accumulation almost identical to the one described previously. Moreover, psoriasiform skin lesions developed on the palms and trunk following the introduction of the biological agent: although similar lesions were observed before, they were now more pronounced. The skin lesions disappeared within several weeks following the fifth application of infliximab.

TNF- α antagonists are included in standard treatment strategies for seronegative spondyloarthropathies; however, their use in the SAPHO syndrome is still considered as off-label [4]. This might change in the future due to new insights into their role on the molecular level [7]

and an increasing number of individual reports suggesting a positive impact on disease activity [4]. Nevertheless, some questions still remain to be answered. Our patient experienced a temporary aggravation of cutaneous lesions, which is in accordance with other authors' findings [8]. The aggravation is probably a side effect of infliximab and not a result of worsening of the disease course. Furthermore, the impact of infliximab on bone tracer uptake should also be addressed. Although an alleviation of osteoarticular complaints was observed soon after the beginning of biological treatment, no regression was observed on the control bone scan after the fourth application of infliximab. In conclusion, there is a need of larger-scale prospective clinical trials, not only to establish evidence-based indications, but also to assess longer-term efficacy of TNF- α antagonists and build reliable follow-up algorithms.

Conflict of Interest

The authors declare that they have no conflict of interest.

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Fig. 1 Whole body technetium 99-m bone scan – (a) anterior and (b) posterior view. Increased tracer uptake was observed in both sternoclavicular joints, the sternum, first ribs bilaterally, fifth and sixth ribs near the costosternal junctions and the anterior portion of the eighth left rib. It was also revealed in the right hip and pubic bone, the pubic symphysis, as well as in the L4 and L5 vertebrae

