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CASE REPORT

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Aseptic spondylitis as the initial manifestation of the SAPHO syndrome

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Abstract We describe the case of a 61-year-old female patient who presented with spondylitis of the lumbar spine. Although the microbiological cultures of the bone biopsy specimens obtained during laminotomy remained negative, the patient was treated with broad-spectrum antimicrobials for 2 months. Eight months later she started to suffer from pain and tenderness in her sternum and the medial portion of her left clavicle. The findings of computed tomography and gallium-labelled isotope scan were indicative of sternoclavicular arthritis. Again, all surgically obtained biopsy specimens yielded negative results in microbiological studies. The diagnosis of the SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteomyelitis) syndrome was then made based on the clinical presentation with recurrent sterile osteitis in two characteristic locations, and the patient was started on immunosuppressive therapy. This case is a reminder that SAPHO may sometimes occur without any skin manifestations. Since this type of patient may be admitted to an orthopedic ward, it is important that orthopedic surgeons are familiar with the syndrome.

Key words SAPHO · Aseptic spondylitis

Introduction

In acute osteomyelitis a specific etiology may be determined in 50%–80% of the patients; the disease is usually caused by bacteria [4]. Among other febrile illnesses mimicking osteomyelitis, a recently recognized clinical entity called the SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteomyelitis) syndrome constitutes an important diagnostic consideration [2, 9, 11, 13]. In a classic form, SAPHO presents with simultaneous bone and skin manifestations. Most commonly, the patient has an aseptic lesion of the spine and/or sternoclavicular region [2, 9, 11]. The typical skin manifestations associated with SAPHO include palmoplantar pustulosis, acne fulminans, and pustular psoriasis. They can either precede or follow the bone disease, or even be totally absent [9–11]. We describe a patient who initially presented with aseptic spondylitis of the lumbar spine. In her case, the diagnosis of SAPHO syndrome was made only 11 months later, after she developed a sterile osteoarticular involvement of her left sternoclavicular region.

Case report

The patient is a 61-year-old woman who presented in June 1995 with a 2-week history of fever, low back pain, and sciatica. On admission to hospital, physical examination revealed deep palpation tenderness in her low back. Patellar reflexes were present and equal, but the Achilles tendon reflexes were absent. There was a Lasegue's sign at 30° in the left leg and at 45° in the right one.

Laboratory examination revealed a hemoglobin value of 112 g/l, a white blood cell count of 3.5×10^9/l, an erythrocyte sedimentation rate (ESR) of 58 mm/h, and a serum C-reactive protein

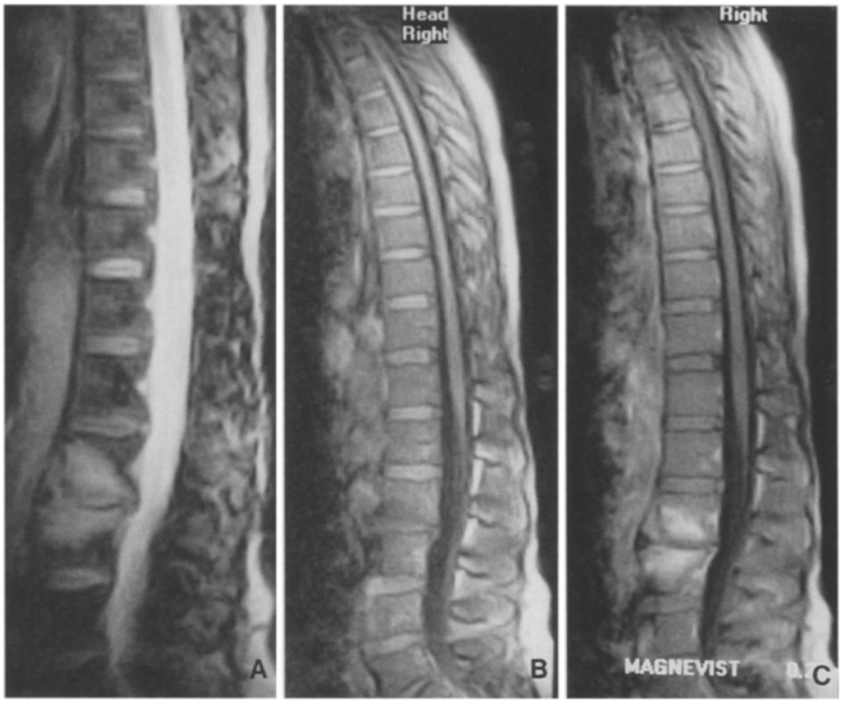

Fig. 1 MRI of the lumbar spine reveals high signal intensity of the L2 and L3 vertebral bodies on a T2-weighted image (**A**), a slightly increased signal intensity on a T1-weighted image (**B**), and distinct contrast-medium enhancement (**C**). An anterior soft tissue mass is also seen

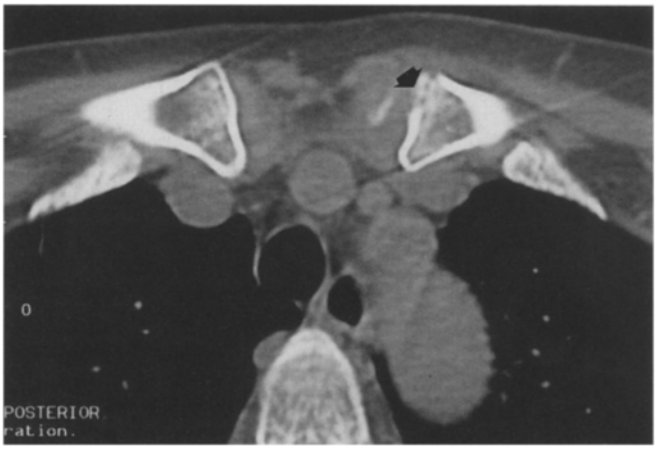

Fig. 2 Iohexol-enhanced computed tomography of the sternoclavicular region reveals erosion of the left clavicular bony joint surface (*arrow*), narrowing of the sternoclavicular joint space, and soft tissue edema, indicating arthritis

(CRP) value of 61 mg/l. Plain radiography and computed tomography (CT) of the lumbar spine revealed changes that were regarded as degenerative. However, MRI of the spine was suggestive of spondylitis of the L2 and L3 vertebrae (Fig. 1). Laminotomy was then performed at that level and revealed inflamed tissues; frank pus, however, was not obtained. Based on the operative findings combined with the clinical presentation of the disease, the diagnosis of spondylodiscitis was made and the patient was administered broad-spectrum antimicrobial treatment intravenously. Unexpectedly, all microbiological staining and culture specimens obtained during the operation yielded negative results, and the histopathological examination of the bone biopsy showed only scanty lymphocytic infiltration indicative of mild inflammation. Intravenous antimicrobial treatment was continued until the patient, 1 month later, was discharged in good condition receiving antibiotics perorally. The total duration of the antimicrobial treatment was 2 months.

In January 1996, the patient started to suffer severe pain and tenderness in her sternum and the medial part of her left clavicle. She had a low-grade fever of 37.8°C, and the serum CRP values varied between 66 and 102 mg/l. On admission to our orthopedic outpatient clinic her left sternoclavicular joint region was swollen, slightly erythematous and tender to palpation. Plain radiography showed no pathological changes. However, CT of the sternoclavicular region revealed findings indicative of arthritis (Fig. 2). Increased metabolic activity in the left sternoclavicular joint was also demonstrated on a gallium-labelled isotopic scan. At the end of March 1996, biopsy samples were obtained from the left sternoclavicular joint region. Again, all specimens for microbiological cultures and staining yielded negative results; histopathology revealed a small amount of lymphocytes as an indication of inflammation.

At this point, the diagnosis of the SAPHO syndrome was made, based on the clinical presentation of the patient with recurrent sterile osteitis in two characteristic locations [9]. Subsequently, she was administered peroral corticosteroids with an initial daily dose of 10 mg prednisolone. This treatment proved of no apparent benefit: the sternoclavicular pain on the left side persisted and the right sternoclavicular region also became affected. Moreover, the patient began to suffer from pain and tenderness in her fingers and wrist joints. In early August 1996, she developed frank synovitis in both her wrists as well as the second, third and fourth metacarpophalangeal joints. She has recently been started on methotrexate therapy.

Discussion

We describe here a patient who initially developed aseptic spondylitis and, later, an aseptic osteoarticular involvement of the sternoclavicular region. Thus, her clinical presentation fulfills the criteria of the SAPHO syndrome. Ac-

cording to French authors [9], recurrent sterile osteomyelitis can be considered sufficient for the diagnosis of SAPHO, even in the absence of skin symptoms. In our patient, the osteitic lesions affected the spine and anterior chest wall, which are the two preferred sites of bone involvement in the SAPHO syndrome [9, 11].

The SAPHO syndrome is classified among the rheumatological diseases with unknown etiology [2, 9], and the patients are usually attended by specialists in rheumatology. Moreover, the majority of cases have been published in rheumatological journals [2, 6, 10, 11, 13]. Therefore, the syndrome may still be unknown to many specialists in other fields. This disease entity was recognized several years ago, but had been previously presented under a variety of clinical diagnoses, including chronic recurrent multifocal osteomyelitis with pustulosis palmoplantaris, acne-associated spondylarthropathy, and sternocosto-clavicular hyperostosis with palmoplantar pustulosis [1, 3, 5–8, 12]. It is assumed that the cases belonging to the SAPHO syndrome represent overlapping manifestations of the same disorder. Since the introduction of the acronym SAPHO in the 1980s [2], knowledge about this syndrome has generally increased. However, the acronym can sometimes cause confusion, because all components of the syndrome are not needed for inclusion into this clinical entity [2, 9, 11, 13]. Patients who only have bone lesions without any skin or synovial manifestations are often initially admitted to orthopedic wards, as was the case with our patient. The diagnosis must then be made by orthopedic surgeons.

The clinical course of the SAPHO syndrome often fluctuates between periods of recovery and deterioration [9, 13]. At the present time, no optimal treatment for SAPHO is known. Antimicrobial therapy is usually not effective [2, 9]. Some patients may benefit from non-steroidal anti-inflammatory drugs, others from systemic corticosteroids. Patients with inflammatory arthritis can respond to intra-articular corticosteroid injections. In addition, there are reports of the successful use of sulphasalazine, colchicine, and methotrexate in anecdotal cases [9].

In conclusion, the possibility of SAPHO syndrome should always be considered in association with recurrent aseptic osteitis. This may save the patient unnecessary surgical procedures and administration of long-term antimicrobial agents. It is especially important that orthopedic surgeons be familiar with this syndrome.

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