

## Case Report

# SAPHO syndrome treated with pamidronate

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### ABSTRACT

A middle aged man presented with a 4 year history of painful swelling of both knees and pustular acne on his back. The MRI scan of the knees showed gross synovitis, so did the histopathology of the synovium. A diagnosis of SAPHO syndrome was made and he was treated with IV pamidronate. His pain reduced and acne completely cleared up on therapy. A dermatologist has rarely made a diagnosis of SAPHO syndrome and treated it.

**Key Words:** Acne, Synovitis, Pamidronate

### INTRODUCTION

French rheumatologists first diagnosed SAPHO syndrome after a nationwide survey in 1987. It is characterized by involvement of the skin and skeletal system. A rheumatologist usually makes the diagnosis, and a consultation with a dermatologist is sought to diagnose and treat the dermatological component. The acronym SAPHO stands for synovitis (inflammation of the synovium of a joint), acne (acne conglobata, fulminans, or the follicular retention triad), pustulosis (palmo-plantar pustulosis), hyperostosis (increase in bone substance), and osteitis (inflammation of bones).

### CASE REPORT

A 40-year-old man presented with painful swelling of both knees since 3 years that was followed by pustular acne on his back. The knees were swollen and painful, limiting his walking. The symptoms varied in intensity over periods of time, never coming back to normal, and at times so severe that he could not get out of his

bed. Medications he had received were a wide range of NSAIDs, systemic corticosteroids, colchicine, sulfasalazine, methotrexate and allopurinol.



**Figure:** MRI scan showing synovitis

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Clinical examination showed gross swelling of both knees, but there was no tenderness or any other sign of inflammation. He had Grade II pustular acne on his upper back. Investigations done previously showed no biochemical or hematological abnormality. The ESR was normal. Examination of the synovial fluid was normal.

Histopathology of the meniscus and synovium showed non-specific inflammation with focal synovitis. X-rays of the knees were normal. MRI of the left knee showed findings suggestive of extensive synovial inflammation associated with moderate effusion. No erosions were seen and the subchondral bone was otherwise normal.

A diagnosis of SAPHO syndrome was made and after a 7-day flush out period the patient was given injection pamidronate 60 mg intravenously in 500 ml normal saline to run in 1 hour. Within 7 days his swelling was noticeably reduced and the pain subsided. The pustular acne cleared up, leaving post-inflammatory marks. In the 4<sup>th</sup> week the effusion began to again build up in the right knee. On day 30 after the initial infusion he was administered 30 mg pamidronate. Three days after the infusion he had pain in the right knee, and was treated with oral ibuprofen for 2 days. His pain subsided along with partial reduction of the effusion. During the entire treatment he had only one transient acne papule on his back that did not become pustular and subsided in a day's time. We initiated treatment with azithromycin 250 mg daily but the patient was then lost for follow-up.

## DISCUSSION

As described by Kahn *et al*, three diagnostic criteria characterize SAPHO syndrome.<sup>[1]</sup>

1. Multifocal osteitis with / without skin lesions,
2. Sterile acute or chronic joint inflammation associated with skin lesions, and
3. Sterile osteitis with skin lesions

According to Kahn, any one of the criteria is sufficient for a diagnosis of SAPHO syndrome. The skin lesions associated with these criteria are of the pustular kind either

1. psoriasis (pustular psoriasis, palmo-plantar pustulosis), or
2. acne (acne conglobata, acne fulminans or follicular occlusion triad).<sup>[1]</sup> Other dermatological conditions associated with the SAPHO syndrome are pyoderma gangrenosum,<sup>[2]</sup> Sweet's syndrome<sup>[3]</sup> and Behçet's syndrome.<sup>[4]</sup> Pustular acne is rare.<sup>[5]</sup>

The psoriatic variety is HLA-B27 negative and the skeletal component involves the front of the chest.<sup>[6]</sup> The most sensitive diagnostic test is a bone scintigraphy using Tc99; a bone or synovial biopsy will clinch the diagnosis. The disease course is usually chronic. Some patients do recover while in others it waxes and wanes.

The etiology is unknown. *Propionibacterium acnes* has been isolated from the bone lesions of patients with the SAPHO syndrome, and it is postulated that these bacteria may act like an antigenic trigger causing the bone and arthritic components of this condition.<sup>[7]</sup>

NSAIDs and sulfasalazine used to be the cornerstone of therapy; this treatment was only symptomatic. Now newer drugs can modify the course of the disease. With infection being considered as the key factor, antibiotics were tried with no response. Macrolides, especially azithromycin, has been effective in some cases, it is postulated that this works by an anti-inflammatory and immunomodulating effect.<sup>[8]</sup> In this patient we had added azithromycin to try to achieve a better control of the disease. Schilling also advocates this dual approach, and the need for a multicenter study to assess this treatment in SAPHO syndrome.<sup>[5]</sup>

Pamidronate is a calcium regulating agent which inhibits the action of osteoclastic cells responsible for bone resorption. Pamidronate has now come to the forefront of treatment. It acts because of its osteoclastic effect but seems to have anti-inflammatory properties too. Both increased bone modeling and inflammatory osteitis characterize SAPHO syndrome, indicating that it may respond favorably to pamidronate's dual mechanism of action.<sup>[9]</sup> Pamidronate seems to be a very effective mode of therapy for patients with the SAPHO syndrome by promoting remission in all components of the disorder, such as bone, joint and skin involvement.<sup>[10]</sup>

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