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PDF issue: 2026-05-11

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(Citation)
Bulletin of allied medical sciences Kobe : BAMS (Kobe),13:193-197

(Issue Date)
1997-12-26

(Resource Type)
departmental bulletin paper

(Version)
Version of Record

(URL)
https://hdl.handle.net/20.500.14094/00188188

KOBE UNIVERSITY

# Chronic Osteomyelitis of the Proximal Femur in SAPHO Syndrome with Avascular Necrosis of the Femoral Head: A Case Report.

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SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) syndrome is relatively rare, that affects anterior chest wall and spine of the skeleton almost exclusively. A 53-year-old patient man presented with palmoplantar pustulosis and left hip pain. Radiographic and magnetic resonance images exhibited an osteolytic lesion demarcated by a thin sclerotic line in the proximal metaphysis of the left femur and avascular necrosis of bilateral femoral heads. Histological and microbiological examination demonstrated that the metaphyseal lesion was sterile chronic osteomyelitis. He was subsequently diagnosed with SAPHO syndrome. As he had a history of excessive ingestion of alcohol and administration of steroid for pustulosis, we speculate that avascular necrosis was not due to osteomyelitis but alcohol or steroid.

## Key Words
SAPHO syndrome,
Avascular necrosis,
Osteomyelitis.

## INTRODUCTION

A variety of similar rheumatic diseases such as psoriatic arthropathy, Reiter's syndrome and enteropathic spondylitis overlap each other to form spondyloarthritides. One of the clinical features of these conditions is possible association with skin manifestations. Another case of this association is palmoplantar pustulosis with sternocostoclavicular hyperostosis, that was first reported by Sasaki in 1967.[1)] Sonozaki et al. described clinical characteristics of the association of these bone and skin lesions on the basis of 53 cases and proposed this clinical entity as pustulotic arthro-osteitis (PAO) in 1981.[2)] They also reported the incidence of 10% of arthro-osteitis in patients with pustulosis.[3)] In recent years, a French group has proposed the acronym SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) syndrome to define a disease entity, including a wider disease spectrum than PAO, with specific inflammatory bone lesions and skin manifesta tions.[4,5)] The pathology of this entity was originally characterized by sterile bone marrow inflammation and neutrophilic skin eruptions.

We report on an unusual case with SAPHO syndrome, who presented

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with chronic osteomyelitis of the proximal femur concomitant with avascular necrosis of the bilateral femoral heads. The diagnosis of SAPHO syndrome and the association of avascular necrosis with osteomyelitis in the present patient are also discussed.

## CASE REPORT

A 53-year-old man presented in December 1996 with slowly progressive pain in the left groin for a month and palmoplantar pustulosis for 3 years (Fig. 1). There was no swelling or pain in the anterior chest or back. At the presentation, dexamethazone (1.5 mg/day) had been prescribed for pustulosis at another hospital. The history of steroid was 2.5 years long with a maximum dose of 3 mg/day of the same steroid. He also had a history of excessive ingestion of alcohol from 20 to 45 years old, that was estimated to be 500 ml of alcohol a week.

Plain radiographs of hip joints demonstrated collapse of the left femoral head and a osteolytic lesion demarcated by a thin sclerotic line adjacent to the lesser trochanter of the ipsilateral femur (Fig. 2a, b). There were no radiographically abnormal signs in the right hip. Magnetic resonance imaging (MRI) depicted a lesion with low-signal-intensity within the left femoral head and a band pattern within the right head on T1-weighted images (Fig. 3a), suggesting that the right was on stage 1 and the left was on stage 2 of avascular necrosis of the femoral heads. The

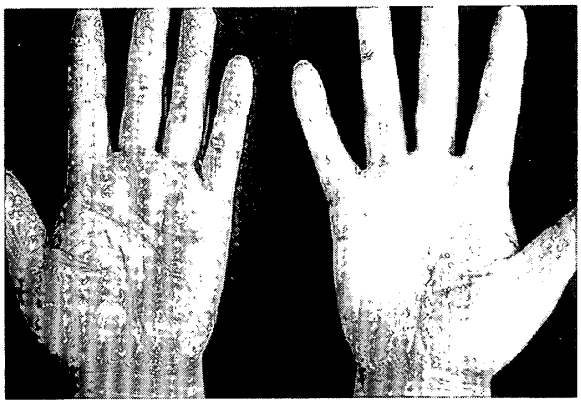

Fig.1. Pustulosis on the palms.

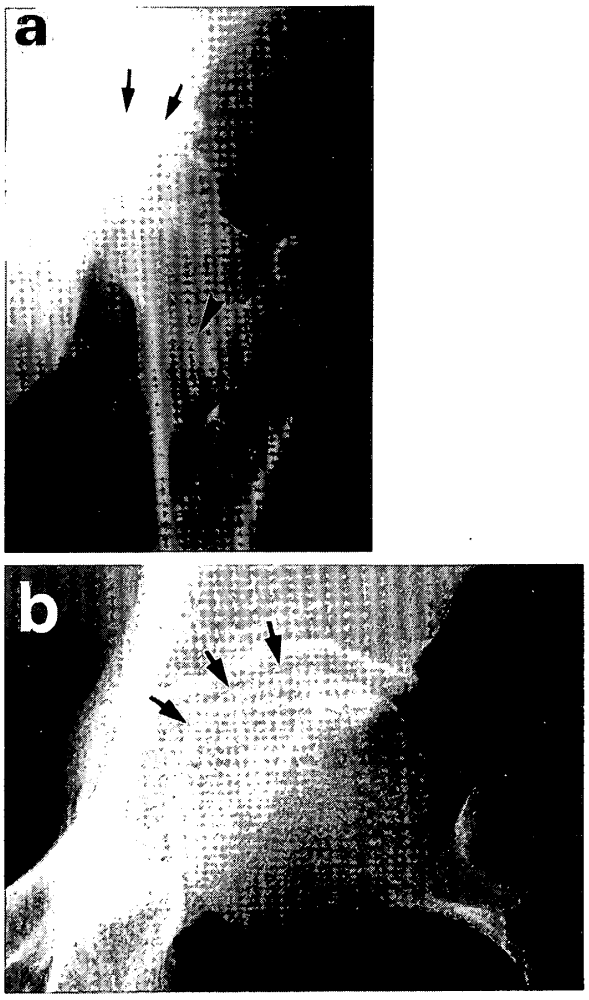


Fig.2. (a) An antero-posterio radiograph of the left hip joint. There were collapse of the weight bearing area of the femoral head (arrow) and a osteolytic lesion (arrowhead) with thin osteosclerotic rim adjacent to the lesser trochanter. (b) A lateral radiograph of the left hip. Note collapse of the head (arrow) seen more apparent to b.

osteolytic lesion was shown as a bone marrow lesion that had low intensity signal on T1-weighted (Fig. 3a) and high intensity signal on T2-weighted MR images (Fig. 3b). Administration of gadolinium enhanced the edge of the trochanter lesion (Fig. 3c). Laborative studies revealed that the serum total protein was 7.6 mg/dl with an abnormal electrophoretic pattern of the serum protein. There was an increased level of gammaglobulin, that accounted for 25% of the total protein. Although the serum concentration of IgG was within normal limits, those of IgA and IgM were elevated to 703 and 273 mg/dl, respectively. An erythrocyte sedimentation rate was slightly elevated to 21 mm/h. A white blood cell (WBC) count, values of hemoglobin and C-reactive protein (CRP) were within normal limits and rheumatoid factor was not detected.

To make a diagnosis of the metaphyseal lesion, open biopsy and iliac bone grafting were performed on February 1997. Histological examination revealed that bone marrow had been replaced by fibrous granulation with moderate degree of mononuclear cell infiltration, identical to that seen in chronic non-specific osteomyelitis (Fig. 4). Bacterial cultures of the bone specimens were sterile. The histology of osteomyelitis together with pustulosis confirmed the diagnosis of SAPHO syndrome.

A month later, anterior rotational osteotomy of the left femoral head was carried out to prevent further deformity of the head. The site of osteotomy was stabilized by a compression hip screw and two cannulated cancellous screws. We are

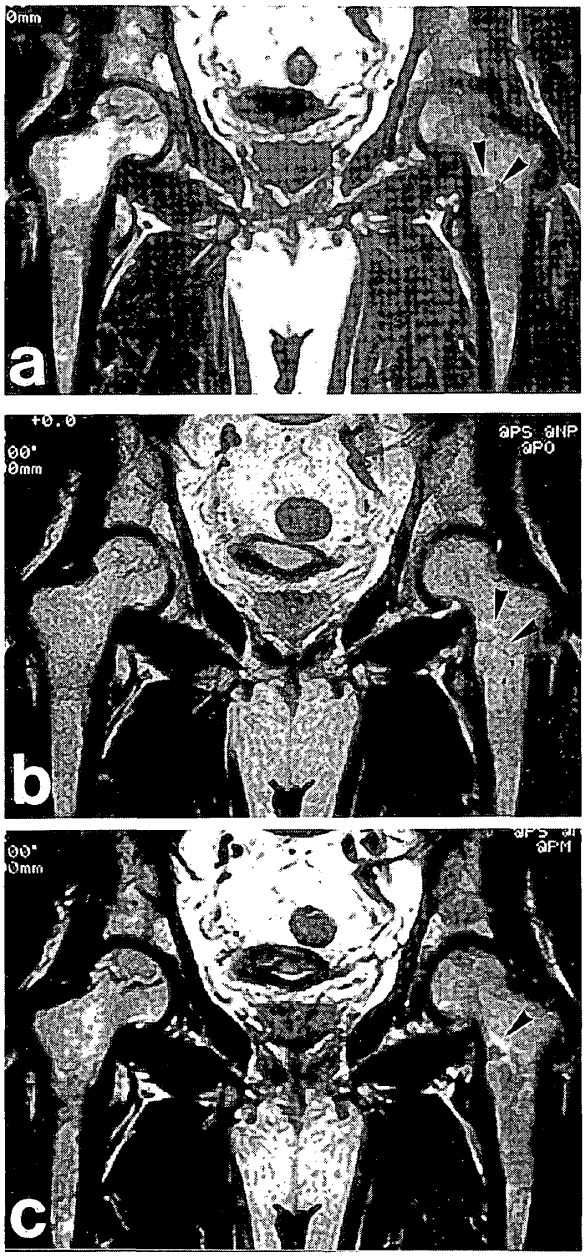


Fig.3. (a) A coronal T1-weighted MR image. A low-signal-intensity lesion in the left femoral head and a band pattern in the right femoral head were recognized. The radiolucent lesion in the metaphysis shown in Fig.2a had low-signal-intensity (arrowhead). (b) A coronal T2-weighted MR image. The lesion in the metaphysis was depicted to have high-signal-intensity (arrowhead). (c) A gadolinium-enhanced MR image. The edge of the metaphyseal lesion was enhanced (arrowhead).

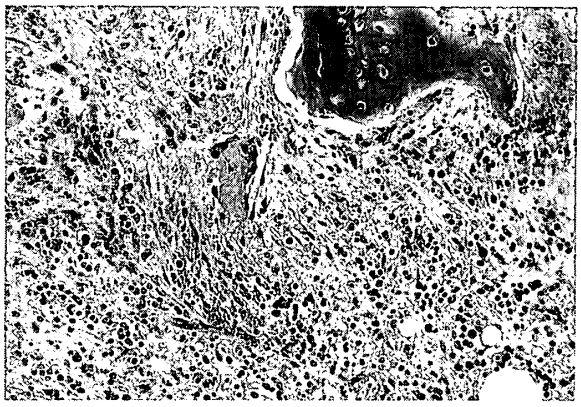

Fig.4. A microscopic appearance of the osteolytic lesion in the metaphysis. Note that normal bone marrow is replaced by fibrous granulation tissue with moderate infiltration by mononuclear inflammatory cells, identical to that seen in chronic bacterial osteomyelitis. (original magnification: x100).

carefully observing the conditions of the osteotomy site and the bilateral heads.

## DISCUSSION

The clinical manifestations of our patient met the criteria for SAPHO syndrome in the presence of pustulosis and sterile osteomyelitis.[4,5] However, the patient's manifestation were not typical of those of SAPHO syndrome in view of the following 2 points. 1) The patient lacked involvement of anterior chest wall or spine, that is present in almost all patients with SAPHO syndrome.[2,4,6] Cotten et al. described that about 30% of patients had involvement of long bones, of which the proximal femur was a rare occasion.[7] Maugars et al. showed that about a half of patients acquired new sites of bone involvement at a mean follow-up of 12.3 years, indicating that osseous lesions are multifocal.[6] The present patient may, thus, have new sites of bone involvement in anterior chest or spine in future.[2] Osteomyelitis of the patient was depicted as a pure osteolytic lesion on radiographs at presentation. Hyperostosis (as included in the acronym SAPHO) with some degree of osteolysis is a common radiographic finding.[2,4,6-8] However, early radiographs can demonstrate only erosive changes followed by hyperostosis or synostosis, as described previously.[7] It is, thus, noteworthy that radiological findings are dependent on the duration of the disease, as is well established in bacterial osteomyelitis.

It was unclear whether avascular necrosis had been linked to osteomyelitis in the metaphysis in a view point of the close proximity of the two lesions. There have been no reports on the association of avascular necrosis with SAPHO syndrome. We speculate that avascular necrosis was not secondary to osteomyelitis from the following reasons. The first is that he had two major risk factors for avascular necrosis. Three mg/day of dexamethazone for pustulosis and 500 ml/week of alcohol could independently induce this deleterious hip disease. As he stopped excessive ingestion of alcohol 8 years before the presentation, necrosis may have been induced not by alcohol but by steroid. The second is that the unilateral involvement of osteomyelitis in the metaphysis can not account for the bilateral involvement of avascular necrosis.

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