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SAPHO Syndrome with a Tumour-like Bony Proliferative Lesion in Distal Femur. A Case Report

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ABSTRACT

SAPHO syndrome is a group of bone and joint abnormalities associated with skin lesions. A 29-year-old male presented with severe acne on his trunk and anterior chest wall, right knee and foot pain. Radiographs and magnetic resonance images showed hyperostosis in the sternocostoclavicular region, sclerosis of one-third of the right distal 5th metatarsal bone and bony outgrowth from the medial condyle of the right femur .The histological findings of the biopsy specimen were consistent with those of old osteomyelitis. All fungal and microbacterial cultures were negative. Pain and swelling of the right knee and foot repeated remission and aggravation. There were no radiological changes of the above-mentioned lesions noted within 4-years follow-up.

INTRODUCTION

In 1987, Chamot *et al.* proposed the term, SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) syndrome, to designate collectively bone and joint abnormalities associated with skin lesions [1, 2]. The common site of skeletal lesions in SAPHO is the chest wall (clavicle, sternum, and sternoclavicular joints). Long bones are occasionally involved. Radiographic changes of the long bone lesions generally suggest either infectious osteomyelitis or a neoplasm such as Ewing's sarcoma[3]. We report a case of SAPHO syndrome having a cauliflower-like bony mass in the left femur together with an anterior chest lesion and sclerosis of the left 5th metatarsal bone. To the best of our knowledge, no cauliflower-like bone protrusion as in the present case has been reported.

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A 29-year-old male had severe acne on his trunk in the summer, in 1991. He had anterior chest pain in October 1999. Plain radiographs demonstrated hyperostosis of the sternoclavicular joints. The pain was relieved by non-steroidal anti-inflammatory drugs (NSAIDs). In 2001, he noticed pain in the right knee and foot, and visited another clinic. He was referred to us after plain radiographs had demonstrated bone tumors in his right femur and right 5th metatarsal bone.

On examination, acne conglobata was found on his head and many acne scars, on his trunk. A bony mass was palpated on the posterior aspect of the medial condyle of the right femur. The range of motion of the right knee was slightly restricted. A mild swelling was seen on the lateral side of the right foot. No redness, tenderness or local heat was found. Laboratory examination revealed a slight elevation of ASO titer (272 Todd). White blood cell count (8,900/ml) and C-reactive protein level (0mg/dl) were normal. Fungal and microbacterial cultures of throat swabs were negative. Plain radiographs showed a mixture of sclerotic and erosive lesions in the sternum including the sternoclavicular and 1st sternocostal joints. The distal metaphysis of the right 5th metatarsal bone was slightly enlarged with dens sclerosis. A tumor-like bony lesion was seen on the medial condyle of the right knee (Fig 1). All lesions were clearly visualized by computed

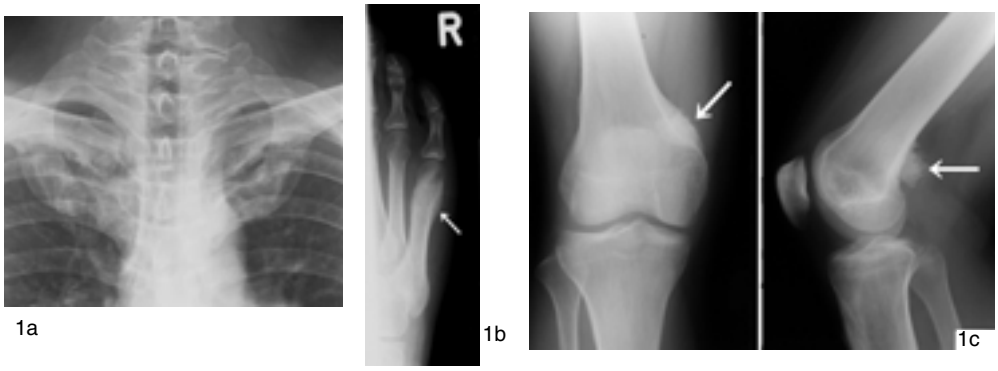
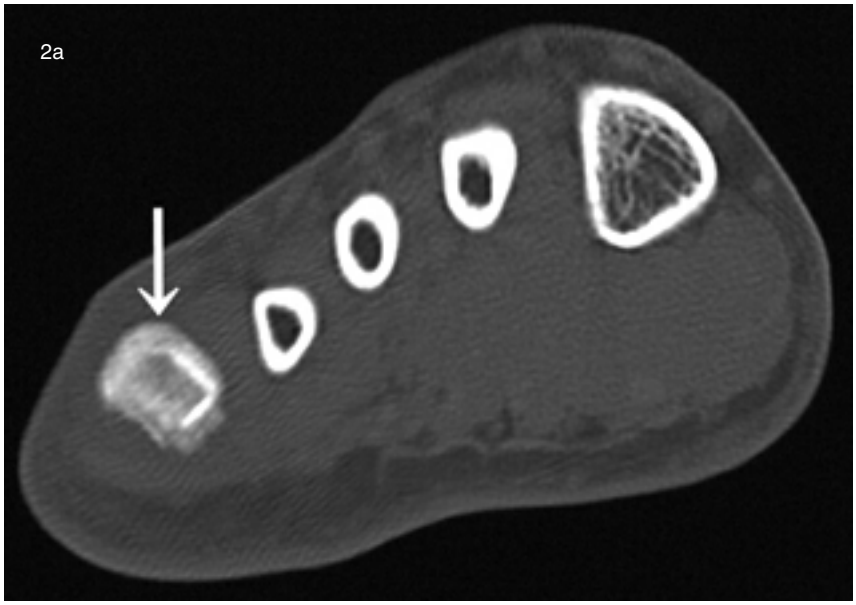


Fig 1. Plain X-ray examination revealed mixture of hyperostosis of sternocostoclavicular region (a), sclerosis (arrow) of the right distal 5th metatarsal bone (b) and tumor-like bony proliferative lesion (arrow) (c) in his right knee medial condyle.

tomography. The lesion of the right femur was in continuity with the underlying cortex. It had irregular surface and hypertrophied lamellar structures inside (Fig 2). Magnetic resonance imaging demonstrated low signal intensity on T1 weighted image, heterogeneous intensity on T2 weighted image in the tumor-like lesion. This lesion and the surrounding soft tissues were enhanced after administration of paramagnetic contrast medium (Fig 3). Bone scintigram showed increased uptakes in the anterior chest wall, right distal femur, and right distal 5th metatarsal bone.

Bone biopsies of the lesions at the right femur and 5th metatarsal bone were performed. All specimens were negative for neoplastic disease. Histological examination of



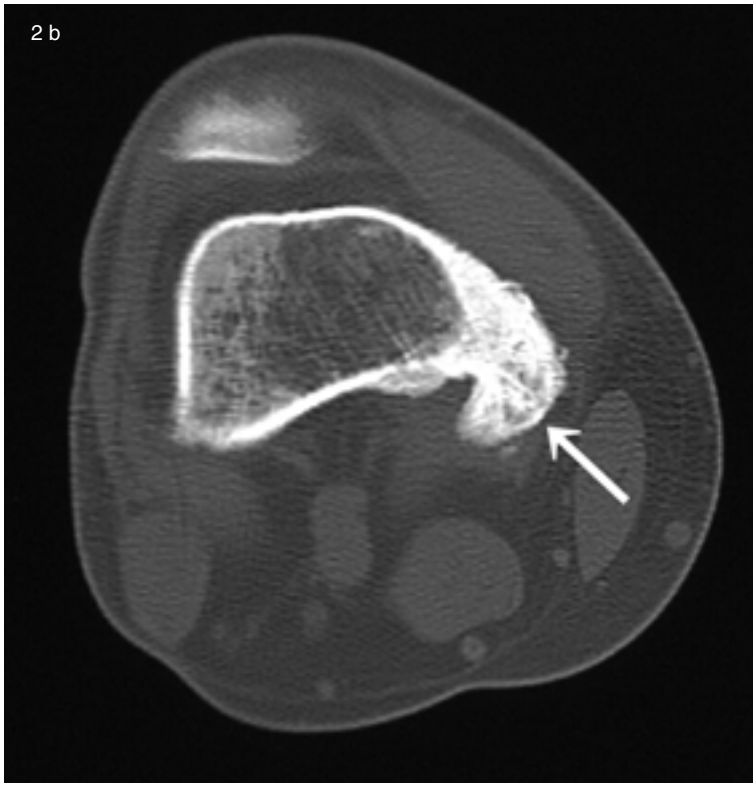


Fig 2. The lesions (arrow) (a: metatarsal lesion, b: distal femoral lesion) were clearly visualized by computed tomography. The tumor of the right femur was in continuity with the underlying cortex. It had irregular surface and hypertrophied lamellar structures inside.

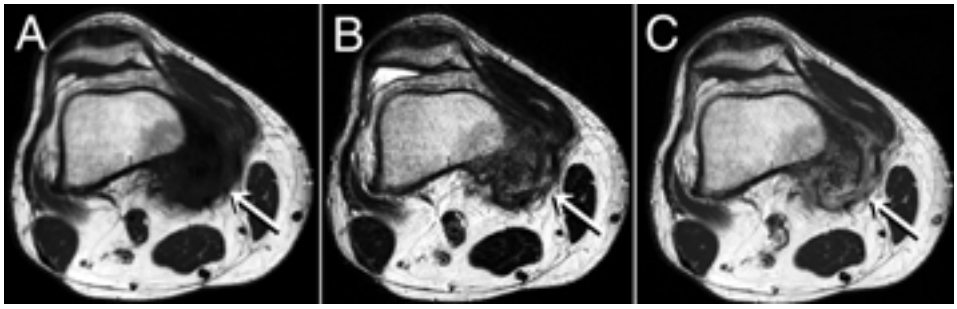


Fig 3. Magnetic resonance imaging demonstrated a femoral lesion (arrow) with low signal intensity on T1 weighted image (A), heterogeneous intensity on T2 weighted image (B) in the tumor. The tumor and the surrounding soft tissues were enhanced after administration of paramagnetic contrast medium (C).

the specimens demonstrated thickened sclerotic trabeculae and marked bone marrow fibrosis with some chronic inflammatory cells (Fig 4). Fungal and microbacterial cultures were negative.

The patient was diagnosed having SAPHO syndrome because of the presence of polyostotic osteomyelitis or bone tumor like lesions, acne, and the histolo1ogical features of the biopsy specimen.

The pain in the right knee and right foot disappeared soon after biopsy. However, skin lesions repeated remission and aggravation. The pain and swelling in the right foot recurred after eight months and disappeared two months later. No progression of the bone lesions was found on plain radiograms within 4-years follow-up.

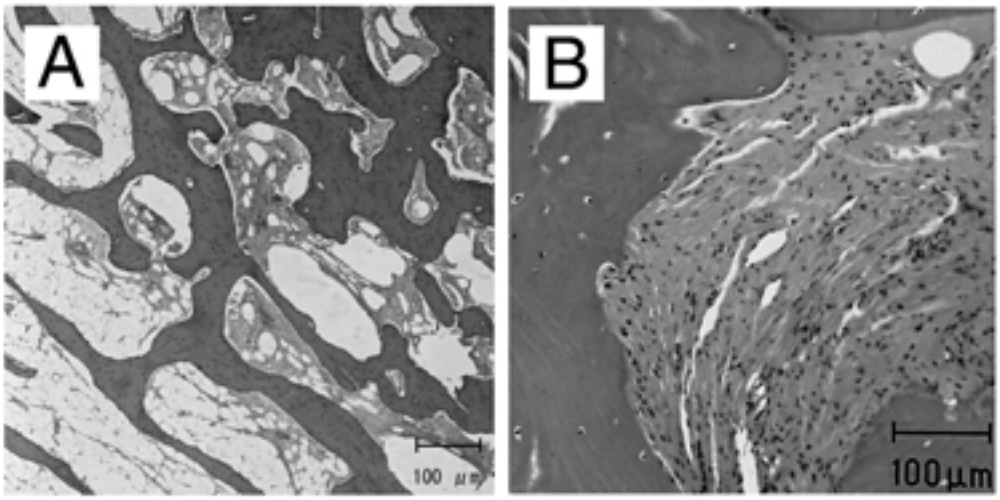


Fig 4. Histological examination of the biopsy specimen demonstrated chronic inflammatory changes, with sclerotic bone trabeculae, prominent bone marrow fibrosis, and scattered lymphocytes. The histological diagnosis was old osteomyelitis. (A: low power view x10, B: high power view x 40)

DISCUSSION

In 1961, Windom *et al*. were the first to describe musculoskeletal problems associated with acne conglobata [4]. In 1987, Chamot *et al*. first proposed an acronym SAPHO syndrome, to describe a group of bone and joint abnormalities associated with skin lesions [1]. Prior to the creation of this acronym, some 250 cases of hyperostosis of the chest wall with pustular eruption in hands or soles had been reported with over 50 designations [5]. Most cases have been reported from Japan and Northern and Western Europe, with a few cases described from United States, Canada, and Great Britain[3]. In Japan, SAPHO syndrome has been paid little attention, probably due to that bone lesions are usually localized to the anterior chest wall and have diagnosed as sternocostoclavicular hyperostosis without checking skin lesions.

Mixture of bone sclerosis and atrophy suggestive of osteomyelitis is one of the bony features of SAPHO syndrome. Hypertrophic bone lesions are often seen in the anterior chest wall. When the anterior chest is involved, adjacent ligaments are also involved by the ossifying process, with production of a sclerotic enthesopathy[3]. Radiographic changes of the long bone lesions usually suggest either infectious osteomyelitis or a primary bone neoplastic lesion, mainly Ewing's sarcoma [3]. In the present case, the location of the femoral lesion well coincided with the attachment of the medial head of the gastrocnemius muscle. The traction of this muscle is thought to be one factor, causing a cauliflower-like bone protrusion. The patients with skin trouble like acne, when having tumor or osteomyelitis like bony lesions should be suspected to have this syndrome.

In 1999, Reith reported that histological findings vary depending on the stages of the disease process. Early lesions contain acute inflammation, edema, and prominent periosteal bone formation, whereas late lesions demonstrate markedly sclerotic bone trabeculae with prominent marrow fibrosis and only mild chronic inflammation [6]. In the present case, four months' duration of symptoms and the histological findings are well consistent with late lesions.

Treatment for SAPHO syndrome has not been standardized yet. NSAIDs are commonly used. However, their efficiency is neither consistent nor complete [2]. Corticosteroids or methotrexate have proven useful in most severe forms, but the efficacy has been variable [7]. In Japan, tonsillectomy has been performed in order to alleviate clinical symptoms associated with the SAPHO syndrome, as the manifestation may be regarded as diseases of focus tonsillitis. However, tonsillectomy was not found to bring any improvements to deformed joints or to reduce intractable pain of sternoclavicular hyperostosis[8]. For the present case, NSAIDs was effective with temporally relief of pain.

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