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SAPHO 증후군
- 증례보고 -

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서무삼 · 박형택 · 박종훈

수장족저 농포증은 특별한 골, 관절계 질환과 관련이 있을 수 있다. 초기 전흉벽에 국한되었던 이러한 질환은 이후 말초 골, 관절계를 침범하는 것으로 밝혀졌다. 무균성 가감염성 골, 관절 질환 역시 심한 여드름과 관련이 있는 것으로 밝혀졌다. 1987년 Kahn 등은 과거 25년간 발표된 특별한 골, 관절 그리고 피부병변에 관한 수많은 보고들을 기술하기 위하여 약성어를 제시하였다. SAPHO 증후군은 Synovitis, Acne, Pustulosis, Hyperostosis and Osteitis syndrome을 뜻한다. 저자들은 SAPHO 증후군으로 의심되는 과골증을 동반한 좌측 쇄골 및 우측 천장 관절 동통과 수장 농포증이 있는 27세 여자 환자 1예를 경험하였기에 보고하고자 한다.

색인 단어: 쇄골, 천장 관절, 수장족저 농포증, SAPHO증후군

INTRODUCTION

The fundamental component of SAPHO syndrome is inflammatory, pseudoinfectious, usually sterile osteitis. The best known and most often described osteitic losions involve the bone and joint of the anterior chest wall. In addition to the anterior chest wall, the pelvic girdle, peripheral bones, and spine are often affected and can be involved in the abscence of sternoclavicular and sternocostal lesions[6]. The associated skin condition mentioned most frequently is palmoplantar pustulosis[6] and various patterns of psoriasis and severe acne can be associated with the characteristic bone lesions[7].

CASE REPORT

The patient was a 27 year old woman who came to our clinic with pain and mild swelling on left clavicular region and right sacroiliac joint. The patient had no history of psoriasis, nor was there any history or clinical evidence of psorisis in her family.

Physical examination of the affected area showed diffuse mild swelling and severe ten-

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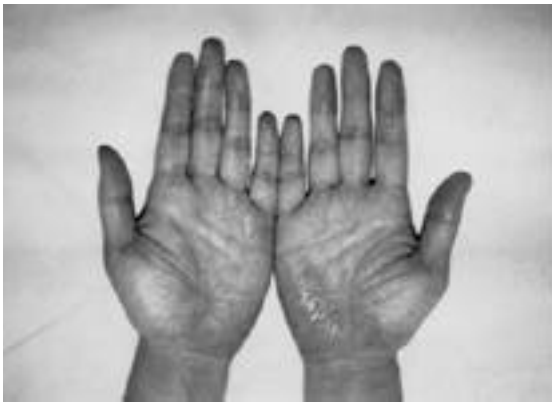

Fig. 1. Pustular skin lesions are shown on the palm.

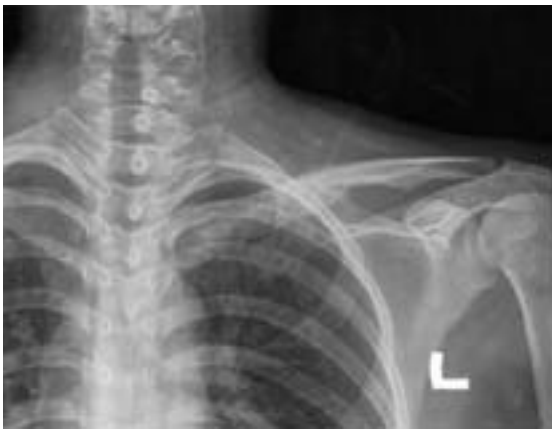

Fig. 2. Linear radiolucent line with periosteal callus formation and osteoporotic change are noted on the left clavicle.

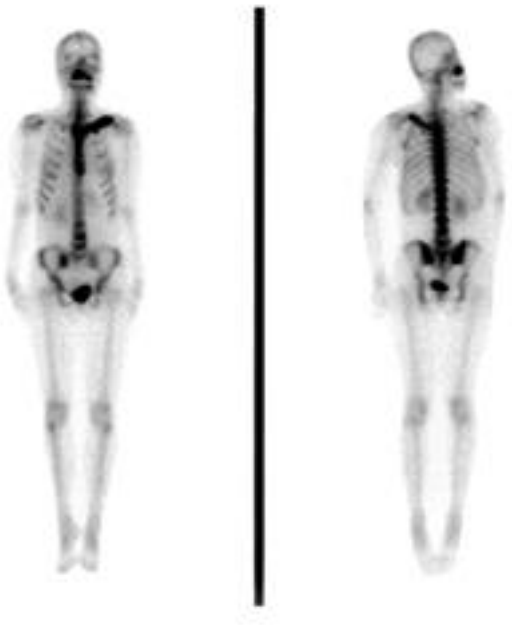

Fig. 3. Radionuclide bone scan imaging with 99 m Tc-methylene diphosphonate shows increased uptake on left clavicle and right sacroiliac joint.

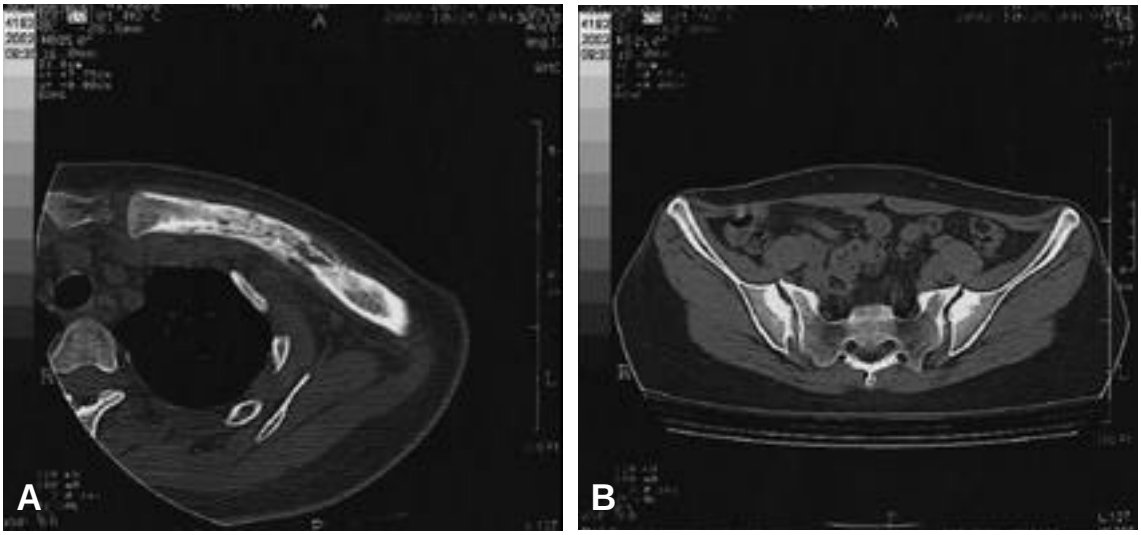

Fig. 4. A. CT scanning of clavicle showed enlargement of the midshaft of clavicle with osteolytic and sclerotic areas. **B**. CT scanning of pelvis showed sclerotic area in right sacroiliac joint.

derness of left clavicular region and right sacroiliac joint. However, the range of shoulder joint motion was relatively well maintained.

Pustules appeared on the patient's palms and soles four months ago, and she was diagnosed by a dermatologist as having pustulosis palmoplantaris (Fig. 1). She was treated with steroid ointment.

A plain radiograph showed linear radiolucent line with periosteal callus formation and osteolytic change in left clavicle (Fig. 2).

A technetium-99 m diphosphonate labelled scintigram of the bones showed increased uptake of the isotope in the left sternoclavicular region and right sacroiliac joint(Fig. 3).

CT scanning of clavicle and pelvis showed enlargement of the midshaft of clavicle with osteolytic and sclerotic areas(Fig. 4).

Hematological tests gave the following results: ESR 54 mm/h WBC 12,700/mm^3. Hemoglobin 11.5 g/al, plateletes 604,000/mm^3, creatinine 0.7 mg/dl, BUN 10.3 mg/dl, calcium 10.1 mg/dl, phosphorus 3.6 mg/dl, SGOT 28 IU/L, SGPT 39 IU/L, urine analysis was normal.

Biopsy of left clavicle was done. Histologi-cal findings showed new bone formation with increased osteoblastic activity (Fig. 5).

General steroid treatment was used. After 3 months later, most of the symptoms were subsided and radiologic finding showed markedly improvement (Fig. 6).

DISCUSSION

In 1987, Kahn et al[12] coined the term SAPHO syndrome (Synovitis, Acne, Pustulosis, Hyperostosis, Osteitis) to designate a group of frequently combined manifestations, of which the most common is osteitis of the anterior chest wall. Over the last 10 years, almost 100 papers, mainly from Japan, Scandinavia, Germany, and France, have been published on similar cases but few reports from Korea. SAPHO syndrome is observed mainly in children and in young and middle-aged adults[8]. It is rarely seen after 60 years of age[2], and more frequent in female[5].

The bone and joint involvement is the common denominator in patients. The anterior chest wall is the most frequent localization[2]. In most cases, spine lesions are seg-

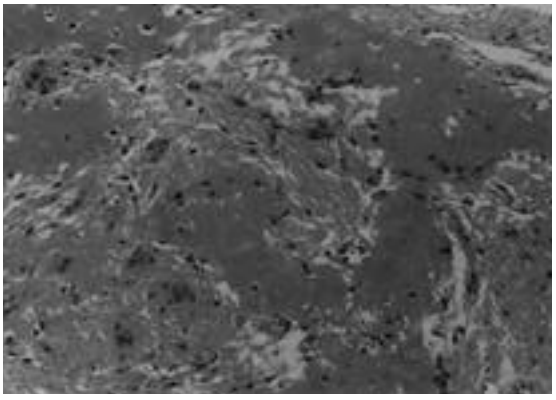

Fig. 5. Histological finding of biopsy of left clavicle shows mature lamella bone with new bone formation and increased osteoblastic activity(H-E stain×200).

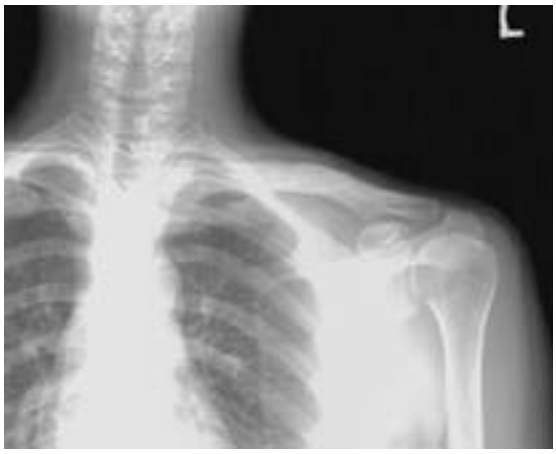

Fig. 6. Radiologic finding of left clavicle showed markedly improvement.

mental, involving two to four adjacent vertebrae. The thoracic vertebrae are the most frequently involved. However, the cervical, lumbar, and even sacral spine may show lesions. In some cases, widespread involvement occurs, usually in combination with sacroiliitis, raising the issue of a possible link with ankylosing spondylitis. Sonography, computed tomographic scan[3], and magnetic resonance contribute little to the identification and localization of lesions. On the other hand, technetium bone scan may prove useful because isotope uptake is increased markedly in all of the above abnormalities and because bone scan is able to detect early involvement including radiographically negative foci. Many publications initiating from the paper by Sonozaki et al[8] suggest that sacroiliitis may be observed in a significant proportion of patients.

Skin lesions consist in palmoplantar pustulosis, palmoplantar pustular psoriasis, other pustular psoriasis, severe acnes, acne conglobata, hidradenitis suppurativa and dissecting cellulitis of the scalp[4].

There is no specific marker for the SAPHO syndrome. Usually, a moderate inflammatory reaction is found with elevated erythrocyte sedimentation rate and C-reactive protein. Alpha-2 and gammaglobulins are elevated slightly in some instances. Blood counts are usually normal. All the markers for different infectious diseases are normal.

Presence of HLA-B27 was correlated with involvement of the anterior chest wall (41.7%) but was much less frequent in patients with palmoplantar pustulosis (12.8%) and with acne (9%) in Khan et al's paper[2].

The main diagnostic problem is, of course, infectious bacterial osteomyelitis. In the past, repeated biopsies with tissue cultures were performed in these patients, and even when results were negative, antibiotics were tried frequently. In almost every case, they proved ineffective. Spinal involvement suggests infectious spondylodisciitis. This leads to invasive, potentially harmful procedures. In cases with prominent peripheral involvement, tumoral conditions are also discussed. Ewing's sarcoma is the more frequently considered, mainly in monostotic cases. Lately, Paget's disease may be considered in a patient with a hypertrophic and sclerotic bone lesion. Pathologic results may be confusing, because an increase in both osteoclasts and osteoblasts can be seen in involved bone[1]. However the patient's age and a careful radiologic and pathologic work-up provide helpful data.

In treatment of SAPHO syndorme, anti-infectious treatment have proved ineffective in most reports in which they were tried[2]. Nonsteroidal anti-inflammatory drugs have been the most frequently used treatments. No single drug has been found to be more superior to the others. Duration of treatment cannot be standardized. Corticosteroids can be useful in the most severe forms, some found them effectiveness, whereas others have negative results.

This paper reports a patient with left clavicular and right sacroiliac joint pain and hyperostosis, pustulosis palmoplantaris. Typical features of SAPHO syndrome were present in our case. In laboratory studies, ESR and WBC were mildly elevated. The arthritis was not transient, but persisted for several months with little remission, failing to NSAIDs. We started steroid treatment, the patient's symptom was relieved. The recognition of SAPHO syndrome can protect individuals from ill-indicated therapeutic

procedures, such as long-term antibiotics, radiotherapy, or surgery wrongly prescribed to treat an alleged infectious or tumorous condition, mimicked by the bone changes. Lastly, patients must be carefully informed of the usually benign if painful course of their condition. This knowledge, in our experience, frequently helps the patient endure the symptoms.

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Abstract

SAPHO Syndrome
- A Case Report -

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Palmoplantar pustulosis may be associated with peculiar bone and joint conditions. Initially restricted to the anterior chest wall, these conditions were later found to involve peripheral bones and joints. The same aseptic pseudoinfectious bone and joint lesions were also found associated with severe acne. In 1987, Kahn et al suggested this acronym to describe a number of reports of peculiar bone, joint, and skin lesions published over the last 25 years. SAPHO sydrome stands for synovitis, acne, pustulosis, hyperostosis, and osteitis syndrome. We report one case of SAPHO syndrome: A 27-year-old female presented left clavicular and right sacroiliac joint pain with hyperostosis and pustulosis palmaris.

Key Words: Clavicle, Sacroiliac joint, Palmoplantar pustulosis, SAPHO syndrome

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