SAPHO Syndrome is a rare disease, and may lead to misdiagnosis of infectious osteomyelitis, resulting in long-term treatment with antibiotics, or osteoblastic metastasis when the patient have primary tumor history.

We report a case of SAPHO syndrome who had a primary cancer history. The patient was a 42-year-old patient suffered from left breast cancer and received operation in 1997.

Bone scintigraphy showed uptake at right sternoclavicular junction. She started to complained about anterior chest wall pain one year later. Series of bone scintigraphy from 1997 to 2003 showed progressive uptake at right sternoclavicular junction, spine, sacroiliac joint and the left sternoclavicular junction. The pain persisted and progressed, and pustular skin lesions over palms and soles also developed in 2003.

CT scan and MRI study revealed multifocal peculiar hyperostotic bone lesions involving bilateral sternoclavicular junction, lumbar vertebra, and right sacroiliac joint, with mild degree surrounding marrow edema around the lesions.

Base on her clinical history, serial chest films, hyperostosis, and pustular skin lesions, possibility of bone metastasis was excluded and a diagnosis of SAPHO syndrome was made.

Key words: SAPHO syndrome, Hyperostosis, Sternoclavicular joint, Acquired Hyperostosis Syndrome

In 1987 a variable group of conditions characterized by the presence of synovitis, acne, pustulosis, hyperostosis, and osteitis was termed the SAPHO syndrome by Chamot et al [1]. It has symptom for the anterior chest wall, characterized by sclerosis, hyperostosis, and arthritis of the sternoclavicular joints and costochondral junctions. It may also affect the spine and sacroiliac joints. The bone lesion may mimic malignancy or infection radiologically. We describe a case of SAPHO syndrome who was a 40-year-old female suffered from breast cancer with the features initially suspected to be multiple bone metastasis.

CASE REPORT

The female had left breast cancer and received modified radical mastectomy and axillary lymph node dissection in 1997. Preoperative technetium 99m bone scintigraphy (Fig. 1a) showed uptake at right sternoclavicular junction.

After one year, she started to complained about anterior chest wall pain. On presentation, the physical examination was normal. Follow-up bone scintigraphy showed uptake at right sternoclavicular junction without obvious interval change as compared to the study in 1997.

In 2002, she visited our outpatient department again due to right lower back pain. This time, the Technetium 99m bone scintigraphy (Fig. 1b) revealed new hot uptake at left sternoclavicular junction, L5 vertebral body, and right sacroiliac joint, and less uptake of right sternoclavicular junction. The new lesions at L5 and right sacroiliac joint were interpreted as traumatic change based on the history that the patient is a had pelvic trauma 2 years ago.

In 2003, the lower back pain persisted and progressed, and pustular skin lesions over palms and soles also developed.

The erythrocyte sedimentation rate was 0.809 mm/h (<0.8) and both the white blood cell count and hemoglobin were normal. Blood cultures were negative.

Radiographs revealed asymmetrical focal increased bone density of the right sternoclavicular junction, L5-S1, and around the right sacroiliac joint.
At the same time, technetium 99m bone scintigraphy (Fig. 1c) demonstrated persisted hot uptake at the left sternoclavicular junction and more extensive and intensive uptake at the right sacroiliac joint and the L5 vertebral body.

The female was initially thought to have osteoblastic metastasis and we arrange magnetic resonance (MR) imaging of lumbar spine and computed tomography (CT) scans of anterior upper chest wall and lumbar spine for her. The MR imaging study (Fig. 2) showed multiple osteoblastic lesions involving right sacral ala, subchondral area of right ilium, L5 vertebra and corners of T12, L1 and L2 vertebrae, with mild degree surrounding marrow and soft tissue edema around the lesions. CT of the anterior chest (Fig. 3) showed hyperostotic lesion in the cartilaginous portion of the anterior aspect of the right first rib and the bilateral sternocostoclavicular area with ankylosis of the right side.

Retrospective inspection of serial chest films from 1997 to 2002 disclosed progressive osteosclerosis of medial aspect of bilateral clavicles and upper sternum and progressive ossification of right sternocostoclavicular joint.

On the basis of her clinical history, serial chest films, hyperostosis, and pustular skin lesions, a diagnosis of SAPHO syndrome was made, instead of multiple osteoblastic metastasis.

She was treated with prednisolone. One month later, the female back pain had resolved. Her disease underwent a period of relative stability, with the symptoms controlled by simple analgesia.

**DISCUSSION**

The acronym SAPHO (for synovitis, acne, pustulosis, hyperostosis, and osteitis syndrome) was first proposed by French rheumatologists (Chamot and coworkers) in 1987 [1]. They discovered an association between several musculoskeletal disorders and various dermatologic conditions. Since their initial description, more than 50 different conditions have been linked to the SAPHO syndrome, and the most commonly mentioned were chronic recurrent multifocal osteomyelitis (CRMO), spondyloarthritis hyperostotica pustulopsoriatica (SHPP), pustulotic arthritides, plasma cell osteomyelitis and sternocostoclavicular hyperostosis (SCCH) [2].

Kahn and Chamot [3] noted that frequently affected bone was enlarged and sclerotic, with pseudopagetic changes, including areas of osteolysis and sclerosis. Long bones exhibited hyperostosis associated with pain, swelling, and increased local heat. The most common musculoskeletal complaint is at...
sternoclavicular junction of anterior chest wall. In cases with vertebral involvement, patients had back pain and stiffness. Lesions frequently involve two to four adjacent vertebrae and are frequently associated with sacroiliitis [3] and spondylodiskitis [4].

Bone scintigraphy shows increased isotope uptake in the anterior chest wall in 70-90% of cases [5].

There are no specific markers for these syndromes. Blood counts are usually normal. C-reactive protein and α1-globulin levels are occasionally elevated as well. The most consistent finding in blood and serum analysis is an elevated erythrocyte sedimentation rate. The elevation usually worsens as symptoms aggravate.

Skin involvement is not absolutely necessary for diagnosis of SAPHO syndrome, and the type of skin condition is variable. Pustulosis of the palms and soles, pustular psoriasis, acne conglobata or fulminans and hydradenitis suppurativa are associated. Skin lesions may be transient and may precede or follow musculoskeletal symptoms by as much as 20 years [6]. For most patients, the interval between onset of skin manifestation and osteoarticular lesions is less than 2 years, but intervals as long as 20 and 38 years have been reported [7].

The etiology and pathophysiology of SAPHO syndrome are unknown.

To further clarify the diagnosis of SAPHO syndrome [8], Kahn et al. established a diagnostic criteria in 1994, including (1) chronic recurrent multifocal osteomyelitis (CRMO), with or without spine or skin lesions, (2) acute, subacute, or chronic arthritis with any of palmoplantar pustulosis, pustular psoriasis, or severe acne, (3) any severe osteitis with any of skin manifestations.

Figure 3. CT of bilateral sternoclavicular junction (A and B), sacrum (C). There are Multifocal hyperostotic lesions involving right sternoclavicular joint, sternal part of left sternoclavicular junction (arrowhead), right sacral ala, subchondral area of right ilium, and L5-S1 vertebra (black arrow), with ankylosis of right sternoclavicular joint (arrow).
Some authors suggest that the SAPHO syndrome is an additional member of the family of seronegative spondyloarthropathies. Sacroilitis occurs in about 50% of patients with the SAPHO syndrome and may be indistinguishable from that seen in ankylosing spondylitis. Vertebral body involvement identical to that seen in ankylosing spondylitis also may occur in SAPHO patients. Other features common to both SAPHO patients and patients with seronegative spondyloarthritis include enthesopathies [9], inflammatory bowel disease [10], and psoriasis. The prevalence of HLA-B27 is higher in SAPHO patients (33%) than in the general people [1], its frequency is less than that seen in the seronegative spondyloarthropathies. Nonetheless, the increased prevalence of this antigen, combined with the striking clinical and radiologic overlap, has suggested to some authors that SAPHO is likely an additional member of this family of disorders.

It maybe difficult to separate SAPHO syndrome and psoriatic arthritis, when combined spondyloarthropathy and dermatologic conditions. However, Sternoclavicular joint involvement was rare in psoriatic arthritis [11]; and DIP joint involvement also rare in SAPHO syndrome.

As the etiology of SAPHO syndrome remains unclear, treatment has been difficult and largely ineffective [12]. Nonsteroidal antiinflammatory drugs have been effective in some cases, but their effect has been inconsistent and incomplete. This female had symptomatic relief by the use of nonsteroidal antiinflammatory drug and steroid.

The major diagnostic pitfall is the misdiagnosis of infectious osteomyelitis, resulting in long-term treatment with antibiotics, which are usually ineffective, and misdiagnosis of osteoblastic metastasis, when the patient have primary tumor history.

Lesions in typical sites are important for a correct diagnosis. Many patients with SAPHO syndrome have involvement of the anterior chest wall, typically the clavicle, the sternum, or one of the first two ribs, and adjacent joints such as the sternoclavicular or sternocostal joints. The spine is the second most common site of SAPHO syndrome in adults and is involved in approximately one third of patients [7]. Bone scan is able to detect early bone involvement, which would not yet be seen radiographically. Computed tomography, and magnetic resonance imaging also contribute to the identification and location of the lesion.

It is important to be aware of SAPHO syndrome to recognize the link between skin involvement and musculoskeletal abnormalities to avoid a misdiagnosis and to differentiate the condition from a malignant process, avoiding unnecessary investigations and inappropriate treatment.

**REFERENCE**

SAPHO syndrome is a rare disease, sometimes mistaken for infective osteitis, leading to long-term antibiotic treatment; or when the patient has a primary tumor history, it may be misdiagnosed as metastatic tumor.

We report a case of a 42-year-old woman with a history of breast cancer who developed SAPHO syndrome. The patient underwent surgery for left breast cancer in 1997. A year later, the patient began to complain of thoracic pain. After 1997 to 2003, a series of nuclear scans showed abnormal uptake in the right thoracic, lumbar, and sacroiliac joints, with progressive joint anomalies. The patient's pain gradually worsened.

We used computed tomography and magnetic resonance imaging to confirm the abnormal bone growth in the two thoracic joints, lumbar vertebrae, and right sacroiliac joints. SAPHO syndrome was diagnosed.

Keywords: SAPHO syndrome, bone proliferation, thoracic joint, reactive bone proliferation syndrome.

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