Imaging Manifestation of Synovitis, Acne, Pustulosis, Hyperostosis and Osteitis Syndrome

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Forty-three years old man with a background history of left radical nephrectomy for renal cell carcinoma (RCC) on surveillance was presented to the rheumatology clinic, Sultan Qaboos University Hospital, Muscat, Oman, in June 2019 with a history of severe pain at manubrium and right sternoclavicular joint associated with acne-like lesions in both palms and soles since two years. On examination, swelling, redness, and tenderness were present over the manubrium and right sternoclavicular joint. There are acne-like lesions in the back, soles and, palms. Laboratory investigations showed elevated inflammatory markers, including erythrocyte sedimentation rate and the total count of white blood cells and neutrophils. Computerized tomography of the chest (CT), bone scintigraphy with single-photon emission computed tomography/CT (SPECT/CT), and whole-body F-18 fluorodeoxyglucose-positron emission tomography/CT (FDG-PET/CT) was done and showed multiple bone lesions. A diagnosis of synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome was made. Non-steroidal anti-inflammatory drugs (NSAIDs), prednisolone, and methotrexate were administered, and the patient's condition improved dramatically. We demonstrate multiple imaging modalities which help to diagnose SAPHO syndrome along with provided history and medical background. Early and accurate diagnosis is essential to reach correct management and good outcome.
Comment

SAPHO is a rare syndrome that was first reported by Chamot et al. in 1987. Many cases have been reported in the West and Japan. The underlying pathophysiology is still unclear, although many hypotheses attributed this condition to a group of genetic, immunologic, and infectious factors.

SAPHO syndrome is a benign condition affecting mainly young and middle-aged adults. This condition could be managed conservatively. Early diagnosis and proper management will improve the clinical outcomes and prognosis.

Clinical history, along with dermatological and radiological manifestations, is essential to make the diagnosis of SAPHO syndrome. Any one of these criteria are considered adequate to diagnose SAPHO syndrome: 1) multifocal osteitis with or without skin lesions; 2) sterile acute, subacute or chronic arthritis associated with pustular psoriasis, palmoplantar pustulosis, acne or hidradenitis suppurativa; and 3) sterile osteitis combined with one of the skin manifestations. However, diagnosis of SAPHO syndrome sometimes could be challenging, especially if skin manifestations are absent. In our case, the patient presented with severe pain at the manubrium and right sternoclavicular joint associated with acne-like lesions in both palms and soles, which meet the criteria of diagnosis.

Different modality imaging like x-ray, CT, bone scintigraphy and magnetic resonance image (MRI) may help to diagnose SAPHO syndrome, but the whole body bone scintigraphy using $^{99m}$Tc-methylene-diphosphonate considers the most sensitive modality in detecting the early bony involvement when these changes in another modality are absent or subtle. Anterior chest wall bones involving both sternoclavicular junction and manubrium sterni consider the most common location for early bony changes of SAPHO syndrome followed by the spine and, to a lesser extent, sacroiliac joints. In our case, these locations were involved apart from the normal appearance of both sacroiliac joints, as shown in [Figures 1,2]. The radiological appearance of SAPHO syndrome depends on the stage of progression, which may start with osteolysis in the early stage, followed by osteitis, hyperostosis, and osteosclerosis. A bone scan usually gives a characteristic pattern of uptake called bull's head appearance centred at the
sternoclavicular joints and manubrium sterni [Figure 1A]. Although plain radiograph is non-specific, sometimes it may show changes if clinically suspected like osteitis and hyperostosis of sternoclavicular joint, sclerotic bony changes of vertebral bodies, and sacroiliitis.\(^3^\)\(^4^\) CT modality gives more details about osteoarticular erosive changes, particularly of the anterior chest wall.\(^4^\) MRI is advised in cases of the suspected active inflammatory process of spondylodiscitis. SAPHO spine lesions may appear as body corner erosions or erosive changes of the disc with narrowing of disc space.\(^2^\)\(^4^\) Our patient had a bone scan [Figure 1A] and CT scan [Figures 2A-C], which showed similar comparable changes to the above-mentioned findings of SAPHO syndrome. FDG PET/CT scan was done to exclude the possibility of sclerotic metastasis in the background of RCC and reported as negative (image not provided).

NSAIDs with or without antibiotics are considered the first-line therapy for SAPHO syndrome.\(^5^\) Many studies have proven the role and usefulness of bisphosphonates, steroids and biologic agents as treatment options, particularly for skin and articular manifestations.\(^2^\)\(^5^\) In our case, NSAIDs, prednisolone, and methotrexate were administered, and the patient condition improved with good outcomes on follow-up.

**Disclosure**
The reporting of the current case does not contravene with the regulations of the local institutional review board. No conflicts of interest declared concerning the publications.

**Authors Contribution**
AA collected the clinical data, medical images, made the literature review, wrote the manuscript draft and revised the manuscript. SR chose the appropriate images and reviewed the manuscript drafts before submissions with error correction.

**References**


Figure 1: (A) Anterior views of the whole body bone scan (WBS) provided with (B) reconstructed SPECT/CT images in sagittal and coronal views revealed intense uptake at the
proximal end of the right clavicle (red arrows) and manubrium sterni (yellow arrows). Faint radiotracer uptake were noted at left sternoclavicular joint and left first rib.

Figure 2: CT scans of the sternum and spine (A,B) revealed bony erosion and hyperostosis changes on the right sternoclavicular joint (red arrows) and sclerotic osteitis changes on either side of the sterno-manubrial joint (yellow arrows). Multiple scattered osteosclerotic lesions in the thoracic and lumbar spine with relatively normal disc spaces (B,C) (white arrows). FDG PET /CT showed no definite focal FDG uptake on these lesions and both sacroiliac joints are grossly normal (images not provided).