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INTERESTING MEDICAL IMAGE

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

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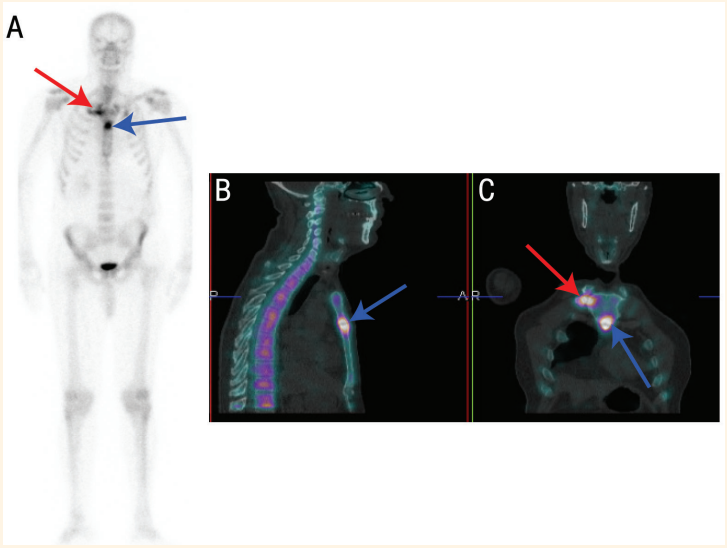

Figure 1: (**A**) Anterior views of the whole body bone scan provided with reconstructed scintigraphy with single-photon emission computed tomography/computerised tomography images in (**B**) sagittal and (**C**) coronal views showing intense uptake at the proximal end of the right clavicle (red arrows) and *manubrium sterni* (blue arrows). Faint radiotracer uptake were noted at the left sternoclavicular joint and left first rib.

A 43-year-old male patient with a history of left radical nephrectomy for renal cell carcinoma (RCC) on surveillance was presented to the rheumatology clinic of a tertiary care hospital in Muscat, Oman, in 2019 with severe pain at the manubrium and right sternoclavicular joint associated with acne-like lesions in both the palms and soles that was present for the past two years. On examination, swelling, redness and tenderness were present over the manubrium and right sternoclavicular joint. There were 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. A computed tomography (CT) of the chest, bone scintigraphy with single-photon emission computed tomography/CT and whole-body F-18 fluorodeoxyglucose-positron emission tomography/CT (FDG-PET/CT) 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, which resulted in a dramatic improvement in the patient's condition. This report demonstrates how multiple imaging modalities can help in diagnosing SAPHO syndrome along with the provided history and medical background. Early and accurate diagnosis is essential for the correct management and achieving a good outcome.

The reporting of the current case does not contravene with the regulations of the local institutional review board.

Comment

SAPHO is a rare syndrome that was first reported by Chamot *et al.* in 1987.[1] Other cases have been reported in the West and Japan.[1,2] The underlying pathophysiology is still unclear, although many hypotheses attributed this condition to a group of genetic, immunologic and infectious factors.[2,3]

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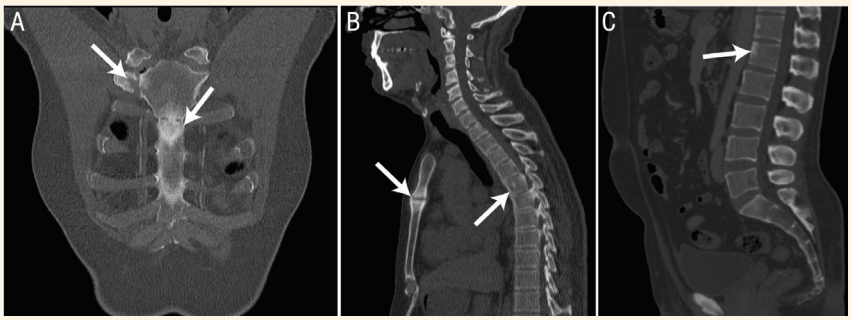

Figure 2: Computed tomography scans of the sternum (**A**) and spine (**B**) showing bony erosion and hyperostosis changes on the right sternoclavicular joint (red arrow) and sclerotic osteitis changes on either side of the sterno-manubrial joint (yellow arrows). Multiple scattered osteosclerotic lesions in the thoracic and lumbar spine (**B** and **C**) with relatively normal disc spaces (white arrows). A whole-body F-18 fluorodeoxyglucose-positron emission tomography/computerised tomography showed no definite focal radiotracer uptake on these lesions and both sacroiliac joints are grossly normal (images not provided).

SAPHO syndrome is a benign condition affecting mainly young and middle-aged adults and can be managed conservatively.[3] Early diagnosis and proper management will improve the clinical outcomes and prognosis.[1]

Clinical history, along with dermatological and radiological manifestations, is essential to diagnose SAPHO syndrome. Any one of these criteria are considered adequate to make the diagnosis: 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.[1,3] However, the diagnosis of SAPHO syndrome sometimes could be challenging, especially if skin manifestations are absent.[1] In the current 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; however, the whole body bone scintigraphy using 99mTc-methylene-diphosphonate is considered the most sensitive modality in detecting early bony involvement when these changes are absent or subtle in other modalities.[2,4] Anterior chest wall bones involving both sternoclavicular junction and manubrium sterni are considered the most common location for early bony changes of SAPHO syndrome, followed by the spine and, to a lesser extent, sacroiliac joints.[1,5] In the current case, these locations were involved apart from the normal appearance of both sacroiliac joints [Figures 1 and 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.[4] 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 such as osteitis and hyperostosis of sternoclavicular joint, sclerotic bony changes of vertebral bodies and sacroiliitis.[3,4] The CT modality gives more details about osteoarticular erosive changes, particularly of the anterior chest wall.[4] An 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] The current patient had a bone scan and CT scan, which showed similar comparable changes to the above-mentioned findings of SAPHO syndrome [Figures 1A and 2]. An FDG-PET/CT scan was performed to exclude the possibility of sclerotic metastasis in the background of RCC and was 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 the current case, NSAIDs, prednisolone and methotrexate were administered resulting in an improvement of the patient's condition with a good outcome on follow-up.

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. Both authors approved the final version of the manuscript.

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