

# Sacroiliitis and Spondylitis with Sternoclavicular Hyperostosis: SAPHO or an Ankylosing Spondylitis Variant?

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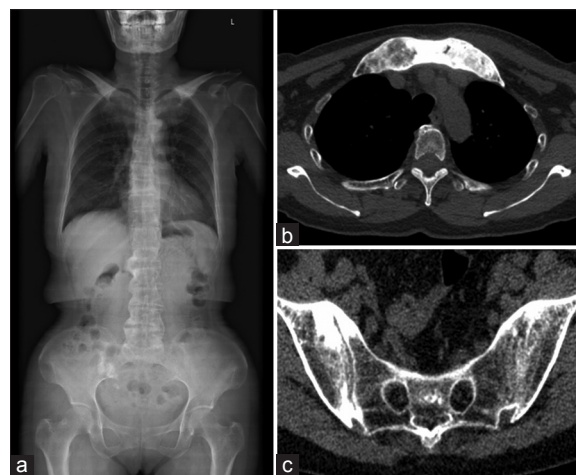
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To the Editor: SAPHO syndrome is a heterogeneous disorder characterized by synovitis, acne, pustulosis, hyperostosis, and osteitis. Its diagnosis can be difficult if there are no typical skin manifestations. We reported a case of SAPHO syndrome which also fulfills the current diagnostic criteria, for ankylosing spondylitis (AS).

A 62-year-old Chinese female patient was evaluated for her bilaterally swelling sternoclavicular joints in our center. She had a 30-year history of nonprogressive lower back pain and spinal stiffness without debilitating motion limitation of the spine and was once diagnosed of AS in our center according to the documented human leukocyte antigen B27 (HLA-B27) positivity. Ten years ago, she began to develop episodic pain in her sternoclavicular area and bilateral shoulders. Localized reddening and tenderness were noticed over her sternoclavicular joints and the medial clavicular areas during those episodes. Her pain could also be mitigated by oral meloxicam, and her daily activity was not affected.

Physical examination showed enlarged sternocostoclavicular joints and deformed shoulders. Motion range of her hip joint, spine, and chest wall appeared normal. No acne or pustulosis-like rashes were noted. Laboratory assays revealed slightly elevated C-reactive protein at 20 ng/ml; erythrocyte sediment rate rose to 50 mm/h; spinal X-ray demonstrated wide-spread osteosclerosis, vertebral erosion, and multilevel bony bridging formation and marginal syndesmophytes [Figure 1a]. Chest computerized tomography (CT) revealed fusing destruction of her sternocostoclavicular joints with obvious hyperostosis [Figure 1b]. Pelvic CT showed obvious joint space narrowing, osteosclerosis and erosion of bilateral sacroiliac joints [Figure 1c]. Her scintigraphic bone scanning showed increased uptake in the sternoclavicular joint, sternum, and bilateral clavicles forming “bullhead sign”. Finally, this patient was treated by oral meloxicam and thalidomide along with yearly infusion of zoledronate. No relapse of joint pain was reported in 6 months follow-up.

Diagnosis of SAPHO syndrome depends on the presence of at least one of three diagnostic criteria including osteoarticular manifestations of severe acne or palmoplantar pustulosis; hyperostosis of the anterior chest wall, limb, or spine; or chronic recurrent multifocal osteomyelitis (CRMO).<sup>[1]</sup> Although



**Figure 1:** (a) Ankylosing change on the spinal X-ray with ivory vertebrae and syndesmophytes on multiple levels. Cortical erosion and hyperostosis of the vertebral bodies with bony bridging were also noted. In addition, sternocostoclavicular hyperostosis with obviously localized enlargement and clavicular osteosclerosis were found. (b) Coronal computerized tomography image showed obviously enlarged sternoclavicular joints with marked erosion, hyperostosis, and osteosclerosis. Cortical erosions and local hyperostosis were noted on the thoracic vertebra. (c) Axial computerized tomography images of the sacroiliac joints showing marginal erosions, profuse sclerosis at the iliac side of the right sacroiliac joint and ankylosing fusion on the left sacroiliac joint.

sternoclavicular joint is the most frequent site of the disease in adults, sacroiliac joint lesion and the spinal lesion are not rare. Paravertebral ossifications in SAPHO may simulate syndesmophyte

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in AS. Erosion of sacroiliac joint accompanied by florid sclerosis and hyperostosis are characteristic of SAPHO spectrum.<sup>[2]</sup> Nevertheless, in adults as high as 30% of SAPHO patients showed HLA-B27 positivity. Many SAPHO cases also fulfilled the diagnostic criteria of AS or spondyloarthropathies (SpA).<sup>[3-5]</sup>

Interestingly, the spinal lesion and sacroiliitis, in this case, occurred 20 years previous to the sternocostoclavicular hyperostosis, and no skin lesion was found all through her 30 year's history. However, unlike typical AS or psoriatic arthritis, SAPHO is a benign disorder. Its spinal lesions are slow in progression and not, particularly debilitating.<sup>[1]</sup> Thus, the largely spared joints mobility and benign prognosis in this patient's spinal lesions after 30 years evolution favored the diagnosis of SAPHO syndrome.

In conclusion, the axial skeletal lesions in SAPHO syndrome appear to be a gray zone overriding the current classification criteria for SAPHO syndrome and SpA. Hyperostosis and CRMO as SAPHO's most specific manifestations may help to distinguish it from classic SpA.

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### Conflicts of interest

There are no conflicts of interest.

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