ANSWER TO PHOTO QUIZ (PAGE 399)

A BONE DISORDER WITH SKIN LESIONS

DIAGNOSIS

At first, the MRI appeared to show spondylodiscitis. For this reason, the patient was referred to internal medicine. However, this diagnosis seemed unlikely because there were no clinical signs of infection and although the MRI did show paravertebral oedema, the disc seemed unaffected.

Based on the combination of vertebral osteitis and pustulosis palmaris on both hands and feet, and a previous episode of sternoclavicular pain and swelling, we suspected synovitis, acne, pustulosis, hyperostosis and osteitis (SAPHO) syndrome. SAPHO syndrome is a rare inflammatory syndrome of bone, joints, and skin with the above-mentioned characteristics.

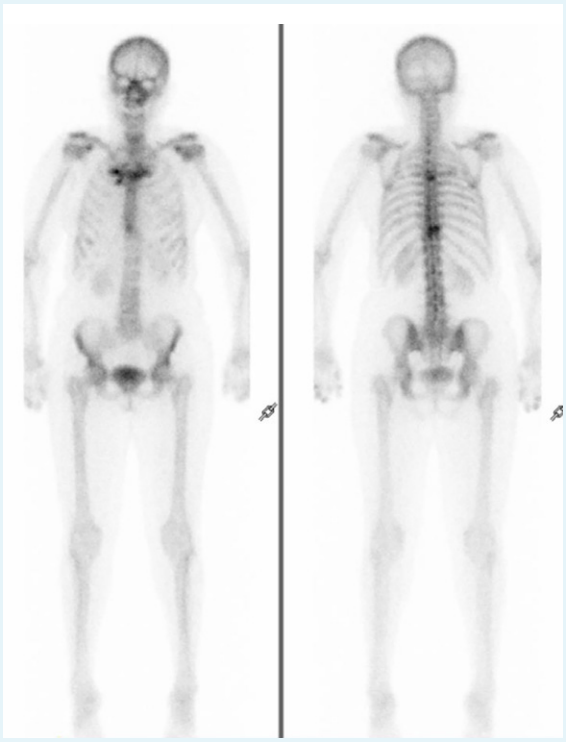
The pathogenesis of the syndrome is still unclear. Aetiology is most likely multifactorial with genetic factors as well as infectious and immunological complements contributing to the disease.[1,2] There are no validated diagnostic criteria for SAPHO syndrome. Diagnosis is based on the patient's history with characteristic combinations of features and typical radiological findings.[2] Other explanations like infection, malignancy, or classic rheumatic diseases should be excluded.

To confirm the diagnosis in this patient, we performed an additional bone scintigraphy showing a so-called 'bull's head' change caused by sternoclavicular hyperostosis, characteristic for SAPHO syndrome with sternoclavicular involvement (figure 3).[1] The diagnosis SAPHO syndrome was made.

Treatment is recommended for all patients to ease symptoms and prevent complications. Initial therapy consists of nonsteroidal anti-inflammatory drugs. Second-line treatment may include a tumour necrosis factor (TNF) inhibitor, methotrexate, bisphosphonates, or anti-interleukin-1 therapy.[2,3] There is little evidence for choice of second-line treatment and recommendations are mostly based on case series. Although treatment can be difficult, long-term prognosis is good if promptly diagnosed and treated.[3]

After starting naproxen (500 mg, 2 times daily), the patient's symptoms improved and she was discharged.

Figure 3. *Bone scintigraphy: sternoclavicular hyperostosis with sternoclavicular involvement and inflammation at Th4 and Th10*



During follow-up, the patient developed peripheral arthritis, also commonly seen in SAPHO syndrome. Due to progression of her disease, she started treatment with additional methotrexate. Because of side effects of methotrexate, she is now starting a TNF inhibitor adalimumab.

REFERENCES

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3. Colina M, Govoni M, Orzincolo C, Trotta F. Clinical and radiologic evolution of synovitis, acne, pustulosis, hyperostosis, and osteitis syndrome: a single center study of a cohort of 71 subjects. Arthritis Rheum. 2009;61(6):813-21.