

Whole-body Bone Scan in the Diagnosis and Treatment of SAPHO Syndrome

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ABSTRACT

Synovitis, acne, pustulosis, hyperostosis, and osteomyelitis (SAPHO) syndrome is rare disease of unknown pathogenesis. Its diagnosis and treatment are difficult. There is no standardised treatment for SAPHO syndrome, and the results of empirical therapy are unpredictable. The whole-body bone scan may help evaluate the efficacy of SAPHO syndrome treatment. The case of a 52-year Chinese male, affected by SAPHO syndrome, is presented and discussed in this report. After the patient received methotrexate, prednisone and non-steroidal anti-inflammatory drugs (NSAIDs) for six months, his clinical symptoms and whole body bone scan signs improved.

Key Words: SAPHO syndrome, Whole-body bone scintigraphy, Treatment efficacy.

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INTRODUCTION

Synovitis, acne, pustulosis, hyperostosis, and osteomyelitis (SAPHO) syndrome is a rare clinical entity involving musculoskeletal and dermatologic systems.¹ The main clinical features are chronic inflammatory osteoarthritis and dermatosis. Chest wall pain, more common in the sternoclavicular joints, along with palmo-plantar pustulosis and acne can point to this uncommon syndrome.² Herein, we report a case of a Chinese male, who was diagnosed as SAPHO syndrome and received methotrexate, prednisone and non-steroidal anti-inflammatory drugs (NSAIDs) for more than six months. Subsequently, his clinical symptoms and whole-body bone scan signs improved.

CASE REPORT

In September 2017, a 52-year Chinese male patient was admitted to our hospital with complaints of pain and swelling in the anterior chest wall and lumbago regions for eight months, without skin lesions.

Physical examination revealed pustulosis on the face, palms and soles. The laboratory evaluation revealed red blood cell count of $4.01 \times 10^{12}/L$ (reference range: $4.3-5.8 \times 10^{12}/L$), hemoglobin 113 g/L (reference range: 130-175 g/L), platelet count $362 \times 10^9/L$ (reference range: $125-350 \times 10^9/L$), erythrocyte sedimentation rate (ESR) 120 mm/h (reference range: 0-20 mm/h), C-reactive protein (CRP) 95.2 mg/L (reference range: 0-7.44 mg/L) and interleukin-6, 39.9 pg/ml (reference range: 0-5.9 pg/ml).

In addition, coagulation function, liver and renal functions, rheumatoid factor, lupus and other autoimmune disease-specific antibodies were all in normal ranges.

Positron emission tomography-computed tomography (PET-CT) showed bone destruction or increased bone density in bilateral sternoclavicular joints, bilateral clavicles, sternum, right first to third ribs, left first to sixth ribs, and abnormal increase of FDG uptake (SUVmax, 3.4); at the same time, no other malignant solid tumors were found by PET-CT imaging (Figure 1). In order to understand the metabolism of the whole body, the whole-body bone imaging was further performed. Whole-body bone scintigraphy (Figure 2a) revealed increased tracer uptake in bilateral sternoclavicular joints, bilateral clavicles, sternum, right first to third ribs, left first to sixth ribs and fourth lumbar vertebrae. The characteristic "bull-head" sign raised suspicion of SAPHO syndrome. According to the diagnostic criteria of Benhamou *et al.*, SAPHO syndrome was finally diagnosed.¹

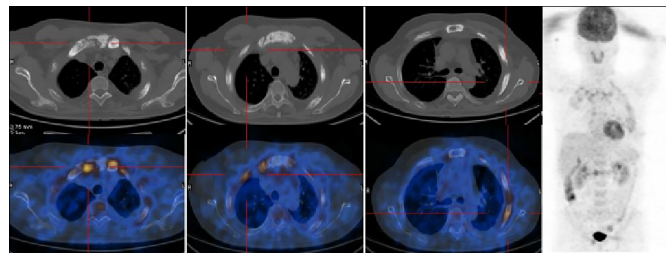


Figure 1: Positron emission tomography-computed tomography (PET-CT) with 18F-FDG showed bone destruction or osteosclerosis and increased tracer uptake in bilateral sternoclavicular joint, bilateral clavicles, sternum and multiple rib joints (SUVmax, 3.4).

The patient was treated with methotrexate, 20 mg/week, prednisone, 10 mg/day and calcium tablet, 1 g/day. The NSAID drug, diclofenac, 150 mg/day, was added to the treatment, which improved the anterior chest wall and lumbago pain.

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After six months, the CRP level and ESR returned to normal. At the same time, the patient stopped taking medicine on his own initiative due to alleviation of the anterior chest wall and lumbago pain. In order to further evaluate the changes of bone metabolism, whole-body bone imaging was performed again. The whole-body bone scan showed decreased uptake of ^{99m}Tc -MDP in the right first to third ribs, left first to sixth ribs and fourth lumbar vertebra (Figure 2b). Single photon emission computed tomography-computed tomography (SPECT-CT) scan was performed to further evaluate bone density changes. Fused SPECT-CT coronal and axial images (Figure 3) showed that the destructive, expansile, osteolytic and osteosclerotic lesions of the sternum, and bilateral ribs were improved.

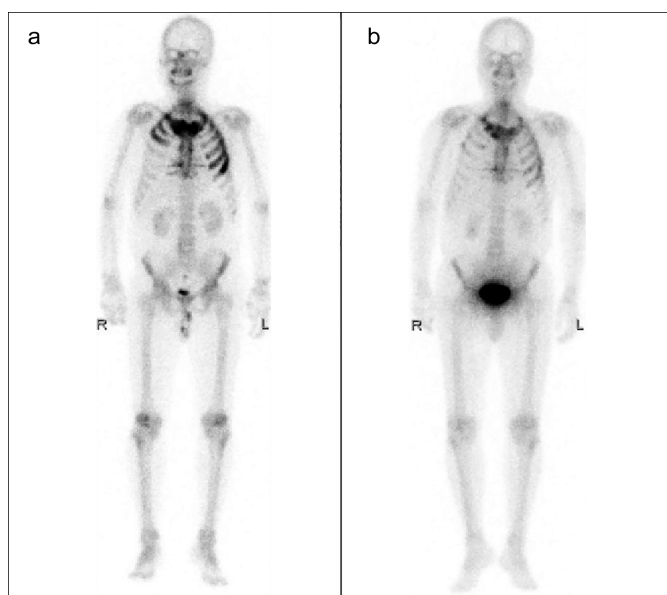


Figure 2: Whole-body bone scan with ^{99m}Tc -MDP images before and after treatment. a) Increased tracer uptake in bilateral sternoclavicular joints, bilateral clavicles, sternum, right first to third ribs, left first to sixth ribs and fourth lumbar vertebra. b) decreased tracer uptake of original lesions.

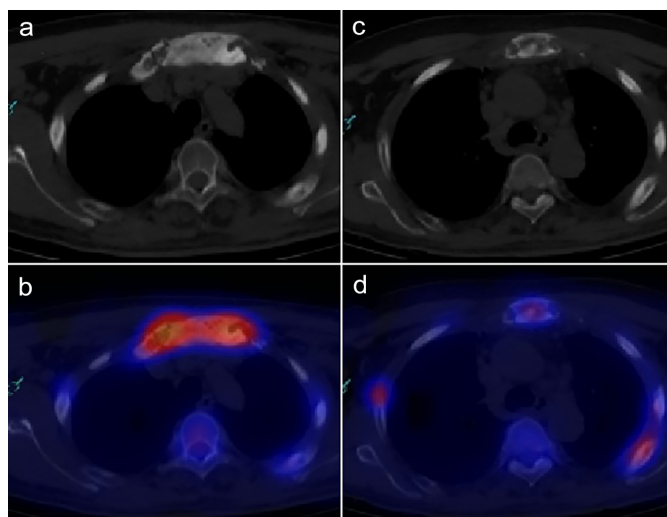


Figure 3: Fused SPECT-CT coronal and axial images (a-d) show the destructive, expansile, osteolytic and sclerotic lesions of the sternum, and bilateral ribs have improved.

DISCUSSION

Chamot *et al.*,² were the first group who proposed the name of SAPHO syndrome in 1987. Whole-body bone scan is a highly sensitive examination method for chest wall lesions in patients with SAPHO syndrome. The pattern of increased tracer uptake in the anterior chest wall may resemble a “bull’s head”, which is the characteristic sign of SAPHO syndrome in bone scans.³

^{18}F -FDG PET/CT may be another useful method, which could find more lesions or concealed lesions. The increase of FDG uptake in the anterior chest wall may precede the appearance of skin lesions and the appearance of obvious abnormality on X-rays. In the advanced stages of SAPHO syndrome, PET/CT is helpful to distinguish active from inactive bone lesions. In our case, this patient had PET/CT scan first, which showed that the lesions had low to moderate ^{18}F -FDG uptake, which was not typical enough to diagnose SAPHO syndrome. The reason of different uptake of imaging agent in PET/CT may be the co-existence of osteolysis and osteosclerosis, which is another feature of SAPHO syndrome. Then, the patient underwent a whole-body bone scan, which revealed the “bull’s head” sign and high uptake of multiple rib joints. Leone *et al.*⁴ have reported that 60–95% bone destructions, which comprise hyperostosis, osteolysis, osteitis, and osteosclerosis, occurred in the area of anterior chest wall, especially in the costochondral and sternoclavicular joints and spine. In our case, the bone scan revealed intense uptake in bilateral sternoclavicular joints, bilateral clavicles, sternum, right first to third ribs, left first to sixth ribs and fourth lumbar vertebra, which were consistent with the above results.

Although there are many researches on the disease, effective treatment is still limited. The current treatment goal for this disease is to relieve symptoms. Skin lesions, such as pustulosis palmaris and plantaris, are difficult to distinguish from psoriasis rash.⁵ However, for highly active bone and joint damage (erosion, hyperosteo-geny and osteoarthritis), long-term use of methotrexate can achieve satisfactory results.⁶ There are several studies which have proved that the use of steroids and methotrexate may obtain an effective treatment result in inhibiting bone damage.⁷⁻⁹ In our case, after six months of treatment with methotrexate, 20 mg/week, the patient felt much better than before, and two years later, the whole-body bone scintigraphy showed decreased uptake by all lesions. This case, on the one hand, has proved the positive effect of methotrexate on the bone damage of SAPHO syndrome; and on the other hand, it has also effectively proved that bone scanning plays an important role in the evaluation of the therapeutic effect in this disease. However, at present, the treatment of SAPHO syndrome is not standardised and has some challenges, such as optimum dose and duration of treatment and compliance by the patients. Therefore, an appropriate treatment plan and regular follow-up are particularly important. Future research should focus on defining the optimum and effective treatment regimen for this uncommon disease.

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PATIENT'S CONSENT:

Informed consent was obtained from the patient to publish the data regarding this case.

CONFLICT OF INTEREST:

The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

CJ: Designed and authored the manuscript.

YZ: Provided concept of the manuscript and proofread the final version before submission and performed the procedure.

XL: Contributed in analysis, acquisition, revision and final proof.

HH: Data collection, and literature review.

CL: Collected the pathological findings and follow-up of the patient.

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