



A Case Report of SAPHO Syndrome Treated with Adalimumab and Review of Related Literature

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Abstract

SAPHO (Synovitis, Acne, Pustulosis, Hyperostosis and Osteitis) Syndrome is a kind of clinical syndrome characterized by skin, bone and bone joint lesions. The clinical manifestations are synovitis, acne, pustulosis, hyperostosis and osteitis. Because of the low incidence rate of this disease, missed diagnosis and misdiagnosis are easy. This paper reports a new case of SAPHO syndrome with early diagnosis and treatment of adalimumab.

Keywords: SAPHO syndrome; Adalimumab; Diagnosis; Treatment

Introduction

SAPHO syndrome was first summarized and described by Chamot to specify a group of diseases related to bone, joint and skin lesions [1]. SAPHO is the acronym of synovitis, acne, pustulosis, hyperostosis and osteitis. The pathogenesis of the disease is still unclear, and the clinical manifestations are complex and diverse. The main characteristics of the disease are bone and joint damage and skin lesions. Sometimes the two occur at different times, and skin lesions may or may not exist in SAPHO syndrome. In addition, there is no specific laboratory examination for the time being, and clinician's lack of understanding of the disease, which makes the disease prone to missed diagnosis and misdiagnosis. At present, there is no unified standard guideline for diagnosis and treatment of SAPHO syndrome, most of which are empirical treatments, consequently, the phenomenon of poor therapeutic effect also exists.

The damage of this disease to the bone system can cause joint deformity, cause irreversible dysfunction, and bring heavy burden to the life of patients. Skin lesions also seriously affect the physical and mental health of patients. Therefore, early diagnosis and treatment are very necessary to alleviate the pain of patients, inhibit the development of the disease, reduce the occurrence of irreversible injury, and achieve the best effect of treatment. A new case of SAPHO syndrome is reported below.

Case Presentation

An 18-year-old female presented with swelling and pain of the left sternoclavicular joint in July 2020 without obvious inducement. She went to another hospital for chest CT examination, which showed that the sternal end of the left clavicle was hypertrophic, and she took oral analgesics (specific drugs are unknown). On November 7th, 2020, due to swelling and pain of the left sternoclavicular joint for more than 4 months, he visited the outpatient department of Rheumatology, Affiliated Hospital of Jiangxi University of traditional Chinese medicine. The symptoms were as follows: Swelling and pain of the left sternoclavicular joint, high local skin temperature, reddish skin color, limited activities such as deep breathing and chest expansion, no pain and discomfort of the remaining joints, no morning stiffness, and multiple pustules left scars on the feet, and a small amount of acne scattered on the face. Appetite and somnus were well, urination and defecation were fine, the tongue was pink and the fur was white, and the pulse was thready. She denied the history of food and drug allergy, hepatitis, tuberculosis and other infectious diseases, family genetic history, and no other special personal history.

Physical examination: No obvious abnormality was found in the systematic examination. The left sternoclavicular joint was red and swollen with tenderness. The sternoclavicular joint was limited in movement. There were multiple pustules and scars left on both feet. There was no tenderness in the pyramids and lumbosacral region.

Auxiliary examination: Chest CT showed that the sternal end of the left clavicle was

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Received Date: 13 Aug 2021

Accepted Date: 21 Sep 2021

Published Date: 24 Sep 2021

Citation:

Xu Wei-dong, Liu Yang, Mao Ying. A Case Report of SAPHO Syndrome Treated with Adalimumab and Review of Related Literature. *Ann Clin Case Rep.* 2021; 6: 1997.

ISSN: 2474-1655

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Figure 1: Chest CT.

hypertrophic and swollen, the articular surface of the sternoclavicular joint was irregular, and the joint space was narrow (Figure 1). HLA-27 typing: negative. Tuberculosis antibody test: Negative.

Clinical diagnosis: SAPHO syndrome

Treatment and follow-up: Adalimumab injection 40 mg, a single dose of subcutaneous injection every two weeks. After 12 weeks, the swelling of sternoclavicular joint subsided and the pain symptoms relieved significantly.

Discussion

SAPHO syndrome is a kind of clinical syndrome characterized by bone and joint damage and skin lesions. The clinical manifestations are synovitis, acne, pustulosis, hyperostosis and osteitis. The clinical manifestations are diverse and the specificity is poor. The cause of the disease is not clear. The existing theories mainly include gene mutation theory, genetic susceptibility theory, and autoimmune theory and *Propionibacterium acnes* infection theory [2]. The incidence rate of this disease is not accurate. There are more women than men in all ages.

Bone and joint damage and skin lesions are the main characteristics of SAPHO syndrome. Bone and joint lesions are usually characterized by synovitis, bone hypertrophy, osteoarthritis, etc. In the early stage of onset, the lesions are mainly inflammatory changes of ligament attachment points. With the development of the disease, osteolysis and bone hyperplasia can gradually appear [3]. The lesions were mainly located in sternum, sternal end of clavicle, first anterior rib and costal cartilage, sternoclavicular joint, sternal stalk body joint, thoracic spine, lumbar spine, etc. [4]. Sternoclavicular joint, sternoclavicular joint and body joint were the most easily involved, accounting for 65% ~ 90%. Spine was also the most frequently involved part, in which thoracic spine was the main involved part, followed by lumbar spine and cervical spine, and caudal vertebra was the least involved [5]. The first case was bone injury in other parts [6]. The results of imageology detection are helpful for the early clinical diagnosis of SAPHO syndrome. The whole body imaging "bull's head sign" is the most characteristic, and the multi-slice spiral CT is the best way to check the chest lesions in the prone parts, while MRI is more sensitive for the early lesions of spine, sacroiliac joint and hip joint [5]. Bone and joint lesions are highly destructive, often causing pain and discomfort in patients, causing joint and bone deformity, causing irreversible dysfunction, causing serious damage to the patient's body. According to the previous diagnostic criteria, bone or joint lesions are necessary for the diagnosis of SAPHO syndrome. Most of the skin lesions of SAPHO syndrome are palmoplantar pustulosis, pustular psoriasis, psoriasis

vulgaris, suppurative hidradenitis and severe acne, etc. Typical skin lesions may or may not exist in patients with SAPHO syndrome, which means that skin lesions are not necessary for the diagnosis of SAPHO syndrome. Clinically, combined with the bone and skin lesions, comprehensive analysis is helpful for diagnosis. At present, there is no unified diagnostic standard for the diagnosis of SAPHO syndrome, and its clinical manifestations are complex and changeable, and the early symptoms are not typical, so it is often missed diagnosis and misdiagnosis. Now we refer to the four criteria recommended in 2012 [7]: 1) Osteoarthritis with aggregate acne or explosive acne or suppurative hidradenitis; 2) osteoarthritis with palmoplantar pustulosis; 3) hyperostosis of upper chest wall, acromegal bone and spine; 4) chronic recurrent multifocal osteomyelitis of axial bone or peripheral bone with or without skin damage; as long as one of the four criteria is consistent, it can be diagnosed with SAPHO syndrome.

The main clinical manifestation of the patient was unilateral sternoclavicular joint inflammation, with pustules left on the foot skin and a small amount of acne on the face. The imaging showed that the sternal end of the left clavicle was hypertrophic and swollen, the sternoclavicular joint surface was irregular, and the joint space was narrow. Laboratory examination showed that HLA-B27 typing was negative, so as to identify the patient with spondyloarthritis. Therefore, combined with the patient's history, clinical manifestations and related auxiliary examination, the diagnosis of SAPHO syndrome was made.

At present, there is no unified treatment standard for SAPHO syndrome, and most of them are empirical treatment. Non-Steroidal Anti-Inflammatory Drugs (NSAIDs), glucocorticoids, Disease-Modifying Anti-Rheumatic Drugs (DMARDs), immunosuppressant's and biological agents are often selected for clinical treatment. If there are serious skin diseases, symptomatic treatment should be given. Among them, non-steroidal anti-inflammatory drugs are usually used as the first-line drugs in the treatment of SAPHO syndrome. With the wide use of biological agents, biological agents are gradually applied to the treatment of this disease. Tumor necrosis factor alpha receptor antagonists and interleukin receptor antagonists are often used as therapeutic options. Early treatment with biological agents can reduce the activity of the disease, inhibit the development of the disease and prevent the occurrence of irreversible injury. According to the patient's medical history, clinical experience and medication intention, adalimumab injection 40 mg was given subcutaneously every two weeks with a single dose, which acquired good therapeutic effect. After 12 weeks, the swelling of sternoclavicular joint subsided and the pain symptoms were significantly relieved.

Because the adverse reactions of adalimumab injection include

severe infection, tuberculosis, invasive fungal infection and other opportunistic infections appear in patients receiving adalimumab treatment, it is necessary to monitor the active tuberculosis symptoms and symptoms of patients receiving adalimumab treatment [8]. According to the need of clinical medication, tuberculosis antibody should be detected regularly.

SAPHO syndrome is rare in clinic, and its early symptoms are not typical. It is easy to be confused with other diseases, so clinicians are challenging in the diagnosis. The diagnosis of SAPHO syndrome mainly depends on the typical imaging findings. At present, there is no unified treatment standard for SAPHO syndrome. Most of the patients choose non-steroidal anti-inflammatory drugs, glucocorticoids, DMARDs, immunosuppressant's and biological agents. It can be seen from this case that early diagnosis and timely treatment and intervention of integrated traditional Chinese and Western medicine can effectively relieve the symptoms of patients and inhibit the development of the disease.

Funding

This study was supported by Young Key Talents Project of TCM in Jiangxi Province.

References

1. Guangju W, Guoan Z. CT features of SAPHO syndrome. *J Clin Radiol*. 2015;34(3):435-9.
2. Shen K, Yang L, Yin G, Xie QB. Sacroiliitis and spondylitis with sternoclavicular hyperostosis: SAPHO or an ankylosing spondylitis variant? *Chin Med J (Engl)*. 2016;129(1):110-1.
3. Xin-Yu L, Yu-Chen L, Chen L, Yu-Qian Y, Yi-Han C, Wei L, et al. Unveiling the mystery of long-term repeated multifocal-bone swelling and pain with acne-like rash. *Med J Peking Union med College Hospital*. 2021.
4. Chamot AM, Benhamou CL, Kahn MF, Beranek L, Kaplan G, Prost A. [Acne-pustulosis-hyperostosis-osteitis syndrome. Results of a national survey. 85 cases]. *Rev Rhum Mal Osteoartic*. 1987;54(3):187-96.
5. Xiangming S, Chuangjian L, Wei G. Analysis for the imaging features and clinical characteristics of SAPHO syndrome. *Hebei Med J*. 2020;42(18):2783-6.
6. Bing-Ju G, Jun L, Li-Zhen P, Yu Q, Li-Song L, Lisong L. Clinical analysis of 5 cases of SAPHO syndrome with mandible osteomyelitis as initial presentation. *China J Oral Maxillofacial Surg*. 2018;16(6):528-32.
7. Nguyen MT, Borchers A, Selmi C. The SAPHO syndrome. *Semin Arthritis Rheum*. 2012;42(3):254-65.
8. Liang W, Zhi-Gang z. Clinical Application of Antibody Drugs in China and Its Pharmaceutical Care Situati. *Drug Evaluat*. 2014;11(6):12-6.