CASE REPORT

Heart failure complicating with SAPHO syndrome

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SUMMARY

A 65-year-old man was referred to our hospital with dyspnoea due to acute heart failure. He presented with swelling in the left clavicle and pustulosis on both soles. An antihypertensive drug and non-invasive positive pressure ventilation improved his condition rapidly. Since all his physical symptoms were compatible with the criteria of SAPHO (synovitis, acne, pustulosis, hyperostosis, osteomyelitis) syndrome, we suspected that the SAPHO syndrome might cause acute heart failure. The aetiology between SAPHO syndrome and heart failure is unclear. Further studies are needed to clarify their relationship.

BACKGROUND

SAPHO (synovitis, acne, pustulosis, hyperostosis, osteomyelitis) syndrome is a rare disease. The rarity and various aspects of this syndrome are attributed to delayed diagnosis. The mechanism of SAPHO syndrome has not been established well. We experienced a rare case of SAPHO syndrome with heart failure.

CASE PRESENTATION

A 65-year-old man was admitted to our hospital with dyspnoea. He called an ambulance because he felt fatigue and headache followed by dyspnoea while he was watching television at midnight. He usually takes non-steroidal anti-inflammatory drugs (NSAIDs) due to recent left chest pain under the clavicle. His vital signs on arrival were: heart rate 140 bpm, blood pressure 218/116 mm Hg, oxygen saturation 90% with 10 L per minute oxygen, respiratory rate 30 breaths per minutes. He was also breaking into a cold sweat due to dyspnoea. His laboratory data were as follows: white cell count 19 400/uL, C reactive protein 22.1 mg/dL, erythrocyte sedimentation rate 112 mm/60 min, brain natriuretic peptide 650 pg/mL. His chest X-ray revealed cardiomegaly and enlargement of the pulmonary artery at the bilateral hilum, suggesting congestive heart failure (figure 1). An echocardiogram showed a low ejection fraction of the left ventricle (46%) without any asynergy. He was diagnosed with acute heart failure and admitted to receive non-invasive positive pressure ventilation and antihypertensive therapy. After the treatment, his symptoms improved rapidly.

Although we tried to detect a cause of heart failure, no major infection or coronary disease was recognised. CT presented bilateral large and swelling clavicles, consistent with hyperostosis (figure 2). He also had persistent back pain and pustulosis palmo-plantaris on both soles since 2 days before admission (figure 3). He was diagnosed with SAPHO

Figure 1 Cardiomegaly and bilateral hilum congestion in chest roentgen.

Figure 2 Hyperostosis of both clavicles in CT.

syndrome and it was hypothesised that the inflammation due to SAPHO might induce heart failure.

OUTCOME AND FOLLOW-UP

On postadmission day 5, he was transferred to the department of cardiology to treat heart failure. An antihypertensive drug and non-invasive positive pressure ventilation improved his condition rapidly.

DISCUSSION

In 1987,¹ SAPHO syndrome was defined as synovitis, acne, pustulosis, hyperostosis and osteitis which might occur at various ages. There is no gender difference in the incidence of SAPHO syndrome. Although various hypotheses regarding bacteriological or immunological mechanisms have been proposed, the exact pathology has not been established yet.²

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Figure 3 Pustulosis palmoplantaris in both soles.

Some criteria and clinical features are compatible to spondyloarthropathy. Benhamou *et al*³ described the criteria, (1) osteoarticular manifestations in severe acne, (2) osteoarticular manifestations in palmoplantar pustulosis, (3) hyperostosis with or without dermatosis, and (4) recurrent multifocal chronic osteomyelitis involving the axial or peripheral skeleton, with or without dermatosis. Kahn and Khan⁴ also described the criteria of SAPHO syndrome, but the prevalent and orthodox criteria have not been established yet.⁵

Venous thrombosis is found as a lethal complication of SAPHO syndrome. Ishizuka and colleagues reported a case of progressive exertional dyspnoea with decreasing left ventricular ejection fraction and frequent non-sustained ventricular tachycardia. This patient showed bilateral subclavian vein occlusion due to SAPHO syndrome. An operation to implant a cardioverter defibrillator was performed to treat refractory ventricular fibrillation.⁶ Several vein obstructions such as the subclavian vein, the iliac vein, the pulmonary vein and the superior vena cava obstruction have been published. Although systemic inflammation might contribute to the hypercoagulopathy, the pathogenesis of venous thromboses of SAPHO syndrome is not elucidated.

Immunologically, human leucocyte antigen (HLA) B27 positive might be associated with SAPHO syndrome, but most patients with SAPHO are negative for HLA B27.⁷ Although the most common osteomyelitis in SAPHO syndrome is in the sternoclavicular joint, symptoms could appear in various lesions. Tsugawa and colleagues described a case with recurrent episodes of headache. MRI of the head revealed thickening and enhancement of the right frontal dura with abnormal signal intensity and enhancement of bone marrow at the same lesions.⁸

Definitive treatment does not exist and is not necessary in the SAPHO syndrome. NSAIDs are widespread and effective. Steroids, colchicine, doxycycline, salazopyrin, interferon and bisphosphonates have been reported to be effective.⁹

Along with the above criteria, the diagnosis as SAPHO syndrome was made in our patient. While taking pain killers, the main target of the treatment was heart failure. Non-invasive positive pressure ventilation and antihypertensive drugs were administered.

Major symptoms of SAPHO syndrome are skin disease or osteitis, which occurs in 84% and 91% of patients, respectively.¹⁰ To the best of our knowledge, this is the first report describing SAPHO syndrome complicating an acute heart failure except for pulmonary embolism.

We hypothesised the following mechanism. First, infection might induce heart failure. Magrey and Khan¹¹ reported that *Propionibacterium acnes*, which is the most frequent pathogenic organism recovered from hyperostosis, might be a cause of the systemic inflammation leading to heart failure. Although blood culture or biopsy was not collected from our patient during the acute phase, no specific organism is usually identified from an osteomyelitis in many cases.

Second, unbearable pain caused by SAPHO syndrome led to after load mismatch. At the time when heart failure happened, he also felt pain.

Third, unknown humoral mediators such as cytokines or leucotrienes caused by SAPHO syndrome might induce severe inflammation. These hypotheses have not been demonstrated yet. Further studies and reports are expected.

Learning points

- ▶ A case of SAPHO (synovitis, acne, pustulosis, hyperostosis, osteomyelitis) syndrome with heart failure was described herein. Its aetiology and pathogenesis is not well understood yet.
- ▶ Our case indicated that heart failure might complicate the SAPHO syndrome. When clinicians are faced with patient with SAPHO syndrome, we have to be aware of heart failure under that condition. Further studies and cases are needed to confirm the mechanism.

Contributors TN and SK joined with the treatment of this patient. SI designed this manuscript. SN gave a final approval to submit this manuscript.

Competing interests None declared.

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