Clinical Case Report

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Purulent constrictive pericarditis caused by Salmonella enteritidis in a patient with adult-onset Still's disease

A case report

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Abstract

Rationale: Purulent pericarditis is a rare and usually fatal disease. Immunodeficiency state and preexisting pericardial effusion can predispose patients to infections. However, we are not aware of similar cases in patients with adult-onset Still's disease (AOSD). In addition, it is seldom caused by *Salmonella* bacteria.

Patient Concerns: We report a 30-year-old woman with dyspnea on exertion and epigastric fullness. She was newly diagnosed with AOSD 4 months previously and medicated with prednisolone.

Diagnoses: Transthoracic echocardiography (TTE) and computed tomography revealed a thickened pericardium with loculations in the pericardial space, consistent with purulent constrictive pericarditis. Subsequent cultures of blood and pericardial fluid yielded *S enteritidis*.

Interventions: She underwent subtotal pericardiectomy through a limited median sternotomy, and antibiotic therapy (ceftriaxone) for 1 month.

Outcomes: The New York Heart Association functional classification downgraded from class III to class I. There was no recurrence during the 1-year follow-up.

Lessons: This case presents an opportunity to highlight the importance of considering purulent pericarditis in patients previously diagnosed with AOSD. High clinical suspicion, early diagnosis, and prompt management can result in a better outcome in purulent pericarditis.

Abbreviations: AOSD = adult-onset Still's Disease, TTE = transthoracic echocardiography.

Keywords: adult-onset Still's disease, constrictive pericarditis, purulent pericarditis, *Salmonella enteritidis*

1. Introduction

Purulent pericarditis is a rare and usually fatal disease. It is characterized by a rapid accumulation of suppurative pericardial fluid. Mortality remains high (40%) despite appropriate therapy, which are closely related to cardiac tamponade, septic shock, and pericardial constriction.[1,2] Predisposing factors include immunodeficiency, previous cardiac surgery, chest trauma, and preexisting pericardial effusion (uremia, congestive heart failure, connective tissue diseases).[3] To the best of our knowledge, purulent pericarditis has been reported in association with following autoimmune diseases such as systemic lupus erythematosus, rheumatoid arthritis, mixed connective tissue disease, and dermatomyositis.[4,5] However, we are not aware of similar cases in patients with adult-onset Still's disease (AOSD). Herein, we report an extremely rare case of AOSD patients with purulent constrictive pericarditis caused by *Salmonella enteritidis*.

2. Case report

A 30-year-old Taiwanese woman presented with intermittent spiking fever and sore throat for 5 days. She also described the migratory pain at sole, ankle, knee in previous 3 weeks. She visited our outpatient department, where salmon-like pink rash was found at her ankle and knee. According to the Yamaguchi criteria, she was newly diagnosed with AOSD. Because of relapsing spiking fever, she was medicated with high-dose systemic corticosteroid therapy for 3 days (methylprednisolone 500 mg/d) then shifted to oral prednisolone (0.5 mg/kg/d). Ten days later, she was readmitted because of intermittent fever and diarrhea. Chest radiography revealed mild left-sided pleural

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effusion. Blood cultures indicated the presence of *S enteritidis* bacteremia. She received intravenous ceftriaxone (2000 mg/d) for 1 week. *S enteritidis* was absent in follow-up blood cultures. Chest radiography and transthoracic echocardiography (TTE) were normal with no evidence of fluid accumulation.

However, she gradually suffered from dyspnea on exertion, nausea with vomiting, and frequent epigastric fullness in recent 2 months. In the emergency department, her initial vital signs were as follows: temperature, 36.2 °C; heart rate, 105 beats/minute; respiratory rate, 24 breaths/minute; blood pressure, 102/76 mmHg; and oxygen saturation, 95%. Chest auscultation revealed bilateral basal rales, and cardiac auscultation revealed mildly diminished heart sound without pericardial knock or friction rub. An electrocardiogram revealed sinus tachycardia without ST segment elevation. Chest radiography revealed massive left-sided pleural effusion. Blood investigations revealed severe infectious disorder, with a white blood cell count of 11800/L, C-reactive protein level of 118.2 mg/L, and procalcitonin level of 0.30 ng/mL. Blood cultures disclosed recurrence of *S enteritidis* bacteremia. She was admitted and treated with intravenous ceftriaxone (2000 mg/d). A pigtail catheter (8.5 French) was inserted and 500 mL of serosanguinous pleural effusion was drained out on the first day.

During hospitalization, the patient remained critically ill in spite of pigtail catheter drainage and antibiotic therapy. TTE was obtained which displayed a thickened pericardium with hyperechoic nonfloating bodies in the pericardial space, resulting in left ventricular posterior wall diastolic collapse (Fig. 1A). In addition, pulsed-wave Doppler analysis displayed exaggerated respiratory variation in mitral inflow velocity (Fig. 1B) and expiratory diastolic flow reversals of prominent hepatic vein, consistent with purulent constrictive pericarditis. Computed tomographic angiography confirmed the findings of TTE with loculated pericardial effusion and consolidation foci in the left lower lobe, and with no evidence of mycotic aneurysm (Fig. 2).

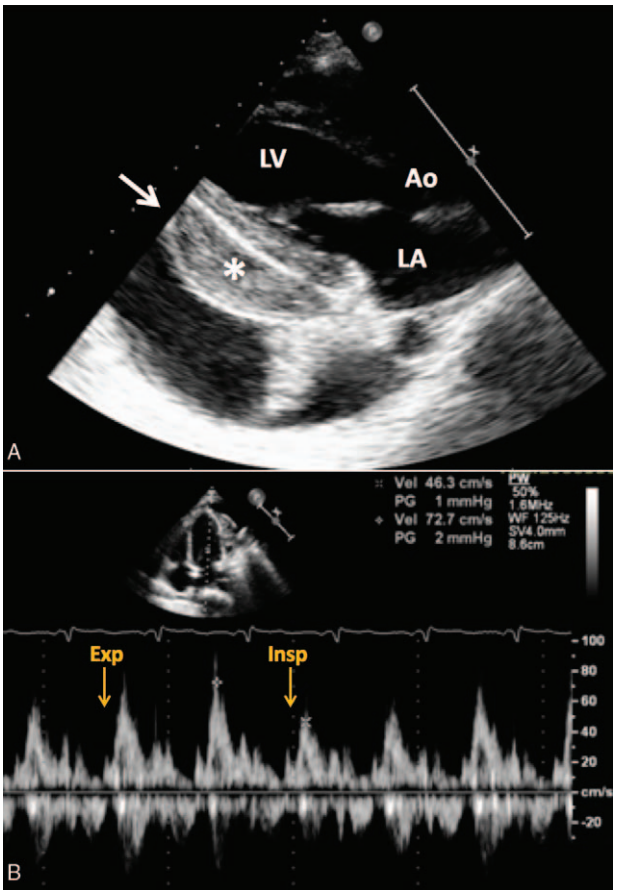

Figure 1. Transthoracic echocardiography. (A) Parasternal long-axis view displayed a thickened pericardium (arrow) with non-floating bodies in the pericardial space (asterisk). Ao = aorta; LA = left atrium, LV = left ventricle. (B) Pulsed-wave Doppler analysis displayed inspiratory decrease and expiratory increase in early mitral flow velocity. Exp = expiration, Insp = inspiration.

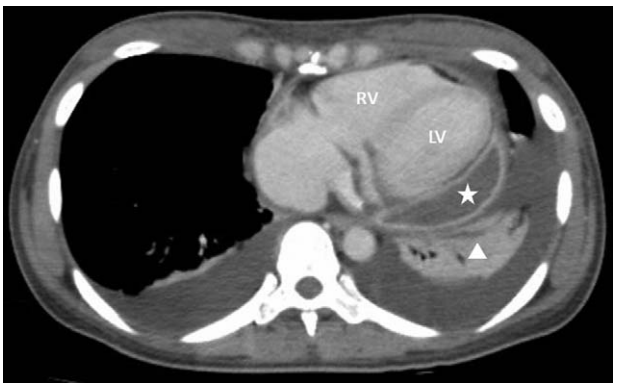

Figure 2. Computed tomographic angiography. Intravenous contrast-enhanced axial images demonstrated a thickened pericardium (arrow) with loculated pericardial effusion (asterisk). Consolidation foci were also noted in the left lower lobe (triangle). LV = left ventricle, RV = right ventricle.

After obtaining informed consent, she underwent subtotal pericardiectomy through a limited (9 cm) median sternotomy. The markedly thickened pericardium was observed and incised vertically by a scalpel. The purulent debris and loculations in the pericardial cavity were manually evacuated and sent for microbial study. Fibrous adhesions between the parietal and visceral pericardium were made free. Then, the pericardium was gently stripped from the right atrium to the right ventricle. Hemodynamic status remarkably improved during the operation, with central vein pressure from 20 to 11 mmHg and mixed venous oxygen saturation from 38% to 79%. Histopathologic examination of the resected pericardium revealed a fibrous thickening. Subsequent cultures of pericardial fluid yielded *S enteritidis*. The patient received the same antibiotic regimen of ceftriaxone (2000 mg/d) in the hospital for 1 month. The New York Heart Association functional classification downgraded from class III to class I. There are no reaccumulation of pericardial effusion and no recurrent constrictive pericarditis on serial postoperative TTE.

3. Discussion

Purulent pericarditis continues to result in a very serious prognosis and high mortality. The population incidence was estimated approximately 1/18,000 persons, with a significant decline attributed mainly to the widespread use of effective antibiotics.[2,3] Since antibiotics introduction into medicine in the 1940 s, the causative pathogens have been changes. *Staphylococcus aureus* has predominated during the 60-year period. Besides, the proportion of gram-negative bacilli and anaerobic organisms are increasing throughout the world.[6]

Salmonella species are gram-negative, flagellated, facultatively anaerobic bacilli that primarily cause gastroenteritis. Once the

bacterial load reaches the threshold level, they may spread into the bloodstream and develop to lethal cardiovascular complications. In contrast to endovascular infections and endocarditis, *Salmonella* species are less likely to involve the pericardium. A previous review of 19 patients, by Ortiz et al[7] in 2014, is considered as the largest study thus far. This review assumed that chronic pericardial effusion might serve as a nidus for *Salmonella* infections. The most common serotype is *S enteritidis* instead of *S typhimurium*, which represented about 50% of reported cases in the last century.[8] This discrepancy is probably due to the raised prevalence of *Salmonella* serogroup D and their unique virulence traits.

Early recognition and diagnosis of purulent pericarditis is still in challenge. Nonspecific systemic inflammatory response syndrome is often the first clinical manifestation, and the classic signs of pericarditis may be absent.[2] It requires a high index of suspicion in high-risk individuals. This case presents an opportunity to highlight the importance of considering purulent pericarditis in patients previously diagnosed with AOSD. Serosal involvement accounts for 25% to 60% of AOSD patients, and is complicated by pericardial effusion.[9] This condition may offer a fertile ground for a secondary infection that arises directly from contiguous intrathoracic source or hematogenous spread.[1–3] Consequently, use of various tracking methods, such as repetitive blood cultures, routine chest radiographies, and serial echocardiographic evaluations, plays a critical role for timely detection of purulent pericarditis.

Prompt management of purulent pericarditis includes drainage of infected fluid along with empiric broad-spectrum antibiotics. Obtained pericardial fluid should be analyzed with Gram, acid-fast, fungal staining and microbial cultures. Antibiotic therapy can later be tailored according to culture and sensitivity results. Percutaneous pericardiocentesis is a safe and simple method to remove bacterial pathogens. Nevertheless, in cases of loculations with fibrin accumulation and dense adhesions, percutaneous drainage may be incomplete and ineffective that pericardial window or extensive pericardiectomy is usually necessary to achieve sufficient drainage[1–3] Guidelines from the European Society of Cardiology emphasize open surgical drainage as the preferable therapy.[1] Moreover, intrapericardial fibrinolysis with urokinase has known as an alternative to prevent recurrence and minimize the risk of constrictive pericarditis.[10] Despite aggressive treatment, purulent pericarditis is still inclined to develop subsequent constrictive pericarditis. In this situation, extensive pericardiectomy is evitable for releasing pericardial restraint. Although most patients had eventually undergone the extensive pericardiectomy, the prognosis is favorable in those with timely management.

In our report, we learn a novel association between AOSD and *Salmonella* purulent pericarditis. Immunodeficient patients with chronic pericardial effusion are predisposed to this fatal disease. Importantly, high clinical suspicion, early diagnosis, and prompt management can produce a better outcome in purulent pericarditis.

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