Anatomy & Physiology: Current Research
ISSN: 2161-0940

Wang et al., Anat Physiol 2017, 7:4
DOI: 10.4172/2161-0940.1000273

Case Report Open Access

Pericardial Effusion and Pericarditis Secondary to Adult Onset Still's Disease

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Received Date: July 15, 2017; **Accepted Date:** July 20, 2017; **Published Date:** July 27, 2017

Abstract

Adult-Onset Still's Disease is an uncommon systemic autoimmune disease with an incidence rate of 0.16 per 100,000. Various systems can be involved including musculoskeletal, cutaneous, hepatic, spleen, and lymphatic system. Cardiopulmonary involvement is found in 30-40% of the cases and can lead to significant morbidity and mortality. Prior cases of myocarditis and pericarditis leading to arrhythmias, heart failure and tamponade have been reported. We present a 24 year-old woman presenting with chest pain and dyspnea, and was found to have pericarditis in addition to pericardial effusion with tamponade physiology. She was successfully treated with drainage of pericardial effusion, corticosteroids and colchicine. The case illustrates the importance of early diagnosis and intervention in ensuring long-term recovery especially from the cardiac complications.

Keywords: Pericardial effusion; Pericarditis; Tamponade physiology; Adult onset still's disease

Introduction

Adult-Onset Still's Disease (ASD) is an uncommon systemic autoimmune disease with an incidence rate of 0.16 per 100,000 [1]. Although genetic and infectious causes have been proposed, the exact cause is still unknown. Suggested pathogens can be viral, Yersinia, and Mycoplasma [2]. Classical clinical presentation of the ASD includes fever, rash, arthritis, and myalgia occurring in 75-95% of the patients [3]. ASD is the diagnosis of exclusion, after infectious, autoimmune and malignancy have been excluded. Various systems can be involved including pharyngeal, liver, spleen, and lymphatic chains. Cardiopulmonary complications may occur and can lead to significant morbidity and mortality, such as pericarditis, pleural effusions, and transient pulmonary infiltrate, found in 30 to 40% of the patients diagnosed with ASD [3]. Rare case of myocarditis leading to arrhythmias, heart failure and tamponade has been reported [4]. Pericarditis is found in 16% in this patient population [3]. We present a 24 year-old woman presenting with chest pain and dyspnea, and found to have pericarditis, and pericardial effusion with tamponade physiology. She was successfully treated with corticosteroids and drainage of pericardial effusion.

Case Presentation

A 24-year-old female presents with 1-2 weeks worsening symmetric bilateral joint arthralgia, chest pain and shortness of breath with inspiration, skin rash, and odynophagia to both liquids and solids. She notes generalized fatigue and difficulty completing tasks at work, as well as intermittent fever. Her symptoms were most pronounced in the hands, knees, ankles and jaw. Her medical history is remarkable for gastro esophageal reflux disease, irritable bowel syndrome, depression, and asthma. On physical exam, the patient was tender to palpation bilaterally in hands, wrists, shoulders, knees and ankles and had decreased passive range of motion (ROM) with pain. Laboratory studies showed WBC 17,400/uL, significant elevation of ESR 107 mm/hr, CRP 221 mg/L, C3 192 mg/dL, ferritin 304 ng/ml, and alkaline phosphatase 131 IU/L. Rheumatoid factor (RF) and antinuclear antibody (ANA) were negative.

Initial echocardiogram showed a left ventricular ejection fraction (EF)>55%, and normal left ventricular diastolic function with mildly dilated left atrium. CT abdomen and pelvis showed hepatomegaly (23 cm), splenomegaly (14.8 cm), small pericardial effusion and small bilateral pleural effusions with adjacent atelectasis, trace peri-hepatic and pelvic free fluid (Figure 1).

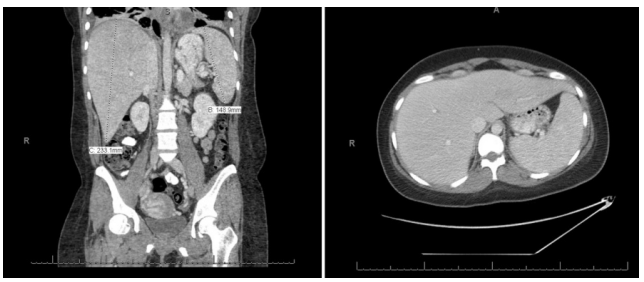

Figure 1: Initial CT of abdomen and pelvis demonstrating small bilateral pleural effusion and a very small pericardial effusion with no other cardiac abnormalities. There are enlargement of liver up to 23 cm and spleen 14.8 cm. There is a trace per hepatic and pelvic free fluid.

Repeat CT 3 days later showed mildly increased bilateral pleural effusions with associated compressive atelectasis, and a pericardial effusion increased from prior examination (Figure 2). A follow-up echocardiogram showed a large circumferential pericardial effusion

with tamponade physiology (Figure 3). She proceeded to have pericardiocentesis with pigtail catheter placement. Thoracic surgery was consulted for a pericardial biopsy, and intraoperatively, a thick adhesion and extensive inflammation was seen in pericardium and epicardium. She was followed with serial echocardiograms noting decreased pericardial effusions and pigtail catheter was removed. The pericardial biopsy showed fibrinous pericarditis with granulation tissue, reactive and hyperplastic mesothelial cells and fibrinous exudate, and mild and acute-on-chronic inflammation.

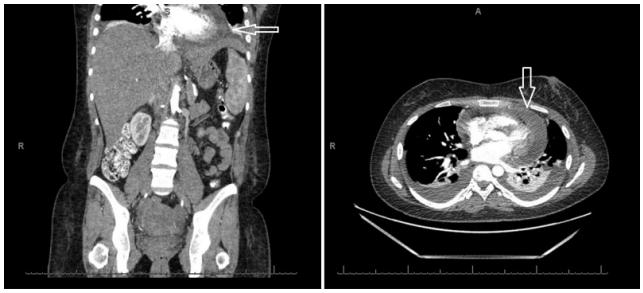

Figure 2: Subsequent CT of abdomen and pelvis 3 days later demonstrates an enlargement of the pericardial effusion (white arrow) with mild rim enhancement along the pericardium. Hepatosplenomegaly are again demonstrated.

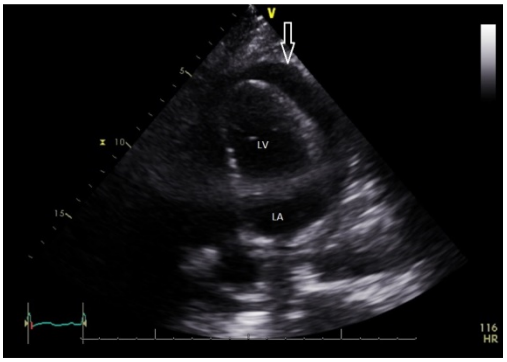

Figure 3: Echocardiogram showing large circumferential pericardial effusion with tamponade physiology. Left ventricular systolic function is normal greater than 55%.

She underwent extensive evaluation with rheumatology, neurology, infectious disease and hematology/oncology. A neurological workup included a CTA of the head and neck, and lumbar puncture, all unremarkable. The presence of splenomegaly prompted a hematology-oncology workup, including a bone marrow biopsy and test for porphyria that were unremarkable. With exclusion of other infectious, neurological, malignant, and other immunological disorders, patient was diagnosed with ASD by meeting Yamaguchi's diagnostic criteria. She was initiated on high-dose steroids intravenously and then transitioned to oral prednisone. She was placed on colchicine for pericarditis. Her symptoms were much improved at the time of discharge. She was discharged and following up with rheumatology for initiation of disease-modifying anti-rheumatic drug (DMARD).

Discussion

While numerous diagnostic criteria have been proposed, Yamaguchi criteria are known for having the highest sensitivity for ASD [5]. At least 5 of the proposed features including at least 2 of the major ones have to be met for the diagnosis. Although not as a part of Yamaguchi diagnostic criteria, laboratory abnormalities such as elevated ESR and CRP, increased serum ferritin level in 70% of cases [6], and abnormal liver function test with lactate dehydrogenase in 75% [7]. Our patient presented with arthralgia at least for 1-2 weeks, showed skin rash, leukocytosis above 10,000/μL, sore throat, hepatosplenomegaly, and negative ANA and RF. She also had elevated ferritin and alkaline phosphatase levels as well. Although a difficult diagnosis, with exclusion of other processes including malignancy, infectious both bacterial and viral, and potential other causes for her symptoms, her overall presentation was consistent with ASD.

The cardiac complications of ASD still remain to be investigated, not due to lack of knowledge on disease pathophysiology but rather making a timely diagnosis. It is a disease of exclusion and thus other more common etiologies such as infection and malignancy are usually pursued first. The lethal complications that may ensue from delayed diagnosis of ASD may lead to unnecessary mortality that may be avoided by early intervention. There is variety of cardiac manifestations to ASD. Pericarditis is found in approximately 10-14% of the patients, 20% of those patients develop pericardial effusion and may have cardiac tamponade [8]. Other uncommon complication is myocarditis in about 3% of ASD cases, and is associated with arrhythmia, atrioventricular (AV) block, and in some instances, overt heart failure [9]. Colina et al reports a patient with ASD presenting with macrophage activation syndrome and acute interstitial myocarditis leading to complete AV block. However, patient went into cardiogenic shock and passed several days later [10]. Carrilho-Ferreira et al describes a case of ASD in a young patient, who exhibited pericarditis with significant pericardial effusion along with tamponade physiology. The patient was treated with therapeutic pericardiocentesis and systemic corticosteroids, which resolved his symptoms [9]. Cardiac tamponade associated with Still's disease is found in less than 20 cases [11]. In a review of literatures on Still's disease and tamponade, 18 cases identified included 8 adults and rest of 10 in children. Among the 10 children, 4 died from the acute illness. Most received combined medical and surgical treatment [11]. It is critical to note that in many cases, pericardial effusion with or without tamponade physiology was the first manifestation of the disease process.

Treatment of the disease is based on the individual presentation and should be based on a multi-disciplined approach. In those with cardiac tamponade, therapeutic pericardial drainage is critical in ensuring hemodynamic stability and control of acute symptoms. With prompt recognition and control of the inflammatory process, positive outcomes are more likely with symptomatic improvement. Treatment of the ASD is based on severity of the disease. Mild condition is managed by non-steroidal anti-inflammatory agents. In those with moderate disease or fail NSAIDS, glucocorticoids are added and tapered as tolerated once disease is controlled. Other agents used may include anakinra, methotrexate in predominant joint symptoms, and TNF inhibitors. Non-biologic DMARDs have been used in ASD such as cyclophosphamide, hydroxychloroquine, azathioprine, intravenous immune globulin [12]. However, in those with severe disease with hepatic and cardiac involvement as in our patient, high-dose

intravenous pulse glucocorticoids followed by oral glucocorticoids and anakinra are recommended [2].

Our case demonstrates an uncommon manifestation of ASD, pericardial effusion with tamponade successfully treated with therapeutic drainage and corticosteroids. Control of the cardiac tamponade as well as treatment of underlying inflammatory process was crucial in this case. Our case illustrated the unpredictability and rapid progression of the disease including cardiac complications; patient developed large pericardial effusion in over 3-day course. Repeat imaging is critical in this case to detect and monitor for new manifestations of the ASD. Despite the challenges, prompt diagnosis of the ASD as well as treatment of potentially lethal presentations is critical to ensuring survival and positive outcome in this patient population.

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