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Cardiac Tamponade: A Life Threatening Complication of Adult still's Disease: Case Report and Review of Literature

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Abstract **Case Report**

Adult onset Still's disease is a systemic inflammatory disorder of unknown etiology characterized by the association of a high spiking fever, an evanescent skin rash, arthritis, and hyperleukocytosis. It is a rare disorder with potentially severe clinical features, including cardiac involvement. This systemic inflammatory disease of unknown origin should be considered in the differential diagnosis of pericarditis, with or without pericardial effusion. Cardiac tamponade is very rare sequelae that require an invasive approach, such as percutaneous or surgical pericardial drainage, in addition to the usual conservative therapy. We describe a case of a cardiac tamponade complicating Still's disease in a 27 year-old male, with a brief review of literature on this entity. This case reminds physicians to not neglect the potential of severe systemic inflammation to lead to fatal complications in this group of patients.

Keywords: Cardiac tamponade, Fever, Still's disease, Complications.

INTRODUCTION

Adult-onset Still's disease (AOSD) is an uncommon inflammatory illness that affects multiple organ systems [1]. It was originally described in children by George Still in 1896 [2] and Bywaters in 1971[3] who described a similar condition in adults. It is a disease of unknown origin that mainly affects patients between the ages of 16 and 35 years, characterized by high fever, arthralgia or arthritis, an evanescent salmon-colored skin rash, and leukocytosis with neutrophilia. It is more prevalent in females and has an estimated incidence of 0, 16 to 0,22 cases per 100,000 individuals. One of the extra-articular presentations is pericarditis which can be rarely complicated with cardiac tamponade [4]. Because there are no pathognomonic features, the diagnosis of AOSD is usually considered after the exclusion of other conditions—particularly infections, neoplasia (above all, lymphoma), and autoimmune disorders (most notably, vasculitis and polymyositis). Of the several proposed sets of diagnostic criteria, the Yamaguchi criteria. Of the several proposed sets of diagnostic criteria, the Yamaguchi criteria emerged as the most sensitive in a comparison by Masson and colleagues [5]. The diagnosis is established by the presence of 5 criteria, including at least 2 major criteria [6].

Table-1: Yamaguchi Diagnostic Criteria for Adult-Onset Still's Disease

Major criteria	• Fever >39°C, lasting 1 week or longer
	• Arthralgia or arthritis, lasting 2 weeks or longer
	• Typical rash
	• Leukocytosis >10 000/mm with >80% polymorphonuclear cells
Minor criteria	• Sore throat
	• Recent development of significant lymphadenopathy
	• Hepatomegaly or splenomegaly
	• Abnormal liver function tests
	• Negative tests for antinuclear antibody and rheumatoid factor (IgM)
Exclusion criteria	• Infections
	• Malignancies
	• Other rheumatic diseases
Five or more criteria are required with 2 or more being major criteria for diagnosis of AOSD	

Abbreviations: AOSD, adult-onset Still's disease; IgM, immunoglobulin M.

Nonetheless, the diagnosis lacks proof and is purely clinical. Close long-term follow-up, with repeated studies, can sometimes yield a different diagnosis [7].

CASE REPORT

A 27-year-old man was admitted with dyspnea and diffuse chest pain. He had been in his usual state of health until 20 days before his admission, when intermittent high fever developed with a temperature of 39 °C. Besides, he reported bilateral and symmetric arthralgia on the wrists and elbows, nausea and vomiting. The patient was previously healthy, had no history of drug abuse, and took no regular medication. He also had no pets and had not traveled recently.

Examination revealed tachycardia with heart rate of 120 bpm, respiratory rate of 25 cpm, and blood oxygen saturation of 98% at room air. The patient was hypotensive with blood pressure of 81/53 mmHg and muffled heart sound. Clinical examination also revealed jugular venous distension, splenomegaly with no lymphadenopathy and salmon-colored rash on the trunk and upper limbs. The electrocardiogram showed sinus tachycardia with electrical alternans, and cardiomegaly in the chest X-ray.

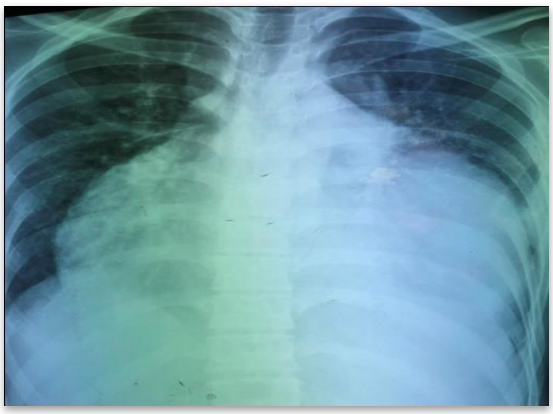

Fig-1: Chest X-ray with enlarged cardiac silhouette

Bedside transthoracic echocardiogram (TTE) showed circumferential pericardial effusion (maximal diameter, 22 mm) and collapse of the right atrium. Immediate pericardiocentesis was performed, draining 450 ml of clear yellow fluid.

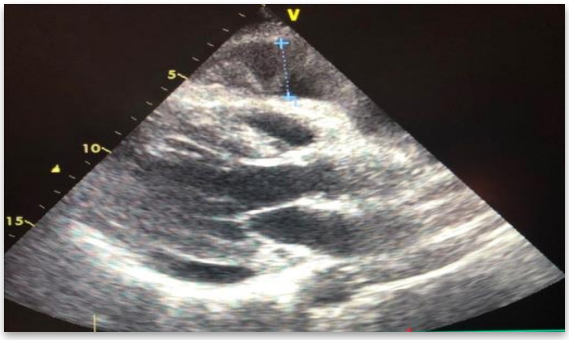

Fig-2: Parasternal long-axis view in echocardiography showing anterior pericardial effusion

Blood testing showed elevated inflammatory markers, with C-reactive protein of 50 mg/L, erythrocyte sedimentation rate of 120 mm/hr and hyperferritinemia of 1288 ng/ml with mildly elevated liver transaminase level. Complete blood count revealed leukocytosis with white of 21,000/uL with 73% of neutrophils, hemoglobin of 10 g/dl, and platelet count of 392,000/uL. Rheumatoid factor (RF) and anti-nuclear antibody (ANA) were negative. Blood and urine cultures were negative. No malignant cells were detected in pericardial fluid. Pericardial fluid was exudate with negative microbiologic examination; histologic analysis didn't detect malignant cells.

Computerized tomographic scans of the chest, abdomen, and pelvis showed mildpericardial effusion and splenomegaly. Bone marrow aspiration results showed normocellular bone marrow with normal maturation and differentiation cells and no blast cells.

The diagnosis of AOSD was established, based on Yamaguchi criteria. The patient was medicated with prednisone (0,5 mg/kg/d), with major clinical and laboratory improvement.

DISCUSSION

Adult onset Still's disease is a rare inflammatory disease that affects multiple organ systems. Patients between the ages of 16 and 35 are most affected [8,9]. Cardiac involvement is common and potentially severe. Pericarditis is observed in 10% to 40% of patients [10, 11] and is complicated, in about 20% of patients, by pericardial effusion [12]. Indeed pericarditis or its sequelae can be the initial manifestation of the disorder [13] but this does not appear to be associated with a worse prognosis [11]. However, as a rare complication, cardiac tamponade can be the initial presentation of AOSD.

Myocarditis is less prevalent (about 3% of all cases) [14] and can be complicated by complete atrioventricular block, tachyarrhythmia, heart failure, or cardiogenic shock [15]. Endocardial involvement is rare, and can present as non-infective endocarditis [16].

Given the frequency and severity of cardiac involvement in AOSD, all patients with this condition should undergo serial echocardiographic evaluation to exclude these complications.

The main cause of this illness remains unclear. Possible contributing factors are genetic predisposition, viral and bacterial infections, neoplasms, and inflammatory processes [17]. AOSD is diagnosed by applying Yamaguchi criteria and ruling out other infectious, neoplastic, and autoimmune causes [18]. Multiple cytokines like interleukins (IL-1 and IL-6) are involved in the pathogenesis of AOSD, making biologic drugs that target interleukins an evolving treatment option [18].

Nonsteroidal anti-inflammatory drugs are first-line treatment options. When necessary, corticosteroids, immunosuppressive drugs (particularly methotrexate and cyclophosphamide), or biologic drugs (tumor necrosis factor inhibitors, interleukin-1 receptor antagonists, and anti-B-cell antibodies) are used [9]. Typically, cardiac involvement responds well to conservative therapy [19]. However, percutaneous or surgical pericardial drainage is mandatory in cases of cardiac tamponade [20]. Adult-onset Still's disease has a good prognosis, with low mortality rates despite a recurrence rate of 10% per year during follow-up [21].

CONCLUSION

Given the frequency and the severity of cardiac involvement in adult onset Still's disease, all patients with this condition should undergo serial echocardiographic evaluation to exclude these complications.

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