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ACUTE SEVERE MITRAL AND AORTIC REGURGITATION ASSOCIATED WITH ADULT-ONSET STILL'S DISEASE

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ABSTRACT

Background: Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disease of unknown etiology usually affecting young adults. The primary clinical features are fever, rash, and arthritis or arthralgia. Cardiac involvement, especially valvular regurgitation, is less common. Only three cases of severe aortic and/or mitral regurgitation associated with AOSD have been reported.

Case Report: A 25-year-old man presented with fever evolving for 15 days, recent exertional dyspnea and diffuse arthromyalgia. On examination, the patient had a body temperature of 39.5°C and a pansystolic murmur at the mitral area radiating to the axilla and an early diastolic decrescendo murmur at the left sternal border with a modest intensity on cardiac auscultation. Laboratory tests revealed important leukocytosis with a predominance of neutrophils. Initial cardiac echocardiography ruled out infectious endocarditis, and revealed valvular dilated heart disease with preserved left ventricular ejection fraction with severe aortic and mitral regurgitation and a minimal tricuspid insufficiency. Pericarditis occurred secondarily. The evolution was marked by the onset of an acute left ventricular insufficiency and worsening of the pericarditis requiring a diuretic. The patient was diagnosed with AOSD using Yamaguchi's classification criteria. He was started on 1 mg/kg/d corticosteroid therapy, and scheduled for double valve replacement. A good outcome was achieved.

Conclusion: It is paramount to identify all the clinical manifestations of AOSD with a systematic screening for cardiac involvement especially when the patient presents with discrete symptoms such as exertional dyspnea. Thereby, early diagnosis and adequate treatment reduce the disease's critical complications.

KEYWORDS

Still's disease, mitral regurgitation, aortic regurgitation, pericarditis, valvulopathy

LEARNING POINTS

- Cardiac involvement in adult-onset Still's disease is not exceptional.
- The most frequent form is pericarditis.
- Valvular manifestations are rare. They can be inaugural and cause severe symptoms with acute heart failure, necessitating systematic screening and intensive treatments

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INTRODUCTION

Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disease of unknown etiology usually affecting young adults. It was first described by Eric Bywaters as a distinct clinical entity in the early 1970s[1]. It is an uncommon disease, with annual incidence ranging from 0.1 to 0.4 cases per 100,000 people in Europe[2]. It has bimodal age distribution; the first peak is between the ages of 15 and 25 and the second is between the ages of 36 and 46. About 70% of patients present with AOSD between 16 and 35 years of age[2]. The primary clinical features are fever, rash, and arthritis or arthralgia, observed in 75% to 95% of patients and they are associated most commonly with leukocytosis and hyperferritinemia[3]. Other common symptoms are pharyngitis, myalgia, lymphadenopathy and splenomegaly and, less often, pericarditis, pleurisy, abdominal pain and hepatomegaly[4]. Among cardiac manifestations, severe valvular involvement is rare. In the literature, only three cases of severe aortic and/or mitral regurgitation associated with AOSD have been reported[5-7]. We report a new case of acute severe mitral and aortic regurgitation in AOSD.

CASE DESCRIPTION

We report the case of a 25-year-old man, with no significant medical history, smoker and drug addict, who presented with fever evolving for 15 days, recent exertional dyspnea and diffuse arthromyalgia. On examination, the patient had a body temperature of 39.5°C, a pansystolic murmur at the mitral area radiating to the axilla and an early diastolic decrescendo murmur at the left sternal border with a modest intensity on cardiac auscultation, sibilant rales on pulmonary auscultation, a proximal muscle weakness of all four limbs, with no obvious infectious site.

Laboratory tests revealed leukocytosis (26,000 cells/µl) with a predominance of neutrophils (22500 elements/µl), neutrophils-to-lymphocytes ratio of 10.7, normochromic anemia 9 g/dl, rhabdomyolysis (creatine phosphokinase, 347 IU/l), hepatic cytolysis (aspartate aminotransferase, 90 IU/l; alanine aminotransferase, 106 IU/l), cholestasis (gamma glutamyl transferase, 201 IU/l), elevated C-reactive protein, 183 mg/l and elevated ferritinemia, 680 ng/ml.

Initial cardiac echocardiography ruled out infectious endocarditis and revealed valvular dilated heart disease with preserved left ventricular ejection fraction and homogeneous kinetics. The patient had severe aortic regurgitation, a moderately remodelled mitral valve with severe regurgitation and minimal tricuspid insufficiency without pulmonary hypertension. There was no pericarditis, initially.

Repeated blood cultures were negative. Testing for antinuclear antibodies and rheumatoid factor were negative. The antistreptolysin O level was <200 IU/ml.

The electroneuromyogram showed abnormalities suggesting myositis. Chest X-rays showed cardiomegaly and bilateral pulmonary edema. Computed tomography scan of the chest abdomen and pelvis showed pericarditis without any other abnormality, and ^{18}F-fluorodeoxyglucose positron emission tomography scan revealed hypermetabolic pericardial effusion, suggesting pericarditis, and the absence of any other pathological focus.

The patient's clinical course was marked by the onset of acute left ventricular insufficiency and worsening of the pericarditis requiring a diuretic.

On the basis of the clinical and laboratory findings, the patient was diagnosed with AOSD using Yamaguchi's classification criteria, with three major criteria, which were fever >39°C lasting for more than one week, arthralgia for more than two weeks, and leukocytosis (26,000 cells/µl) with 86% of neutrophils; and with two minor criteria, which were abnormal liver function tests and negative antinuclear antibodies and rheumatoid factor, and after exclusion of other etiologies. The patient was subsequently started on 1 mg/kg/d corticosteroid therapy and scheduled for double valve replacement. A good outcome was achieved, with disappearance of fever and arthromyalgia, improvement in general, hemodynamic and respiratory status, and regression of the biological inflammatory syndrome, cytolysis and hepatic cholestasis.

DISCUSSION

AOSD is a multi-systemic inflammatory disorder characterized by several distinguished manifestations and patients may present multiple complications, affecting the disease outcome. Cardiac involvement may affect all layers of the heart, including myocarditis, tamponade or endocarditis, with pericarditis being the most frequent manifestation[8]. Valvular involvement has been rarely reported, with only three cases in the literature[5-7].

Our case is the fourth one highlighting the potential severity of valvular manifestations in AOSD.

The three reported cases involved two women and a young man experiencing valvular regurgitation causing cardiogenic shock in two of them at initial presentation[5,6]. The only reported case with severe regurgitation of both the aortic and mitral valves was reported by Lee et al.[5]. In all these cases, dyspnea was the first sign leading to further investigations.

In the case reported by Çamlı Babayiğit et al.[7], the patient's valvular disease worsened 5 years after the diagnosis of AOSD, while she took anti-inflammatory drugs intermittently and according to the frequency of AOSD flares.

Bodard et al.[8] compared 28 patients with cardiac involvement to 68 AOSD patients with no cardiac manifestations and found that the first group was more likely to have refractory disease and to require biotherapy.

No specific guidelines for AOSD cardiac manifestations exist. Corticosteroid monotherapy or associated with a biologic must be initiated as early as possible in patients with life-threatening symptoms[9,10]. These

cardiac manifestations should indicate intensive immunosuppressive therapy to prevent cardiac shock[7].

It is paramount to identify all the clinical manifestations of AOSD with systematic screening for cardiac involvement, especially when the patient presents with discrete symptoms, such as exertional dyspnea, because. early diagnosis and adequate treatment reduce the risk of critical complications.

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