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Adult-Onset Still's Disease: A Rare Cause of Acute Severe Mitral Regurgitation

CASE REPORT

INTRODUCTION

Adult-onset Still's disease (AOSD) is a rare systemic inflammatory condition, typically presenting with fever, polyarthritis, and a transient maculopapular rash. Approximately 30% of AOSD cases involve cardiac complications. While pericarditis and myocarditis are the most commonly reported cardiac manifestations, valvular involvement is relatively rare.[1] Here, we present a case that underscores the potential for sudden onset and rapid progression of valve involvement in patients with AOSD, even with ongoing treatment. This highlights the importance of systematic cardiac screening for all patients diagnosed with AOSD.

CASE REPORT

A 62-year-old woman presented with worsening shortness of breath for 2 weeks. She was hospitalized with a suspected diagnosis of asthma due to complaints of cough and dyspnea. Despite undergoing 2 weeks of bronchodilator treatment, her shortness of breath and exertion intolerance persisted. She applied to our cardiology clinic.

She was diagnosed with AOSD 5 years ago. Methotrexate, prednisolone, and immunomodulatory drugs were intermittently administered based on the frequency of her attacks. The patient had been undergoing leflunomide treatment for 2 years when she was admitted to the cardiology clinic, and her last attack was on May 2023. Since Still's disease can also affect the heart, the patient had undergone echocardiography at regular intervals since the diagnosis. Her echocardiography was performed 2 months ago, showing a left ventricular ejection fraction (LVEF) value of 60% and mild mitral and tricuspid regurgitation. No abnormalities were noted regarding the valve appearance. Previous transthoracic echocardiography (TTE) examinations showed similar findings.

On admission, she was afebrile with a heart rate of 109 beats per minute and a respiratory rate of 24 breaths per minute. Her blood pressure was 110/52 mm Hg. Her oxygen saturation on room air was 86%. The patient appeared to be in moderate distress, exhibiting bilateral lung crackles and 1+ leg edema. She had a pansystolic murmur at the mitral area. Electrocardiogram revealed sinus tachycardia without signs of ischemia. The initial chest X-ray suggested pulmonary congestion and pleural effusion. Transthoracic echocardiography revealed a flail posterior mitral leaflet accompanied by severe mitral regurgitation, with no evidence of vegetation observed (Figure 1). The left ventricle was mildly dilated, with normal systolic function (LVEF: 60%). The right ventricle appeared normal in size and function, while tricuspid valve regurgitation was also noted to be severe. Based on the European Society of Cardiology 2023 diagnostic criteria for infective endocarditis, a thorough diagnostic process is followed, which includes major and minor criteria.[2] Transesophageal echocardiography (TEE) was performed quickly to exclude infective endocarditis and chordal rupture. Transesophageal echocardiography revealed a flail posterior mitral leaflet and an anteriorly directed jet of severe mitral regurgitation (Figure 2) with no sign of vegetation. The other major criterion for infective endocarditis is positive blood cultures. In our case, repeated blood cultures were taken, and all were negative. A definitive diagnosis

Ezgi Çamlı Babayiğit[1]
Emre Şener[1]
Erdi Babayiğit[2]

[1]*Department of Cardiology, Eskişehir City Hospital, Eskişehir, Türkiye*
[2]*Department of Cardiology, Eskişehir Osmangazi University, Eskişehir, Türkiye*

Corresponding author:
Ezgi Çamlı Babayiğit
✉ ezgi_iel@hotmail.com

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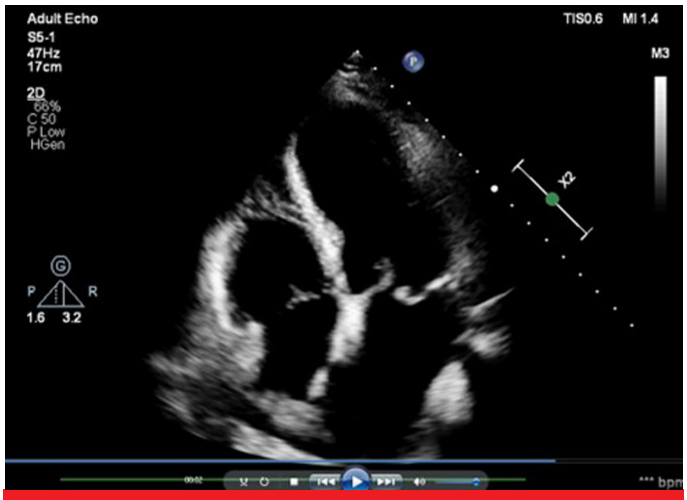

Figure 1. Transthoracic echocardiogram (apical 4 chamber view), showing a flail posterior mitral leaflet.

necessitates the fulfillment of either 2 major criteria, 1 major and 3 minor criteria, or 5 minor criteria. Conversely, a possible diagnosis can be established by meeting 1 major and 1 minor criterion or 3 minor criteria. Since neither definite nor possible diagnostic criteria were met, infective endocarditis was not considered a viable diagnosis in our case. Antibiotic therapy was not given in consultation with an infectious disease specialist.

In blood examination, white cell counts 5.6 10^3/L (4.2-10.8 10^3/L), erythrocyte sedimentation rate (ESR) 22 mm/h (0-20 mm/h), C-reactive protein (CRP) 10.5 mg/L (0-5 mg/L), Brain-natriuretic peptide (BNP) was 286.3 ng/L (0-100 ng/L). She was hospitalized and started on intravenous furosemide 40 mg q.i.d., and oral metoprolol tartrate 50 mg o.d. Since determining the etiology in heart valve patients is also important for treatment and follow-up, we also reviewed the possible causes of mitral insufficiency while the patient's treatment was continuing. Barlow disease and

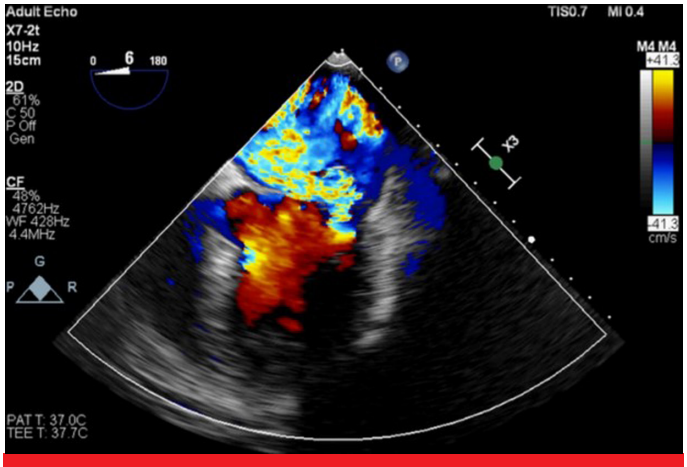

Figure 2. Transesophageal echocardiography (at 0°), showing a flail posterior mitral leaflet and an anteriorly directed jet of severe mitral regurgitation.

fibroelastic deficiency are 2 forms of degenerative mitral valve disease. Patients with Barlow disease are typically young (<60 years), more often women, and followed up for years to decades before interventions. Disease progression is slow. On the other hand, patients with fibroelastic deficiency are typically of middle to advanced age and present with a relatively short history of dyspnea, primarily due to chordal rupture. Our patient's profile did not fit either of them.

Since we could not explain the rapidly developing mitral valve insufficiency and prolapse of the patient with other possible etiologies, we thought that it might be related to Still's disease. After the patient's congestion was reduced, she was referred to the cardiovascular surgery unit for intervention on the mitral and tricuspid valves. Before the valve operation, angiography was performed, and no critical stenosis was found, and ischemic etiology was also ruled out. She underwent a mitral valve replacement (MVR) and tricuspid annuloplasty operation on February 13, 2024.

During follow-up, she remained stable without signs of dyspnea. Her international normalized ratio was maintained within the therapeutic range under warfarin treatment. She was followed by the rheumatology clinic, and her leflunomide treatment was continued. Follow-up TTE demonstrated a normally functioning mitral prosthesis, an LVEF of 60%, and mild tricuspid valve regurgitation. During the follow-up, informed consent was obtained from the patient, and a publication was planned as a case report.

DISCUSSION

Adult-onset Still's disease is a rare multisystemic, autoinflammatory disorder of unknown etiology.[3,4] Its incidence is estimated to range between 0.16 and 0.4 cases per 100 000 individuals, with a prevalence rate varying from 1 to 34 cases per 1 million people.[5] The major clinical features include intermittent spiking fever, transient salmon-colored maculopapular rash, polyarthralgia, sore throat, neutrophilia, lymphadenopathy, liver dysfunction, hepatosplenomegaly, increased ESR, and hyperferritinemia.[6] There are various sets of classification criteria for diagnosis, with Yamaguchi's and Fautrel's criteria being the most used.[7,8]

In a study by Bodard et al[9] involving 28 patients diagnosed with AOSD, the rate of cardiac involvement was found to be 29%. This included pericarditis (61%, n = 17), tamponades (18%, n = 5), myocarditis (18%, n = 5), and 1 case of non-infectious endocarditis (4%). The findings of cardiac involvement were present before or during the diagnostic stage in 89% of patients, before any specific treatment was initiated. Zenagui and De Coninck[10] previously described a case involving a 38-year-old man diagnosed with non-infectious aortic valve endocarditis associated with AOSD who was successfully treated with 1 mg/kg prednisolone. Therefore, especially in newly diagnosed patients, early initiation of immunosuppressive therapy should be considered to control inflammation and prevent hemodynamic instability. In this

case report, our patient had already been diagnosed with AOSD and was undergoing treatment. However, a recent echocardiography performed just 2 months ago revealed no severe pathology.

In a case report by Lee et al[11] in 2005, a 55-year-old woman with AOSD complicated by acute severe mitral and aortic insufficiency was presented. The patient underwent MVR and Bentall's operation. Like our case, the patient's mitral regurgitation, initially mild at the time of diagnosis, progressed to severe within 1 month, and severe aortic insufficiency developed as well. This situation highlights the potential for acute and severe valve disease to develop rapidly in AOSD patients, even in the absence of prior significant cardiac abnormalities.

CONCLUSION

In summary, when findings suggestive of cardiac involvement are present, echocardiography should be repeated more frequently. If suspicion persists, further examination should be conducted using TEE and other imaging techniques. Early detection and monitoring of cardiac complications are crucial for timely intervention and management in AOSD patients.

We did not use any artificial intelligence (AI)– assisted technologies (such as Large Language Models [LLMs], chatbots, or image creators) in the production of submitted work.

Informed Consent: Informed consent was obtained from the patient.

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