

Heavy Calcification of Left Atrium and Calcific Mass on Mitral Valve: An Unusual Presentation of Ankylosing Spondylitis

Sol Atriyumda Ciddi Kalsifikasyon ve Mitral Kapak Üzerinde Kalsifik Kitle: Ankilozan Spondilitin Sıra Dışı Bir Prezantasyonu

ABSTRACT

Ankylosing spondylitis is a complex chronic disease that starts from the spine and sacroiliac joint, causes joint stiffness, and affects the quality of life. Aortic dilatation, aortic valve insufficiency, and heart block due to conduction system involvement are the leading accompanying cardiac pathologies. In this case report, we aimed to show a rare cardiac involvement of left atrial calcification, pericardial involvement, and formation of a calcified inflammatory mass that might be misdiagnosed as vegetation on the mitral valve in a late-stage ankylosing spondylitis patient, by using multimodality imaging and histopathological diagnosis.

Keywords: Ankylosing spondylitis, cardiovascular imaging, valvular disease

ÖZET

Ankilozan Spondilit (AS), omurga ve sakroiliak eklemlerden başlayan, eklem sertliğine neden olan ve yaşam kalitesini etkileyen karmaşık kronik bir hastalıktır. Aort dilatasyonu, aort kapak yetersizliği ve iletim sistemi tutulumuna bağlı kalp bloğu eşlik eden kardiyak patolojilerin başında gelir. Bu olgu sunumunda, geç evre bir ankilozan spondilit hastasında nadir görülen bir kalp tutulumu olan sol atriyal kalsifikasyon, perikardiyal tutulum ve mitral kapakta vejetasyon olarak yanlış tanılanabilecek kalsifiye enflamatuvar kitle oluşumunu, multimodalite görüntüleme ve histopatolojik tanı yöntemleri ile göstermeyi amaçladık.

Anahtar Kelimeler: Ankilozan Spondilit, kapak hastalığı, kardiyovasküler görüntüleme

Ankylosing spondylitis (AS) is a complex chronic disease that causes stiffness in joints starting from the spine and sacroiliac joint. The main cardiac pathologies that occur during the course of the disease are aortic dilatation, aortic valve insufficiency,

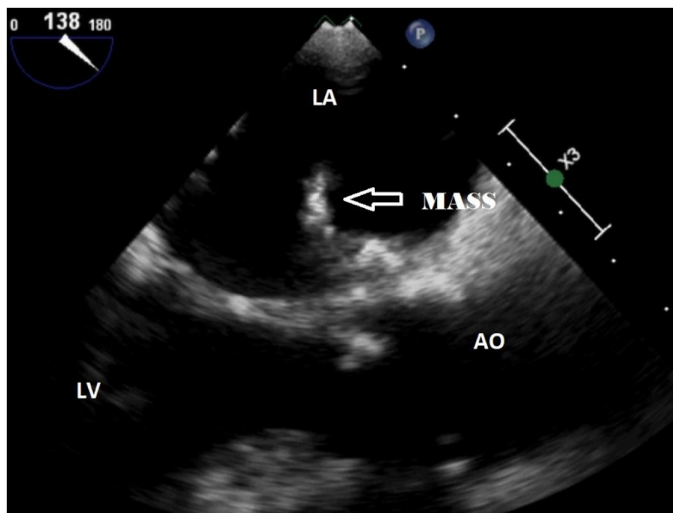


Figure 1. Calcific mass on mitral valve. LA, left atrium; LV, left ventricle; AO, aorta.

CASE REPORT OLGU SUNUMU

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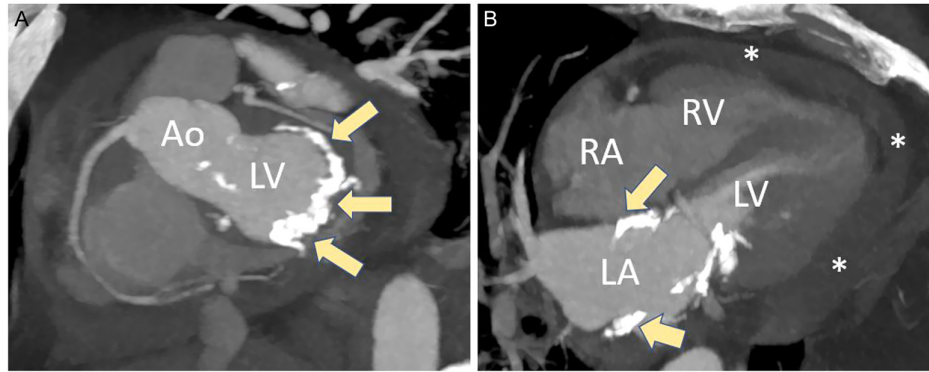


Figure 2. (A).The short-axis contrast-enhanced cardiac CT image through the mitral annulus level depicts mitral annulus calcification (arrows). **(B).** The 4 chamber CT image shows calcific plaques on the left atrial wall (arrows). Notice the moderate pericardial effusion. CT, computed tomography.

and heart blocks due to conduction system involvement. Other cardiac manifestations are mitral valve insufficiency, pericarditis, and cardiomyopathy. In this report, we aimed to discuss a very rare case with a vegetation-like mass on the mitral valve and heavy calcification of left atrium due to AS.

Case Report

A 57-year-old male patient, who was suffering from dyspnea, was referred to our clinic with an initial diagnosis of infective endocarditis with elevated C-reactive protein (113 mg/dL), increased sedimentation rate (65 mm/h), and troponin I levels accompanying a suspicious mass on mitral valve on echocardiographic examination. The patient was diagnosed with ankylosing spondylitis 15 years ago, but routine controls had not been performed for years. The patient had a kyphotic appearance with severe movement restriction and when questioned, he stated that he had not received any treatment for his disease. Indeed, our echocardiographic examination revealed a mobile mass at the tip of the anterior mitral leaflet with suspicion of vegetation.

On transesophageal echocardiography (TEE) examination, a mobile mass with a size of 2.18 × 0.53 cm, starting from the anterior annulus of the mitral valve and extending to the interatrial septum, with hypo-hyperechogenic areas and acoustic shadowing, was detected (Figure 1, Supplementary Videos 1 and 2)*. Concomitantly, computed tomography (CT) images revealed pericardial involvement with moderate pericardial effusion, as well as irregular thickening and calcification were observed in all walls of the left atrium, more prominent in the anterior wall (Figure 2).

Although the infective endocarditis team did not consider infective endocarditis at forefront, fluorodeoxyglucose (FDG)-positron emission tomography (PET) was performed to exclude that diagnosis completely and at this imaging, no involvement compatible with the infection was found on mitral valve.

Since TEE images suggested a calcified mass rather than an active vegetation (Figure 3 and Supplementary Video 3) and no bacterial reproduction was observed in blood cultures, diagnosis of infective endocarditis became less likely. In addition, other common pathologies that might cause calcification, such as chronic kidney disease, hyperparathyroidism, tuberculosis or sarcoidosis, were excluded.

As the mass was larger than 1 cm, it was decided via the heart team to operate the patient to prevent the risk of embolization. Complete mass excision was performed. Pathological examination of the mass revealed signs of active chronic non-specific inflammation, without any pattern of vegetation. On the basis of these findings, the inflammatory mass formation on the mitral valve and severe involvement of the left atrium indicated chronic inflammation in the patient with advanced stage ankylosing spondylitis. After the surgery, the patient was referred to a rheumatologist for follow-up and medical treatment.

Informed consent obtained from the patient.

Discussion

Ankylosing spondylitis is a chronic, inflammatory, and rheumatic disease, mainly affecting the sacroiliac joint and spine. The frequency of cardiovascular involvement in AS varies between 10% and 30%.¹

As a result of aortitis, which is the main cardiac involvement of AS, aortic valve insufficiency and aortic root enlargement develop.^{2,3} Aortic involvement extends toward the interventricular septum and this leads to the development of conduction disorders.³

Typical changes in the heart valves are fibrosis and thickening and retraction of the valves in the early period.⁴ These changes cause progressive insufficiency of the valve.

Mitral valve insufficiency in AS can occur in 2 ways; the first is due to left ventricular dilatation secondary to severe aortic insufficiency and the second way is the extension of subaortic fibrotic thick tissue to the mitral valve anterior leaflet.⁵

ABBREVIATIONS

Ao	Aorta
AS	Ankylosing spondylitis
CT	Computed tomography
FDG PET	Fluorodeoxyglucose positron emission tomography
LA	Left atrium
LV	Left ventricle
RA	Right atrium
RV	Right ventricle
TEE	Transesophageal echocardiography
3D	Three dimensional

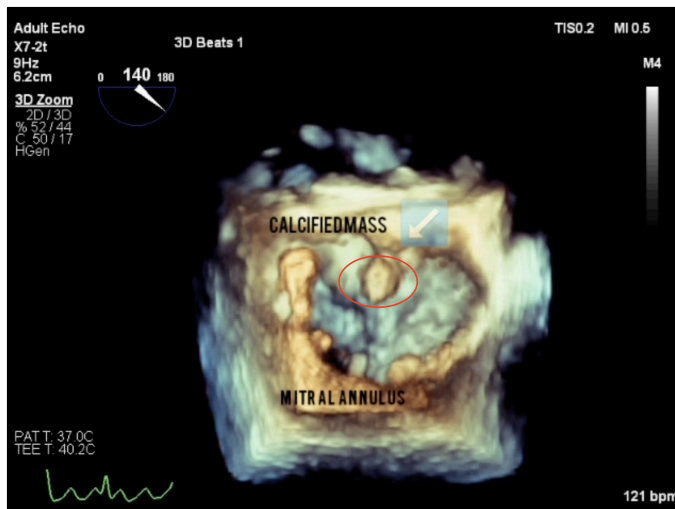


Figure 3. Three-dimensional TEE image of the calcified mass, on top of mitral valve (marked with red circle).

A case of mitral stenosis due to AS has been reported in the literature and it has been speculated that subaortic fibrotic expansion has led to fibrotic thickening of the mitral valve. However, it does not seem possible to rule out acute rheumatic fever with anamnesis in this patient.⁶

In a case reported by Hoppmann et al⁵ in the literature, vegetation on the diffuse fibrotic and thickened aortic valve was reported in a patient with ankylosing spondylitis. Although the main pathology of valve involvement in ankylosing spondylitis is diffuse thickening, infective endocarditis should always be kept in mind when a mass is detected on the valve.⁷

Our patient was first investigated for infective endocarditis because of the presence of a mobile large mass on the mitral valve. However, this diagnosis was excluded with the results of laboratory tests, negative blood cultures, and multimodality imaging. Widespread calcification of the left atrium on both TEE and CT suggests that it may be associated with the advanced stage of the disease. Interestingly, our patient had mitral valve and left atrial involvement without a serious involvement of the aortic valve, such as aortic regurgitation or aortic root dilatation. Cardiac CT revealed the extent of calcification (Figure 2). Histopathological evaluation revealed fibrotic calcification and hyalinization due to AS, and consequently, vegetation and malignant mass were excluded.

Conclusion

In this case report, we presented a calcified inflammatory mass on the mitral valve, which might be misdiagnosed as

vegetation, and concomitant left atrial calcification, and pericardial involvement in a patient with advanced stage ankylosing spondylitis.

Informed Consent: Written informed consent was obtained from all the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – S.T.K., G.B.G.; Design – S.T.K., G.B.G.; Supervision – G.B.G.; Data Collection and/or Processing – S.T.K., G.B.G., B.O., Ç.T., S.Ö.; Analysis and/or Interpretation – S.T.K., G.B.G.; Literature Search – S.T.K.; Writing Manuscript – S.T.K., G.B.G.; Critical Review – G.B.G.

Declaration of Interests: The authors have no conflict of interest to declare.

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*Supplementary video files associated with this article can be found in the online version of the journal.

Supplementary Video 1: Mobile mass starting from the anterior annulus of the mitral valve. LA, left atrium; LV, left ventricle; AA, ascending aorta.

Supplementary Video 2: Mobile mass starting from the anterior annulus of the mitral valve, biplane view. LA, left atrium; LV, left ventricle; AA, ascending aorta.

Supplementary Video 3: 3D image of the calcified mass, on top of mitral valve.

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