CASE REPORT

Mitral annular calcification-related calcified amorphous tumor in a patient with normal renal functions: A case report

Böbrek fonksiyonu normal olan hastada mitral anüler kalsifikasyonla ilişkili kalsifiye amorf tümör: Bir olgu sunumu

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Summary– A calcified amorphous tumor (CAT) of the heart is a non-neoplastic mass, characterized by nodules of calcium on a background of amorphous fibrinous material. Mitral annular calcification (MAC) is one of the conditions most frequently associated with CAT. Most MAC-related CAT cases are reported in end-stage renal disease (ESRD) patients. This report is a description of a patient with normal renal function who was incidentally diagnosed with a CAT arising from MAC. An asymptomatic 43-year-old man with normal renal function underwent echocardiography. The echocardiography results revealed a highly mobile hyperechoic mass attached to the atrial side of the mitral annulus superimposed on a heavily calcified mitral annulus. Cardiac surgery and histopathological evaluation confirmed a diagnosis of MAC-related CAT. While MAC-related CAT is usually associated with ESRD patients, the precise etiology and management strategy of this benign tumor remain unclear. The rare incidence of this tumor in patients with normal renal function highlights the importance of further research of the pathology of this uncommon condition.

Özet– Kalbin kalsifiye amorf tümörü (KAT) neoplazik olmayan bir kitle olup amorf fibrinli materiyal üstünde kalsiyum nodülleri ile karakterizedir. Mitral anüler kalsifikasyon (MAK) en büyük sıklıkla KAT ile ilişkili durumlardan biridir. MAK ile ilişkili KAT olgularının çoğu son dönem böbrek hastalarında (SDBH) bildirilmiştir. Bu olgu raporu rastlantısal olarak MAK'dan kaynaklanan KAT tanısı alan böbrek fonksiyonu normal bir hastayı tanımlamaktadır. Böbrek fonksiyonu normal 43 yaşında semptomsuz bir erkek hastaya ekokardiyografi uygulandı. Ekokardiyografi sonuçları çok mobil, mitral kapağın atriyal yüzüne yapışık, son derece kalsifiye mitral kapağı örten hiperekoik bir kitleyi ortaya çıkardı. Kalp cerrahisi ve histopatolojik değerlendirme MAK ile ilişkili KAT tanısını doğruladı. MAK ile ilişkili KAT genellikle SDBH olanlarla ilişkilendirilmesine rağmen bu iyi huylu tümörün kesin etiyoloji ve sağaltım stratejisi hâlâ net değildir. Böbrek fonksiyonları normal hastalarda bu tümörün seyrek görülmesi bu olağandışı durumun patolojisini daha fazla araştırmanın önemini vurgulamaktadır.

A calcified amorphous tumor (CAT) of the heart is a non-neoplastic mass characterized by nodules of calcium on a background of amorphous fibrinous material. Mitral annular calcification (MAC) is one of the conditions most frequently associated with CAT.[1] Due to a high risk of embolization, MAC-related CAT should be considered unique in comparison with other instances of CAT.[2] Most MAC-related CAT cases are reported in end-stage renal disease

Abbreviations:

CAT	*Calcified amorphous tumor*
ESRD	*End-stage renal disease*
MAC	*Mitral annular calcification*

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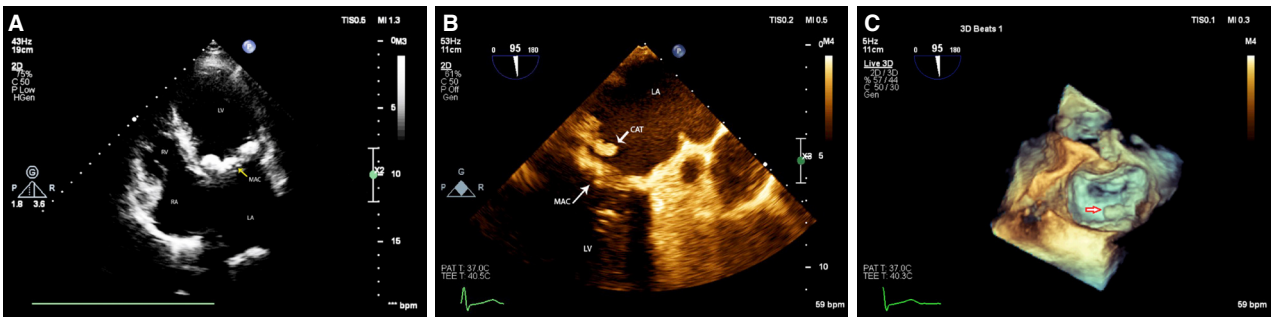

Figure 1. **(A)** Transthoracic echocardiography image in apical 4-chamber view shows a bulky mitral annular calcification (MAC; arrow). **(B)** Transesophageal echocardiography image in the mid-esophageal 2-chamber view shows a bulky mitral annular calcification (MAC), associated with a calcified mass on the atrial side of the mitral annulus, suggestive of a calcified amorphous tumor (CAT). **(C)** Three-dimensional transesophageal echocardiography image of the mitral valve in en-face view shows a large hyperechoic mass (arrow) suggesting a calcified amorphous tumor. LA: Left atrium; LV: left ventricle; RA: Right atrium; RV: Right ventricle.

(ESRD) patients; however, there are exceptions, including the present case, a patient with normal kidney function.[3,4]

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A 43-year-old man presented due to a heart murmur found during a regular medical examination. The patient was asymptomatic, without limitation of effort. There was no apparent medical history of diabetes mellitus, hypertension, hyperlipidemia, cigarette smoking, or thromboembolism. The physical examination revealed an afebrile patient with normal vital signs; however, cardiac auscultation depicted a dominant mid-systolic harsh murmur heard at the second right intercostal space concomitant with a low-pitched rumbling murmur detected at the apex.

Transthoracic and 3-dimensional transesophageal echocardiography showed a highly mobile hyperechoic mass (2.0x0.6 cm) attached to the atrial side of the mitral annulus superimposed on a heavily calcified mitral annulus, resulting in severe mitral stenosis and moderate mitral regurgitation (Fig. 1a-c, Videos 1–3*). A calcified aortic valve and moderate aortic stenosis were also observed using echocardiography (Video 4*). All of the laboratory test results, including renal, liver, and thyroid function, and levels of

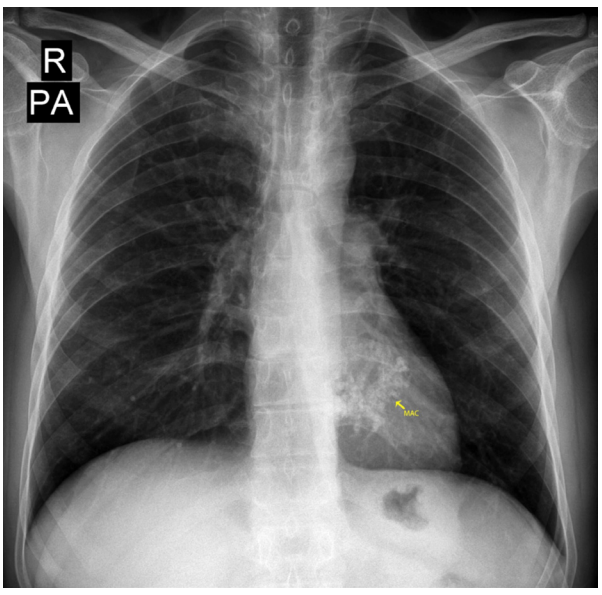

Figure 2. A chest X-ray showed a normal cardiothoracic ratio with a calcified dense opacity (arrow) suggestive of bulky mitral annular calcification (MAC).

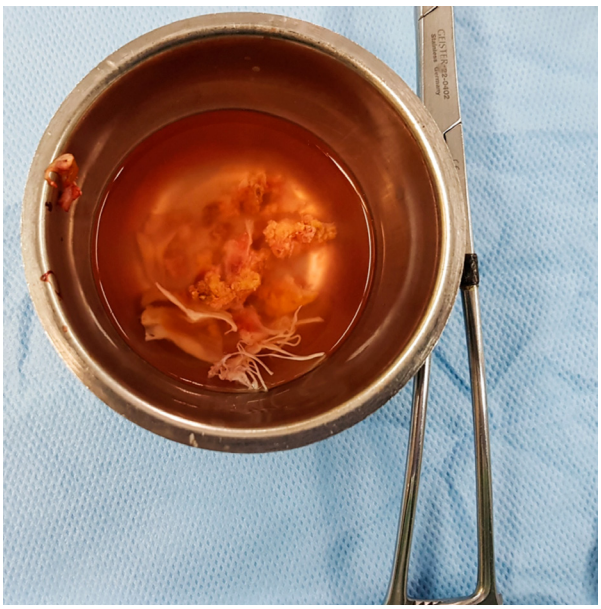

Figure 3. The mitral valve, the calcified mass, and a large calcification of the mitral annulus were resected.

calcium, phosphorus, parathyroid hormone, and uric acid, were within the normal range. Moreover, the C-reactive protein level and erythrocyte sedimentation rate were also within normal limits. A chest X-ray illustrated a normal cardiothoracic ratio with a calcified dense opacity prominently embedded in the mitral annulus site (Fig. 2). An electrocardiogram demonstrated an inverted T-wave in leads III and an aVF with no remarkable ST-depression. Coronary angiography revealed normal epicardial coronary arteries. As the mass was highly mobile and prone to embolization, cardiac surgery was performed, and intraoperative findings indicated heavy calcification of the mitral annulus extending to the left ventricular endocardial surface. The calcified mass on the atrial surface was resected, and both the mitral and aortic valves were replaced with mechanical prostheses (Fig. 3).

A histopathological evaluation of the resected mass illustrated dense calcium deposits surrounded by a fibrinous background, consistent with a calcified amorphous tumor. The patient recovered after 7 days and was discharged from the hospital without any complications.

DISCUSSION

MAC-related CAT represents a subgroup of CAT, and is usually associated with ESRD.[2,4–8] Our patient is one of a few MAC-related CAT patients in the literature who had normal renal function. The exact pathogenesis of cardiac CAT is not well-known, abnormalities in calcium-phosphorus metabolism and certain genes implicated in valvular calcification may contribute to the pathogenesis of this rare tumor.[6,9] The growth rate of a CAT is largely unknown, but may be relatively fast, especially in cases of MAC-related CAT.[1] Although CAT may be found incidentally, the most common presenting symptoms are dyspnea, embolic events, and syncope.[1] CAT has been introduced as a possible clue to the bridge between MAC and stroke. The best management strategy for this tumor remains a debate,[10] but surgical resection seems to be a good option due to the high probability of mass embolism.

The precise etiology and management strategy for this benign tumor remains unclear. To the best of our knowledge, our patient is the fourth case reported in a patient with normal renal function. This highlights the importance of further research on the pathology of this uncommon condition.

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

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Keywords: Calcified amorphous tumor; echocardiography; end-stage renal disease; mitral annular calcification.

Anahtar sözcükler: Kalsifiye amorf tümör; ekokardiyografi; son dönem böbrek hastalığı; mitral kapak kalsifikasyonu.