Atypical mitral annular calcification mimicking an intracardiac tumor

İntrakardiyak tümörü andıran mitral anülüs kalsifikasyonu

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Mitral annular calcification (MAC) is a common echocardiographic finding in geriatric population and is usually seen in the posterior atrioventricular groove. In general, MAC does not affect mitral valve functions. Intramyocardial extension is rare. A 67-year-old woman presented with shortness of breath and palpitation. She had a history of hypertension. Physical examination was unremarkable except for arrhythmia and raised blood pressure. The electrocardiogram showed atrial fibrillation with a ventricular rate of 80/min and an incomplete right bundle branch block. A chest radiogram showed a moderately enlarged heart silhouette, transthoracic echocardiography demonstrated a round echogenic mass in the posterior periannular region between the base of the posterior mitral leaflet and contiguous left ventricular wall, suggestive of a cardiac tumor. There were no findings of mitral stenosis or regurgitation in 2D, color and spectral Doppler imaging. Unenhanced cardiac magnetic resonance imaging (MRI) revealed involvement of the posterior mitral annulus and posterobasal myocardial wall, and calcified nature of the mass with no signal intensity. Contrast-enhanced MRI showed no perfusion of the mass. The mass was diagnosed as MAC extending from the posterior mitral annulus to the adjacent myocardial wall.

Key words: Calcinosis; heart valve diseases/etiology; magnetic resonance imaging; mitral valve/pathology.

Mitral annular calcification (MAC), a degenerative process of the fibrous support structure of the mitral apparatus, is a common echocardiographic finding in geriatric population especially in females.^[1] It is associated with known atherosclerotic risk factors such as age, diabetes mellitus, hypertension, and hyperlipidemia and is usually seen in the posterior atrioventricular groove, sometimes extending to the

Mitral anülüs kalsifikasyonu özelikle yaşlı kişilerde görülen oldukça yaygın bir ekokardiyografik bulgudur. Sıklıkla posterior atrioventriküler olukta görülür. Genellikle mitral kapak fonksiyonlarında önemli bir anormalliğe yol açmaz. İntramiyokardiyal uzanımı nadirdir. Öyküsünde hipertansiyon tanısı olan 67 yaşında kadın hasta nefes darlığı ve carpıntı yakınmalarıyla başvurdu. Fizik muayenesinde aritmi ve artmış kan basıncı dışında özellik yoktu. Elektrokardiyogramda, ventriküler hızı 80/dk olan atriyal fibrilasyon ve inkomplet sağ dal bloku izlendi. Göğüs grafisinde orta derecede genişlemiş kalp görüntüsü izlendi; transtorasik ekokardiyografide, posterior perianüler bölgede, posterior mitral yaprakçık tabanı ile sol ventrikül duvarı arasında, kalp tümörünü andıran yuvarlak ekojenik bir kitleye rastlandı. İkiboyutlu, renkli ve spektral Doppler görüntülemede mitral darlık ya da yetersizlik bulgusu yoktu. Kontrastsız kardiyak manyetik rezonans görüntülemede (MRG), sinyal yoğunluğu göstermeyen kalsifiye kitlenin posterior mitral anülüsü ve posterobazal miyokard duvarını tuttuğu görüldü. Kontrastlı MRG'de perfüzyon izlenmedi. Tanı, posterior mitral anülüsten yanındaki miyokard duvarına uzanım gösteren mitral anülüs kalsifikasyonu şeklinde kondu.

Anahtar sözcükler: Kalsinoz; kalp kapağı hastalığı/etyoloji; manyetik rezonans görüntüleme; mitral kapağı/patoloji.

mitral annulus and the base of the mitral leaflets. However, unusual and extensive presentation of MAC has been reported due to primary and secondary hyperparathyroidism which is frequently related to chronic renal failure.^[2] In this report, we presented a case in which unusual and extensive MAC was encountered, simulating a cardiac tumor.

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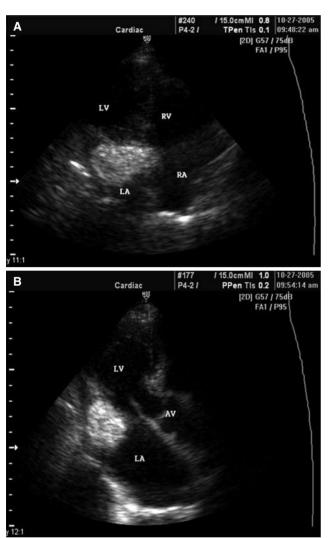


Figure 1. Modified apical (A) four-chamber and (B) fivechamber views showing a round echogenic mass.

CASE REPORT

A 67-year-old woman was admitted to our institution with shortness of breath and palpitation. She had a history of hypertension. Her physical examination was unremarkable except for arrhythmia and raised blood pressure. The ECG showed atrial fibrillation with a ventricular rate of 80/min and an incomplete right bundle branch block. Complete blood count, electrolytes, and creatinine were within normal limits. Serum parathormone level was 36 pg/ml (normal range: 15-65 pg/ml). Upon observation of a moderately enlarged heart silhouette on her chest radiogram, transthoracic echocardiographic (TTE) examination was performed, which demonstrated a round echogenic mass in the posterior periannular region between the base of the posterior mitral leaflet and contiguous left ventricular wall (Fig. 1). There were no findings

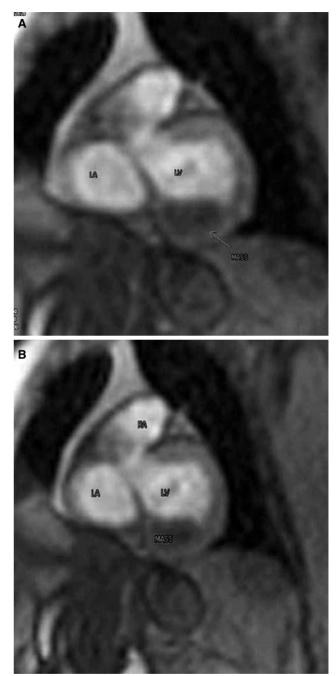


Figure 2. (A) Unenhanced MRI view showing involvement of the posterior mitral annulus and adjacent myocardium. (B) Contrastenhanced MRI showing no perfusion of the mass.

of mitral stenosis or regurgitation in two-dimensional, color and spectral Doppler imaging. Cardiac magnetic resonance imaging (MRI) was planned for further evaluation and differential diagnosis of the mass. Unenhanced cardiac MRI frames revealed involvement of the posterior mitral annulus and posterobasal myocardial wall, and calcified nature of the mass, as demonstrated by the lack of any signal intensity on all sequences. In enhanced MRI, neither the first-pass acquisition frame nor delayed contrast-enhanced acquisition frames showed perfusion of the mass (Fig. 2). The mass was diagnosed as MAC extending from the posterior mitral annulus to the adjacent myocardial wall.

DISCUSSION

Mitral annular calcification is a chronic and degenerative process involving the fibrous ring of the mitral apparatus.^[1] It occurs more often in women and in the elderly. In general, MAC does not affect mitral valve functions; however, it can rarely cause mitral regurgitation and less commonly mitral stenosis.^[3] In extreme cases, calcification can grow posteriorly into the ventricular myocardium. However, intramyocardial extension is rare. Several reports have shown an association between extensive, atypical MAC and abnormal calcium metabolism.^[2,4] A relationship was demonstrated previously between chronic renal failure (CRF)-associated hypercalcemia and accelerated deposition of calcium in the mitral annulus, leaflets, and in individual myocardial fibers.^[2] The frequency of MAC has been reported as 50% in patients with CRF, receiving hemodialysis for 10 years or longer.^[5] An unusually intense, mass-like involvement and intramyocardial extension has also been described in these patients. Mitral annular abscess,^[6] lipomatosis of the atrioventricular groove, and enlarged lymph nodes and tumors should be considered in the differential diagnosis of round echogenic structures adjacent to the left atrioventricular groove.[7] However, intramyocardial calcification of the left ventricle has a wide differential diagnosis. The most obvious cause for myocardial calcification is scar formation following a myocardial infarct. Intramural cardiac myxoma,^[8] primary cardiac osteosarcoma and metastatic osteosarcoma,^[9] hydatid cyst,^[10] teratoma, angioma, and rhabdomyoma are other unusual causes of atypical calcifications or calcified cardiac masses. Although symptoms and echocardiographic characteristics can provide adequate data in the majority of cases, MRI may provide additional information for the differential diagnosis. The mass may exhibit a calcified nature with high signal intensity. This case illustrates the potential role of cardiac MR imaging in the evaluation

and differential diagnosis of atypical and extensive MAC mimicking a cardiac tumor.

REFERENCES

- Boon A, Cheriex E, Lodder J, Kessels F. Cardiac valve calcification: characteristics of patients with calcification of the mitral annulus or aortic valve. Heart 1997; 78:472-4.
- Depace NL, Rohrer AH, Kotler MN, Brezin JH, Parry WR. Rapidly progressing, massive mitral annular calcification. Occurrence in a patient with chronic renal failure. Arch Intern Med 1981;141:1663-5.
- Aronow WS, Kronzon I. Correlation of prevalence and severity of mitral regurgitation and mitral stenosis determined by Doppler echocardiography with physical signs of mitral regurgitation and mitral stenosis in 100 patients aged 62 to 100 years with mitral anular calcium. Am J Cardiol 1987;60:1189-90.
- Bittrick J, D'Cruz IA, Wall BM, Mansour N, Mangold T. Differences and similarities between patients with and without end-stage renal disease, with regard to location of intracardiac calcification. Echocardiography 2002;19:1-6.
- Al-Absi AI, Wall BM, Aslam N, Mangold TA, Lamar KD, Wan JY, et al. Predictors of mortality in end-stage renal disease patients with mitral annulus calcification. Am J Med Sci 2006;331:124-30.
- Daniel WG, Mugge A, Martin RP, Lindert O, Hausmann D, Nonnast-Daniel B, et al. Improvement in the diagnosis of abscesses associated with endocarditis by transesophageal echocardiography. N Engl J Med 1991; 324:795-800.
- Zuber M, Oechslin E, Jenni R. Echogenic structures in the left atrioventricular groove: diagnostic pitfalls. J Am Soc Echocardiogr 1998;11:381-6.
- Rendon F, Agosti J, Llorente A, Rodrigo D, Montes K. Intramural cardiac myxoma in left ventricular wall: an unusual location. Asian Cardiovasc Thorac Ann 2002; 10:170-2.
- Nowrangi SK, Ammash NM, Edwards WD, Breen JF, Edmonson J. Calcified left ventricular mass: unusual clinical, echocardiographic, and computed tomographic findings of primary cardiac osteosarcoma. Mayo Clin Proc 2000;75:743-7.
- Ben-Hamda K, Maatouk F, Ben-Farhat M, Betbout F, Gamra H, Addad F, et al. Eighteen-year experience with echinococcosus of the heart: clinical and echocardiographic features in 14 patients. Int J Cardiol 2003;91:145-51.