

Multiple angiomatous mitral valve cysts leading to floppy mitral valve syndrome

Floppy mitral kapak sendromuna yol açan çok sayıda anjiyomatöz mitral kapak kisti

Nazmi Gültekin, M.D., Emine Küçükateş, M.D., Murat Ersanlı, M.D., Erhan Kansız, M.D.

Department of Cardiology, Cardiology Institute, İstanbul University, İstanbul

We report an extremely rare case of cystic-tumor like formations that originated from the mitral valve tissue affected by verrucous endocarditis, leading to floppy mitral valve syndrome. These cystic tumoral formations were discovered during two-dimensional echocardiographic examination of a 46 year-old woman with cardiac symptoms of palpitation, dyspnea, and exertional angina pectoris. Multiple cysts were attached to the anterior mitral leaflet, resulting in pansystolic pseudoparachute-like floppy mitral valve prolapse, and severe mitral regurgitation. The patient underwent prosthetic mitral valve replacement following removal of the mitral valve and multiple cystic-tumoral formations. She had an uneventful postoperative course. Histological diagnosis was diffuse angiomatous cystic development of vasculatory tumor-like structures due to verrucous endocarditis.

Key words: Echocardiography; heart valve diseases/ultrasonography; mitral valve insufficiency; mitral valve prolapse/pathology/surgery.

The majority of patients with floppy mitral valves resulting in mitral valve prolapse have structural abnormalities that may be defined by echocardiography. A spectrum of floppy mitral valve structure has been demonstrated by echocardiography, with mitral regurgitation occurring more frequently in patients with multiple and more severe anatomic abnormalities. In addition to the presence of prolapse and regurgitation, the assessment of leaflet thickness, leaflet length, annular diameter, and chordal length is fundamental to the definition and stratification of patients with mitral valve prolapse associated with floppy mitral valve.

Bu yazıda, verüköz endokardit tutulumu olan mitral kapak dokusundan kaynaklanan ve *floppy* mitral kapak sendromuna yol açan irili ufaklı çok sayıda anjiyomatöz tipte kistik ve tümöre benzer yapılaşmanın görüldüğü son derece nadir bir olgu sunuldu. Bu kistik tümoral oluşumlar, çarpıntı, nefes darlığı ve eforla gelişen angina pectoris gibi semptomları olan 46 yaşındaki bir kadın hastanın ikiboyutlu ekokardiyografi ile incelenmesi sırasında ortaya çıkarıldı. Ön mitral kapakçığa tutunmuş olan çok sayıda kistik lezyon, pansistolik psödoparüşüt-benzeri mitral kapağı prolapsusu ve ciddi mitral yetersizliğine yol açmıştı. Mitral kapağın ve kistik tümöral oluşumların cerrahi olarak çıkarılmasından sonra hastaya prostetik mitral kapak takıldı. Ameliyat sonrası dönemde herhangi bir sorunla karşılaşılmadı. Histopatolojik tanı, verüköz endokardite bağlı vaskülatör tümör benzeri yapılardan diffüz anjiyomatöz tipte kistik gelişim olarak bildirildi.

Anahtar sözcükler: Ekokardiyografi; kalp kapağı hastalığı/ultrasonografi; mitral kapağı yetersizliği; mitral kapağı prolapsusu/patoloji/cerrahi.

CASE REPORT

A 46 year-old woman was admitted with symptoms of palpitation, dyspnea, and angina pectoris on exertion of eight-year history. These complaints were also present for a year on minor and minimal exercise related to housework. No information on personal or familiar cardiac anamnesis could be derived from the patient. On admission, she was evaluated as NYHA class III-IV. On cardiac examination, the apical impulse was weak and an S₃ was heard. P₂ was loud and there was a grade 3-4/6 apical pansystolic murmur. The electrocardiogram showed normal sinus rhythm with P mitrale. There was left

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Correspondence: Dr. Nazmi Gültekin. İ.Ü. Kardiyoloji Enstitüsü, Kardiyoloji Anabilim Dalı, 34034 Haseki, İstanbul.
Tel: 0212 - 459 20 00 / 29510 Fax: 0212 - 459 20 69 e-mail: nngultekin@yahoo.com

ventricular enlargement with a cardiothoracic ratio of 0.56 on a chest X-ray.

Echocardiography revealed multiple cysts attached to the anterior mitral leaflet, pansystolic pseudoparachute-like floppy mitral valve prolapse, and severe mitral regurgitation (Fig. 1). The anterior and posterior mitral leaflets were enlarged and deformed. The patient could not be stabilized clinically with digitalis, diuretics, and vasodilators, and she was submitted to open heart surgery. Multiple cystic-tumoral formations were observed with the removal of the mitral valve (Fig 2). Diffuse tumor-like multiple cysts of varying size originated from the mitral valve tissue. The pulmonary artery was slightly dilated. There was global cardiomegaly. The intracardiac tumoral-cystic formation protruded into the left atrium at surgical exploration and gross anatomic-pathologic examination. One of multiple cystic formations was giant, bloodsucker-shaped, and of saccular formation. The mitral valve was replaced with a Björk-Shiley 33-mm prosthetic valve and the patient had an uneventful postoperative course. Myocardial hypertrophy was determined in the left atrial appendage. The suspected diagnosis was histologically confirmed as diffuse angiomatic cystic development of vasculatory tumor-like structures due to verrucous endocarditis.

DISCUSSION

Mitral prolapse is a parachute-like protrusion of the valve into the left atrium. The floppy mitral valve prolapses in such a dynamic manner that it becomes a space-occupying lesion within the left atrium. Prolapsing floppy mitral valve also causes mitral valvular regurgitation.^[1-3] With the onset and gradual progression of mitral valvular regurgitation, altera-

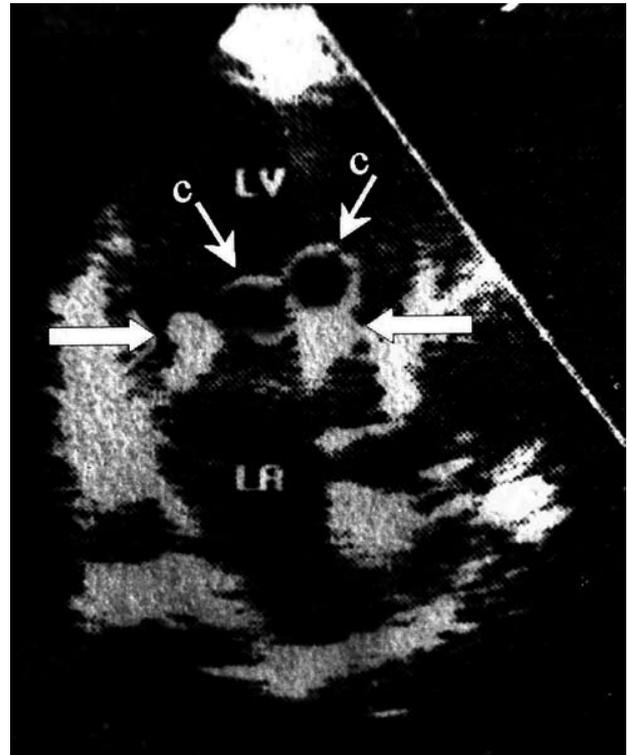


Figure 1. Echocardiography showing a pseudoparachute-like formation associated with multiple cysts attached to the anterior mitral leaflet leading to floppy mitral valve syndrome and severe mitral regurgitation. LV: Left ventricle; LA: Left atrium; C: cysts (arrows).

tions in the size and performance of the left atrial and ventricular chambers occur, resulting in left atrial and ventricular myopathy.^[1] Typically, prolapsed mitral valve tissue is shiny, grey-white edematous, and contains increased mucopolysaccharides in the zona spongiosa and chordae tendinae, and exhibits myxoid balloon cell degeneration (floppy mitral valve syndrome); however, in our case, these histopathologic features were not observed and the specimens exhib-

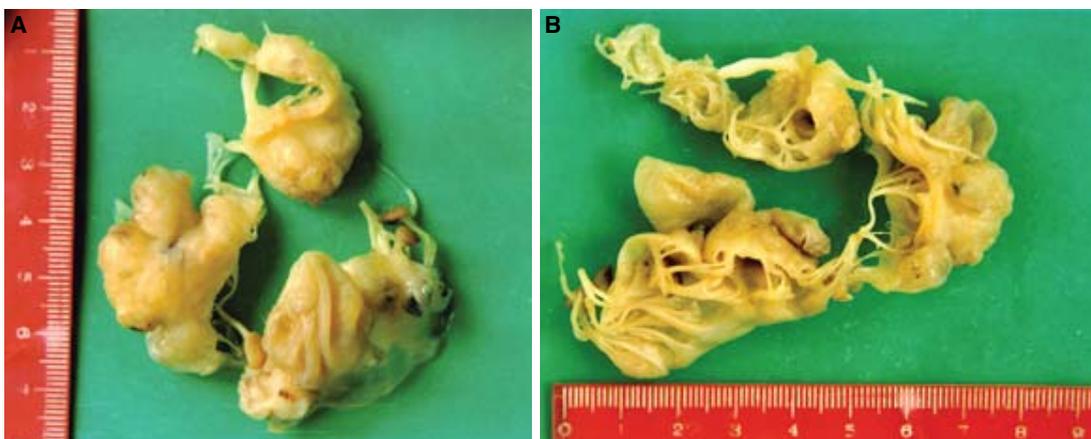


Figure 2. (A) Ventricular and (B) atrial views of the excised mitral valve, multiple angiomatic cysts associated with tumor-like formation due to verrucous endocarditis.

ited only multiple angiomatous mitral valve cysts of vasculatory tumor-like formation.^[2,4]

Blood cysts are congenital cysts located in the endocardium, particularly along the lines of closure of heart valves. They are lined by flattened endothelium and filled with nonorganized blood.^[5] Mitral valve blood cysts associated with floppy mitral valve are an extremely rare entity. They can be formed by trapping of blood in sacculated dissolved collagenous fibrous layers. If they cause functional derangement of the mitral valve, they must be excised surgically with or without prosthetic valve replacement.^[6] In our case, the floppy mitral valve was removed surgically along with the multiple cysts and was replaced with a prosthetic valve.

Mitral valve blood cysts can also be associated with mitral valve hamartoma.^[6-8] Hamartoma is a benign tumor-like nodule composed of an overgrowth of mature cells and tissues that are normally present in the affected tissue, but shows disorganization often with one element predominating. There are angiomatous subtypes, but hamartoma is usually a solid mass such as a cardiac rhabdomyoma.^[9] Our case also differed from mitral valve hemangioma or hamartoma histopathologically.

In conclusion, this case is a striking echocardiographic example of multiple cysts associated with floppy mitral valve, which was further confirmed by macroscopic and microscopic diagnosis as a tumor-like cystic lesion secondary to verrucous endocarditis. In addition, a cystic-tumoral lesion of angiomatous subtype arising from verrucous endocarditis associat-

ed with floppy mitral valve syndrome is an extremely rare entity causing severe mitral regurgitation and requiring mitral valve surgery.

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