

Accessory mitral papillary muscle causing severe aortic insufficiency

Aksesuar mitral papiller kasın yol açtığı ileri aort yetersizliği

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Accessory mitral papillary muscle originating from the interventricular septum is a rare congenital anomaly. A 20-year-old male patient presented with a complaint of exertional dyspnea. On cardiac examination, a grade 3/4 diastolic murmur was heard over the right upper parasternal area, and the apical pulsations were easily palpable over the precordium. Transthoracic echocardiography showed severe aortic regurgitation, dilatation of the left ventricle, and an accessory papillary muscle with its chordae, extending from the interventricular septum to the anterior mitral leaflet in the left ventricular outflow tract (LVOT). There was no mitral regurgitation. Color Doppler imaging showed turbulence set up by the abnormal mitral attachment and an associated mild pressure gradient of 20 mmHg across the LVOT. At surgery for aortic valve replacement, degenerative changes in the aortic leaflets were noted. The accessory papillary muscle was spared to maintain mitral valve functions and an aortic bileaflet mechanical prosthetic valve was implanted. During eight months of follow-up, he was well without any signs of left ventricular systolic dysfunction and mitral regurgitation, with a functioning prosthetic valve.

Key words: Adult; aortic valve insufficiency/etiology; echocardiography; mitral valve; papillary muscles/abnormalities.

Accessory mitral papillary muscle is an uncommon congenital cardiac malformation in young adults. Most of the cases in the literature are associated with left ventricular outflow tract (LVOT) obstruction and related symptoms. This anomaly may also coexist with hypertrophic cardiomyopathy and other congenital malformations; however, aortic valve degeneration associated with the presence of an accessory papillary muscle across the LVOT is an unusual condition.^[1,2] It has been well-demonstrated that subaortic pathologies such as discrete mem-

İnterventriküler septumdan köken alan aksesuar mitral papiller kas nadir bir doğuştan bozukluktur. Yirmi yaşındaki erkek hasta eforla gelişen nefes darlığı yakınmasıyla başvurdu. Kalp muayenesinde sağ üst parasternal bölgede 3/4 dereceli diyastolik üfürüm duyuldu; apikal pulsasyonlar prekordiyum üzerinde kolayca alınabiliyordu. Transtorasik ekokardiyografide ciddi aort yetersizliği, sol ventrikül genişlemesi ve kordu ile birlikte aksesuar papiller kas saptandı. Kordun interventriküler septumdan sol ventrikül çıkış yolunda (SVÇY) anterior mitral yaprakçığa kadar uzandığı gözlemlendi. Mitral yetersizlik yoktu. Renkli Doppler görüntüleme anormal mitral bağlantısının türbülans oluşturduğu izlendi ve SVÇY'de 20 mmHg'lik hafif basınç gradiyenti ölçüldü. Hastanın aort kapağını değiştirmek için yapılan ameliyatta aort yaprakçıklarında dejeneratif değişiklikler görüldü. Mitral kapak fonksiyonunun korunması için aksesuar papiller kas yerinde bırakıldı ve iki yaprakçıklı mekanik prostetik aort kapağı takıldı. Sekiz aylık takip döneminde hastada sol ventrikül sistolik disfonksiyonu ve mitral yetersizliğe ait bir bulguya rastlanmadı, prostetik kapağın fonksiyonu normal idi.

Anahtar sözcükler: Erişkin; aort kapağı yetersizliği/etyoloji; ekokardiyografi; mitral kapağı; papiller kas/anormallik.

brane or ventricular septal defect can be associated with jet flow across the ventricular outflow tract and may damage the aortic leaflets leading to secondary regurgitation.^[3] Therefore, turbulence and jet flow caused by abnormal mitral attachments can be the cause for the degeneration of the aortic valve leaflets and associated valvular insufficiency. Herein, we present a case in which nonobstructive septal attachments of the anterior leaflet of the mitral valve were incidentally found during investigation for aortic regurgitation.

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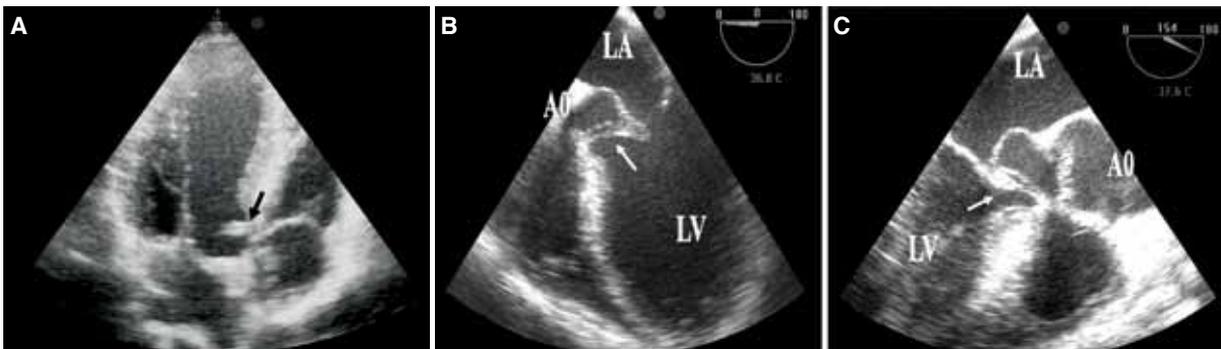


Figure 1. (A) Preoperative transthoracic echocardiography shows accessory papillary muscle (black arrow) across the left ventricular outflow tract. Transesophageal echocardiography views in (B) diastole and (C) systole show accessory papillary muscle (white arrows) originating from the interventricular septum with its chordae extending to the anterior mitral leaflet. Ao: Aorta; LA: Left atrium; LV: Left ventricle.

CASE REPORT

A 20-year-old male patient presented with a complaint of exertional dyspnea that increased in severity within the past two months. His medical history was unremarkable. On cardiac examination, a grade 3/4 soft diastolic murmur was heard over the right upper parasternal area, and the apical pulsations were easily palpable over the precordium. He was normotensive but had an increased pulse pressure. The electrocardiogram showed sinus rhythm and left ventricular hypertrophy with left axis deviation evident in lateral chest derivations. A chest roentgenogram showed an increased cardiothoracic index. Transthoracic echocardiography (TTE) showed severe aortic regurgitation, dilatation of the left ventricle (internal dimension in diastole and systole: 63 mm and 45 mm, respectively) and an ejection fraction of 55%. There was an accessory papillary muscle with its chordae in the subaortic area in addition to the anterior and posterior mitral leaflets (Fig. 1a). There was no mitral regurgitation or stenosis, nor an evident dilatation of the aortic root. A subsequent transesophageal echocardiography (TEE) examination confirmed the presence of an accessory mitral papillary muscle and an associated chordae extending from the interventricular septum to the edge of the anterior leaflet in the LVOT, together with severe aortic regurgitation (Fig. 1b-c, 2). Color Doppler imaging showed turbulence set up by the abnormal mitral attachment and an associated mild pressure gradient of 20 mmHg across the LVOT. It was thought that the turbulence by this large accessory papillary muscle would possibly lead to degenerative changes in the leaflets of the aortic valve and to severe aortic valvular insufficiency in such a young patient. The patient was scheduled to an elective procedure involving aortic valve replacement.

Operation was performed under cardiopulmonary bypass and moderate systemic hypothermia. Following cardiac arrest and the aortotomy incision, degeneration of the aortic leaflets was noted. There was neither calcification nor thickening of the leaflets and annulus of the aortic valve, suggesting an underlying infectious or inflammatory process. Following resection of the degenerative leaflets, the accessory papillary muscle was inspected through the aortic orifice. It originated from the interventricular septum and attached to the anterior leaflet. This anomalous subvalvular apparatus of the mitral valve was spared to preserve mitral valve function, because TEE examination had not shown a significant pressure gradient in the LVOT or evident mitral insufficiency. The procedure was completed uneventfully after implantation of a 21-mm St. Jude aortic bileaflet mechanical prosthetic valve. The patient was discharged on postoperative day 5 with anticoagulant therapy and a

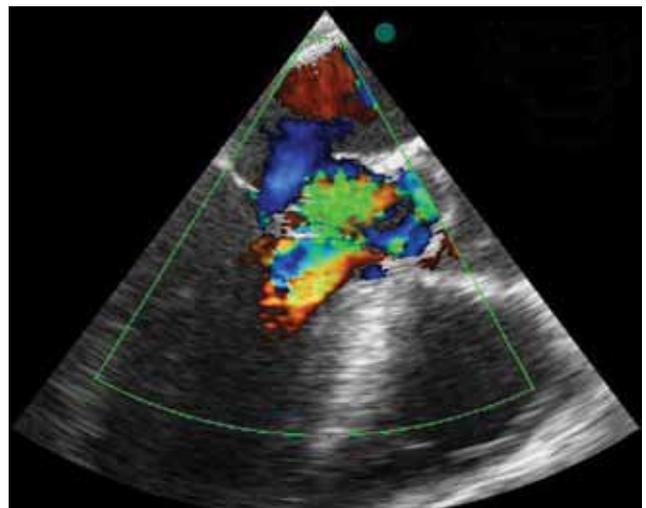


Figure 2. Intraoperative transesophageal echocardiography image shows severe aortic regurgitation.

favorable outcome. During eight months of follow-up, he was well without cardiac symptoms. Control TTE examination did not show left ventricular systolic dysfunction or any abnormality of the prosthetic valve function. There was no mitral regurgitation.

DISCUSSION

Anomalies of the subvalvular apparatus of the mitral valve are unusual congenital malformations and generally present in infancy or early childhood. These structural anomalies are frequently associated with LVOT obstruction or hypertrophic cardiomyopathy.^[1,2] In particular, an accessory papillary muscle originating from the interventricular septum, with attachment to the anterior leaflet of the mitral valve is an unusual entity in adults. Although there are many reports of septal attachments of the mitral valve, it is not clear how frequently an accessory papillary muscle presents in young adults.

Anomalies of the mitral subvalvular apparatus can present in four different forms: anomalous insertion of papillary muscle directly into the anterior mitral leaflet as the most common type,^[4] extensive fusion of papillary muscles with the ventricular septum or left ventricular free wall, abnormal chordae tendineae (false cords) attaching to the ventricular septum or free wall, and accessory papillary muscles. All these abnormalities may tether the mitral leaflets toward the septum and produce LVOT obstruction or mitral regurgitation. Their long-term course in adults is unknown. It is unknown whether an asymptomatic accessory papillary muscle in the LVOT would ultimately result in severe aortic valve degeneration and related symptoms requiring surgical intervention.

The variability of subvalvular anatomy and papillary muscle alignment is most likely related to abnormal valve morphogenesis.^[5] After 5 to 6 weeks of gestation, the left ventricular septum undergoes involution with both chordae and papillary muscles originating from the septal muscle tissue. Failure of origin from the ventricular wall or an abnormal origin of papillary muscles would explain the accessory papillary muscle originating from the interventricular septum. On the other hand, the presence of chordal structures across the LVOT, arising from a hypertrophic accessory papillary muscle, would cause turbulent systolic jets or blood flow leading to aortic valvular degeneration and insufficiency.^[1] Damage to the aortic valve due to subvalvular systolic jet flow has been implicated as the main cause of valvular aortic regurgitation in subval-

ular pathologies such as discrete subvalvular aortic stenosis.^[6,7] In such cases, repetitive trauma from turbulent blood flow causes fibrosis of the leaflet, and can play an important role in the retraction of the valve leaflets.

Septal attachments of the mitral valve are usually symptomatic, and clinical presentation varies with associated anomalies such as ventricular septal defect, fibromuscular tunnel, or hypertrophy of the interventricular muscular septum. Tethering of the mitral valve because of thickening and retraction of anomalous papillary muscle would restrict its normal excursion during systole out of the LVOT. Systolic blood flow may cause a Venturi effect leading to anterior motion of the anterior mitral valve cusp, resulting in LVOT obstruction or mitral insufficiency. When there is co-existing hypertrophy of accessory papillary muscle or interventricular septum, systolic anterior motion of the mitral valve with LVOT obstruction may be evident.^[4,8] Thus, echocardiography plays a vital role in the evaluation of these structures and their relation to cardiac functions.

Surgical resection of accessory papillary muscle causing a pressure gradient across the LVOT may be necessary to relieve LVOT obstruction, mitral insufficiency, and related clinical symptoms. Aortic valve replacement is indicated, as in our case, when aortic insufficiency develops as a result of valvular degeneration associated with the turbulence by subvalvular accessory papillary muscle and its chordae.

In conclusion, an accessory mitral papillary muscle originating from the interventricular septum across the LVOT is rare in young adults and may be the cause of degeneration of the aortic valve leaflets leading to valvular insufficiency.

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