Muscularisation of the chordae tendineae: an unusual etiology for aortic insufficiency

Muskularize korda tendinea: aortik yetmezliğin etyolojisinde az rastlanan bir neden

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Introduction

We can classify left ventricular (LV) chordae tendineae into three groups. The first group inserted on the free edge of the mitral leaflet, the second group 6-8 mm from the edge on the LV surface of the leaflet and the third group (on the posterior leaflet only) inserted to the basal portion of the leaflet (1,2). On average 25 -chordae insert on the mitral valve. Chordal branching has so many variations that some remain unbranched or branching into three cords before insertion into the leaflet (3-6). The LV usually has two papillary muscles (PM) (posteromedial and anterolateral), both arise from the LV free wall, unlike the right ventricle (RV), without any papillary muscles arising from the septum. Both PM are attached to the middle third of the LV with broad bases, although occasionally accessory PMs may arise in the apical third (3,5). The interactions between the mitral valve and left ventricle are complex and not yet completely understood. However, continuity between the papillary muscles and the mitral annulus is probably the most important factor in this relationship because severance of the chordae tendineae in experimental animals causes a significant drop in left ventricular systolic function as assessed by load independent parameters (7,8).

Chordae muscularis constitutes an uncommon variant of chordal configuration that a chord may be muscular and fleshy instead of being a tendinous structure either in its whole length or sausage-shaped with a muscular central segment and tendinous origin and insertion. This anomaly is known as muscularisation of the chordae which is an autopsy, surgical or echocardiography finding, reveals that the papillary muscle inserts directly on a leaflet without any intervening to chorda tendinea (3).

Case Report

Our case is a 20-year-old white male, admitted to our clinic for the evaluation of shortness of breath. There was a diastolic murmur best heard at end expiration at the fourth intercostal space intersecting the midclavicular line without any spread. His electrocardiogram showed sinus rhythm with right axis deviation and marked both left and right ventricular hypertrophy by voltage with ST and T wave strain pattern. His chest X-ray revealed mild cardiac silhouette enlargement detected at base.

His transthoracic echocardiography (TTE) was performed with Vingmed system V (GE, Horton, NORWAY) with 2.5 MHz probe and transesophageal echocardiography (TEE) was performed with the same machine utilizing a 5 MHz TEE probe (9). On TTE, the LV chamber was mildly dilated with normal right ventricular (RV) size and normal wall motion, mild hypertrophy of both septal (IVS) and lateral LV walls. His color and continuous wave Doppler (CW) examination of the aortic valve was consistent with moderate to severe aortic insufficiency (AI) with an uncertain band localized at subaortic level. To rule out the etiology of AI, TEE was performed and revealed LV dilatation with mildly thickened aortic cusp edges, moderate to severe AI and muscular chordae originating from the lateral wall of the LV inserting on the left coronary cusp (LCC) of the aortic valve. In every heart cycle of the LV, the muscular chordae contracts and pulls down the LCC causing a coaptation anomaly giving rise to AI (Fig. 1). After TEE, his cardiac catheterization revealed normal left ventriculogram with mildly dilated LV and no gradient was recorded at the aortic valve or left ventricular outflow tract level. His aortography revealed severe AI. We recommend aortic valve replacement with the excision of the papillary muscle. Although our patient was informed in terms of all the complications (arrhythmia, embolism, heart failure etc) and his survey regarding with the AI, he refused the surgical procedure.

The patient was discharged on the seventh day with Warfarin therapy.

Discussion

The LV usually has two papillary muscles; both arise from the LV free wall. Our case has two papillary muscles in LV and a muscular structure originating from the lateral wall of LV inserting directly on to the LCC of the aortic valve. This is a very rare congenital heart anomaly and very hard to diagnose, when the papillary muscle inserts directly on a leaflet without any intervening to chorda tendinea (3,10,11). These anomalies are usually asymptomatic and are diagnosed at autopsy. They mostly cause

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strokes or transient ischemic attacks due to cerebral embolism. The reason of the muscular tissue replacing the chorda tendinea lies in the embryologic development of the chorda tendinea, papillary muscles and valve tissue, when all these structures arise by delamination of the primitive endocardium (3). Transesophageal echocardiography, which provides detailed information both for the mitral valve anatomy and the papillary muscles, is a useful laboratory tool regarding with the diagnosis of muscularisation of the chordae tendineae. All examination procedures and measurements were obtained according to American Society of Echocardiography (ASE) guidelines in this case (9). This anomaly is known as muscularisation of the chordae, which is an autopsy, surgical or echocardiographic finding (3,9,10). The incidence, the affecting population around the world or the male female ratio is not clear according to the literature (4,8).

If muscularisation of the chordae is diagnosed on echocardiography, warfarin or antiplatelet treatment is useful prevent thromboembolic events. When the reports of cases are taken into account it can be easily seen that if there is an embolic event suggesting the presence of muscularisation of the chordae, surgical intervention will be necessary.

Although the patient was informed in terms of all the complications (arrhythmia, embolism etc.) and his survey, the patient refused the surgical procedure.

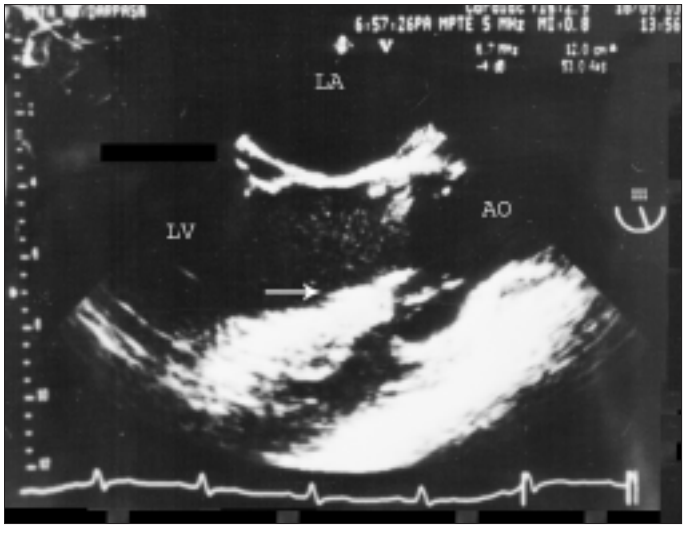

Figure 1. Midesophageal level at 111º TEE view; a cord-like structure originating from the lateral wall of LV inserting on an aortic cusp is seen

TEE- transesophageal echocardiography, LV- left ventricle

As we conclude; this is a rare congenital anomaly causing AI and according to literature and to the best of our knowledge this is the first case reporting muscularisation of the chordae tendineae in Turkish people.

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