ORIGINAL RESEARCH PAPER

Cardiology

KEY WORDS: Aberrant chordae, LV apical chordae attachment, 4D XStrain Echocardiogrpahy.

ORIGINAL RESEARCH ARTICLE DISTINCTIVE ECHOCARDIOGRAPHIC IMAGES OF ACCESSORY CHORDAE TENDINEAE INSERTION INTO LEFT VENTRICULAR APEX: DETAILED CASE STUDY WITH REVIEW OF LITERATURE

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ABSTRACT

On routine trans thoracic echocardiography (TTE) in an asymptomatic 33 years old hypertensive male, we identified an aberrant chordae tendineae (ACT) arising from the tip of anterior mitral leaflet (AML) and then descending towards the LV apex, traversing The left ventricular (LV) cavity in the form of a striking echodense vertical band, running parallel and adjacent to the ventricular septum and finally attaching to LV apex in a characteristic "Y" shaped arrangement .

INTRODUCTION

The mitral valve (MV) complex consists of functional units, which include the fibrous trigones, chordae tendineae and papillary muscles. Tendinous cords and papillary muscles connect MV leaflets to the LV free wall like shrouds of a parachute [1,2] (Figure 1-4).

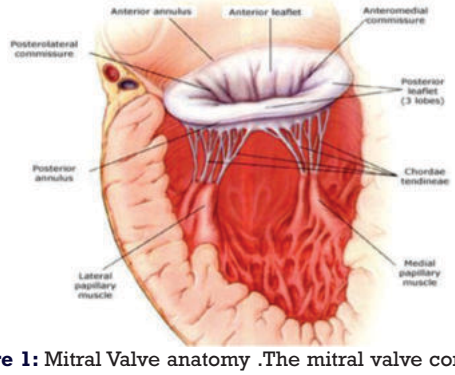

Figure 1: Mitral Valve anatomy .The mitral valve consists of themitral valve annulus, anterior and posterior leaflets, chordae tendineae and the papillary muscles.

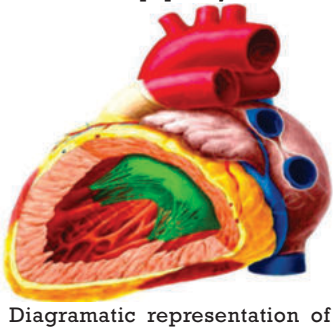

Figure 2: Diagramatic representation of mitral valve apparatus.

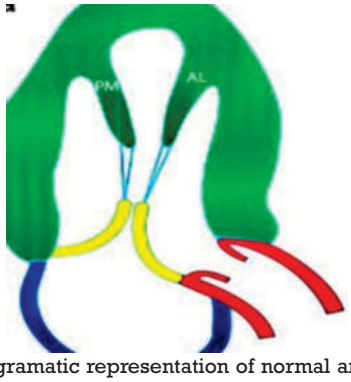

Figure 3: Diagramatic representation of normal anatomy of mitral valve apparatus in the vertical long-axis projection.The papillary muscles originate from the free wall of the LV attached to trabecula carnea.The papillary muscles give rise to multiple chordae tendineae (blue) which attach to the mitral valve (yellow).

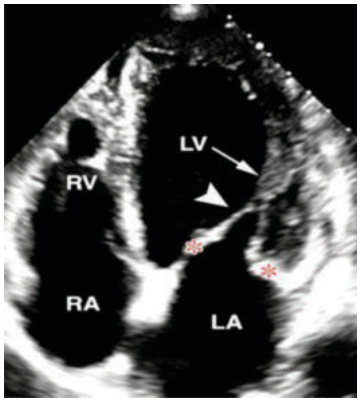

Figure 4: Apical 4 Chamber View shows an anterolateral papillary muscle (arrow) and a chordae tendineae (arrowhead) attached to the anterior and posterior mitral leaflets (*).

The MV and its subvalvular apparatus are integral parts of LV and play an important role on its geometry and systolic function. Anomalies of mitral subvalvular apparatus includes differing types of papillary muscles and chordae tendineae.

Anomalous chordal insertion to interventricular septum is a rare anomaly with an incidence of 1 per 26,000 echocardiograms in adult [4]. A clinical syndrome caused by rupture of chordae tendineae (CT) of mitral valve has been recognized with increasing frequency [5-7]. The rupture may be associated with a fulminant onset followed by rapid eventful course or it may produce mild symptoms with a chronic course or it may be completely asymptomatic [5-7]. The anomalous insertions may also present with arrhythmias, palpitations, syncope and rarely sudden death [8,9].

General Characteristics of Mitral Valve Chordae Tendineae

The chordae tendineae are fibrous strings that originate from tiny nipples on the apical portion of the two left ventricular papillary muscles or directly from the ventricular wall. Those that insert into the valve are true chordae tendineae, those that insert elsewhere, for example, into the muscle, are false chordae tendineae.

Careful investigation of the relationship of chordal

morphology to the site of insertion of individual chordae guided the authors Lam et al [3] to propose the new classification (Figure 5).

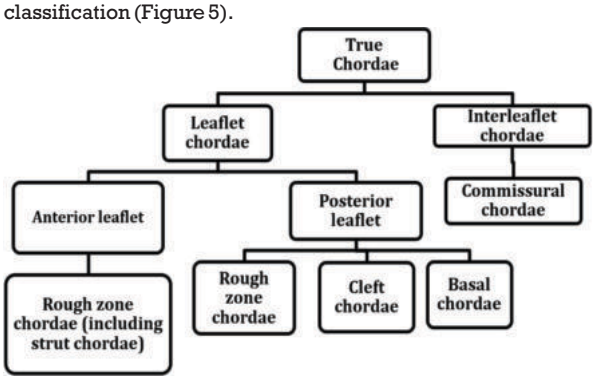

Figure 5: Classification of true chordae tendineae of the mitral valve (adapted from Lam et al [3])

This classification emphasizes a distinction between leaflet chordae and the fanlike interleaflet or commissural chordae. It defines three distinct types of leaflet chordae. The first type is termed "rough zone chordae" because they insert into the distal rough portion of the anterior and posterior leaflets. Two of the rough zone chordae of the anterior leaflet are very thick and are named strut chordae. The second type of leaflet chordae, basal chordae, insert into the base of the posterior leaflet. The third type are cleft chordae which insert into the indentations or clefts between the scallops of the posterior leaflet.

Using the classification a close clinicopathologic correlation in cases of mitral regurgitation secondary to ruptured chordae tendineae may better delineate the clinical spectrum of this syndrome. It might be expected that rupture of the strut chordae of the anterior leaflet might cause more severe regurgitation than rupture of other types of mitral valve chordae tendineae.

We are reporting a unique case report of an aberrant insertion of chordae tendineae to the LV apex in an asymptomatic hypertensive male. Although ECG, X-ray chest (PA) and clinical evaluation were non-informative, however, routine TTE was able to delineate this peculiar lesion.

Case Report

A 33 year old asymptomatic adult male was referred to us for routine TTE because of history of hypertension for two years.

4 Dimensional XStrain Echocardiography (4D XStrainE)

TTE was performed by 4D XStrain system of Esaote, Italy in the left lateral decubitus position. There was presence of concentric hypertrophy of left ventricle with a left ventricular mass of 218 gm in end diastole (Figure 6a).

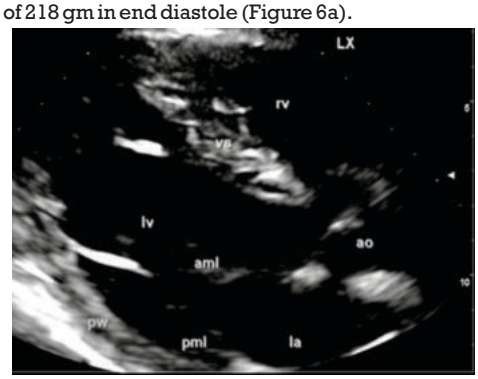

Figure 6a: Long axis depicts concentric hypertrophy of LV

There was no regional wall motion abnormality detected and his biplane LV ejection fraction and volumes were normal (Figure 6b).

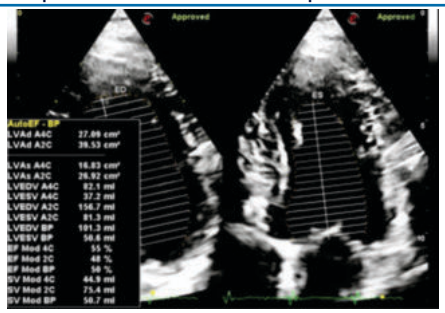

Figure 6b: Apical 2 Chamber and 4 Chamber view delineates normal biplane Simpson LV ejection fraction and volumes.

The fascinating finding was the presence of a striking aberrant echodense chordae tendineae attached to the LV apex in a "Y" shaped pattern (Figure 7) and traversing the LV cavity in a vertical fashion lying parallel and adjacent to the interventricular septum and finally inserting to the tips of anterior mitral leaflet (Figure 8).

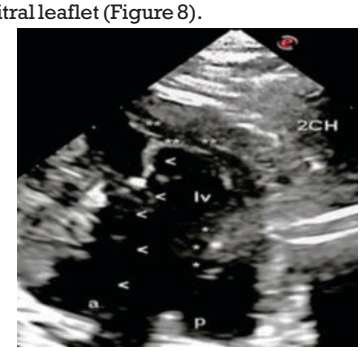

Figure 7: Apical 2 Chamber View defines the chordae attachment to LV apex in a "Y" shaped pattern. Moreover, aberrant chordae can be seen traversing upwards into the LV cavity in a vertical fashion, parallel and adjoining the ventricular septum and finally attraching to the anterior mitral leaflet. a, anterior mitral leaflet, p, posterior mitral leaflet

Figure 8: Apical 2 Chamber View depicts the chordal attachment sites: 1. To the LV apex (<), and 2. To the LV free wall (*)

Furthermore, another characteristics feature was the presence of two chordal insertion sites after arising from the anterior mitral leaflet: a) to the LV apex, as detailed earlier and b) to the antero-lateral LV wall (Figure 9).

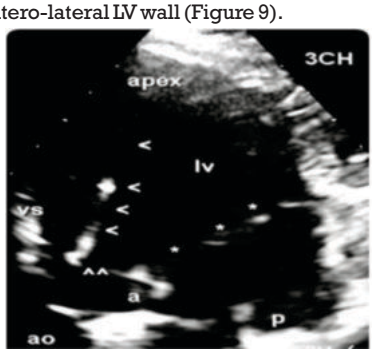

Figure 9: Apical 3 Chamber View reveals a conspicuous

attachment of chordae at 2 sites after arising from the anterior mitral leaflet: 1. LV apex, and 2. LV free wall. a, anterior mitral leaflet, p, posterior mitral leaflet.

Conspicuously, laminar, non-turbulent flow was noted in LVOT consistent with unobstructed circulation (Figure 10).

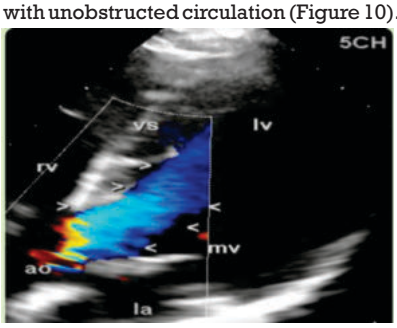

Figure 10: In the Apical 5 Chamber View a smooth laminar flow is identified in the LVOT, consistent with unobstructed circulation.

DISCUSSION

The mitral chordae tendineae which typically number 25, are thin, fibrous structures composed of collagen interwoven with elastin fibers, which extend from the papillary muscles to attach to the mitral leaflets. The chordae provide the critical function of anchoring the mitral leaflets during systole, allowing for symmetric coaptation and preventing prolapse of the leaflets into the LA.

The chordae are attached to the LV via the papillary muscles. During systole, the papillary muscles contract to facilitate closure of the leaflets via the chordae. The papillary muscles arise from the area between the apical and middle thirds of the LV free wall and are divided into two groups: the anterolateral and the Posteromedial papillary muscles [3].

MV apparatus consist of left atrioventricular orifice and its annulus, two valve leaflets, chordae tendineae and two papillary muscles. Function of the valve is dependant on the anatomic and mechanical structure of all these parts [10]. MV is a continuous attachment around the entire mitral orifice rather than two distinct leaflets [11]. Its free edge has dips. There are two deep and constant indentations called anterolateral and posteromedial Commisures. The chordae attached here are called commissural chordae.

The anterior leaflet is semicircular or triangular. The leaflets consists of two zones. There is a deep crescentic rough zone and a clear zone. The anterior leaflet has no basal zone. The posterior leaflet usually has two clefts and has rough, clear and basal zones. However, it does not have any strut chordae among its rough zone chordae [3].

True chordae of mitral valve consist of anterolateral or posteromedial commissural chordae and other types of leaflet chordae [3]. Rough zone chordae include strut chordae. The posterior leaflet has rough zone, cleft, and basal chordae. Anterolateral and posteromedial commissural chordae arise by a single stem fanning out immediately into radiating strands attached to the smooth free margin of the commisure.

The anterior leaflet has two specific chordae. They are very thick and sturdy. They arise from tips of anterolateral and posteromedial papillary muscles to attach near the line of valvar closure posteromedially (4-5 o'clock position) and anterolaterally

(7-8 o'clock position), respectively. They are strut chordae, visible in over 90% of the hearts [3]. Their zone of attachment was earlier termed as the critical point of tendinous insertion of the anterior leaflet [12].

Most true chordae have a single stem and divide into branches soon after their origin from papillary muscle Fan-shaped chordae have short stems and branch profusely to attach to the valve leaflets. Rough zone chordae usually have a single stem which splits into three strands. Basal chordae are solitary cords and pass from the ventricular wall to the valve leaflet [13].

The chordae tendineae form an important part of MV apparatus connecting papillary muscles to the valve leaflets. Chordae tendineae rupture will lead to mitral incompetence due to loss of support to the valve leaflet [14, 15]. In mitral incompetence, repair should include restoring the chordae also. It requires knowledge of type of repair and the number of chordal substitutes for reimplantation [16]. There is thickening and fusion of chordae in rheumatic mitral stenosis which leads to disturbances in their functioning. Valve leaflets are unable to open during diastole. This gains importance when the deformity is to such an extent that the function of the valve gets disrupted and clinical symptoms appear.

The fan-like arrangement of the commissural chordae would facilitate the anterior and posterior leaflets to come into contact with commissural region by hinge-like movements. Because of its position the posteromedial commissural chordae would be a more accurate guide in performing commisurotomies [17]. Atrioventricular valve repair may be preferred by cardiac surgeons instead of valve replacement [18]. Chordae replacement or repair has proved to be very useful in mitral valve repair [19]. A detailed knowledge of morphology of AV valves, therefore is important while performing surgeries.

Our patient is currently asymptomatic and because the aberrant chordae is not causing any systolic or diastolic functional abnormalities of LV, was thus advised to get a yearly echocardiographic review of his atypical lesion.

CONCLUSION

Although anomalous insertion of chordae may be asymptomatic, yet in rare individuals are associated with catastrophic outcome. Routine TTE can characterise these lesions with accuracy. Thus it is imperative that every academic and practicing cardiac physician must be aware of such situations and recommend standard echocardiography to precisely rule out these cardiac aberrancies in patients presenting with atypical cardiac symptoms.

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Purpose Of Article: Academic

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