



Aberrant Mitral Valve Chordae Tendineae Insertion into the Left Ventricular Apex: A Unique Case Report

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Abstract

A 33 years asymptomatic hypertensive male underwent a routine color echocardiography for assessment of overall cardiac anatomy, left ventricle (LV) regional wall motion abnormality, LV wall thickness, systolic and diastolic functions. A striking echodense vertical band was visualized in the LV while visualizing the apical two, three and four chamber views. Nevertheless, despite its anatomic location, there was no evidence that the anomalous mitral valve chordae tendineae was causing any regurgitation of mitral valve or obstruction to left ventricular outflow tract (LVOT). To the best of our knowledge this is the first case report of chordae attachment to the LV apex.

Keywords: Aberrant mitral valve chordae tendineae, abnormal mitral valve apparatus, 4D X Strain Echocardiography.

Introduction

Anomalies of the mitral subvalvular apparatus can include differing types of papillary muscles and chordae tendineae. Direct insertion of the anomalous papillary muscle into the anterior mitral leaflet or chordae tendineae to the ventricular septum is common findings related to these anomalies.¹ Chordae by conventional nomenclature are divided into primary, inserting into the margin of the leaflet, secondary as inserting into the ventricular surface of leaflet and tertiary as inserting into the basal portion of posterior leaflet. Insertion at other sites like free wall or septum is considered anomalous resulting

due to aberrant growth of endocardial cushion. Anomalous insertion of chordae to interventricular septum can be asymptomatic or it can present with chest pain, palpitation, mild to severe LVOT obstruction causing exercise intolerance, syncope on exertion or early repolarisation in ECG.² We report a unique case of anomalous insertion of chordae to left ventricular (LV) apex in a young adult male. Although clinical evaluation and ECG were non-informative in this case, routine trans thoracic echocardiography (TTE) was able to delineate this characteristic lesion. Although being a rare entity, it can be ignored so far as the outcome is concerned.

Case

History of Clinical Presentation: A 33 year old adult male was referred to us for routine TTE because of history of hypertension for two years. The pleasant healthy looking adult was having a weight of 80 kg, height 171 cm2 last, SPO₂ 98 % at room air, Pulse rate 85/min. and BP 166/100 mmHg in the Right arm in sitting position. He was absolutely asymptomatic without any history of

chest pain, shortness of breath, palpitation or syncope. There was nothing remarkable in the cardiovascular examination. He was having normal heart sounds and there was absence of murmurs or clicks on auscultation. Moreover his resting ECG and X-Ray chest (PA) were inconspicuous (Figure 1).The pathological reports to rule out the presence of Diabetes, Thyroid and Dyslipidemia were all in the normal range.

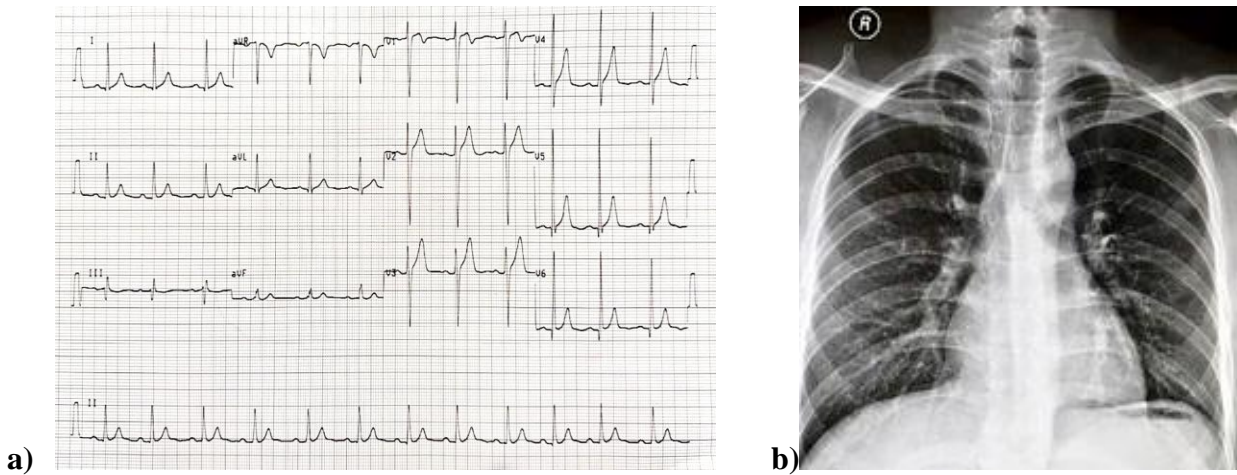


Figure 1: a) Normal ECG, b) Normal X ray Chest PA view

4 Dimensional X Strain Echocardiography (4D X Strain E): TTE was performed by 4D X Strain 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 2a). There was no regional wall motion abnormality detected and his biplane LV ejection fraction and volumes were normal (Figure 2b).

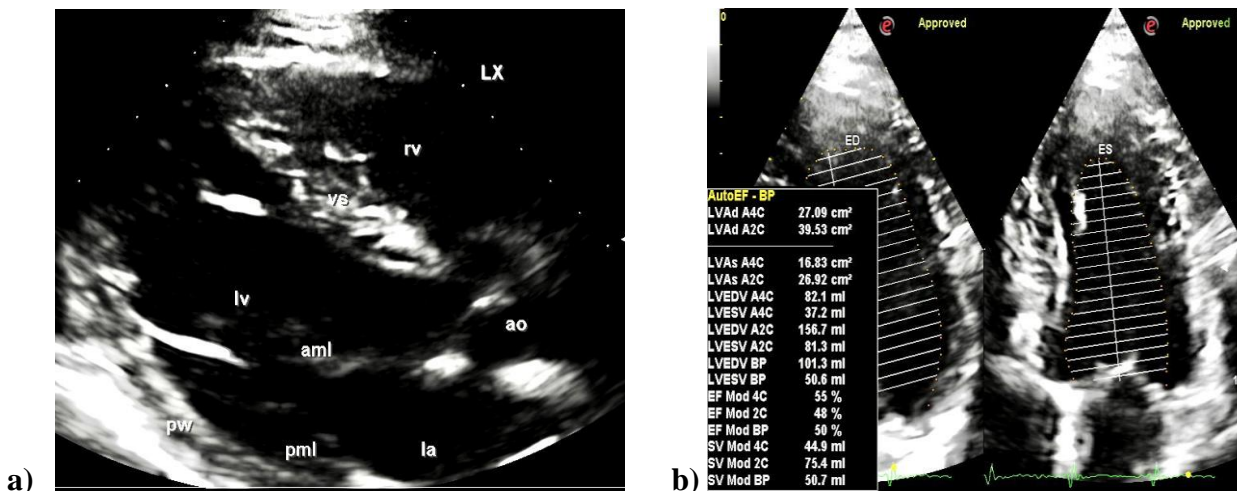


Figure 2: a) 4DXStrainE in long axis view b) by biplane Simpson method LVEF and LV volumes were normal

The fascinating findings was the presence of a striking aberrant echodense chordae tendineae attached to the LV apex in a “Y” shaped pattern

and traversing the LV cavity lying parallel and close to the interventricular septum and attaching to the anterior mitral leaflet(Figure 3a, 3b).

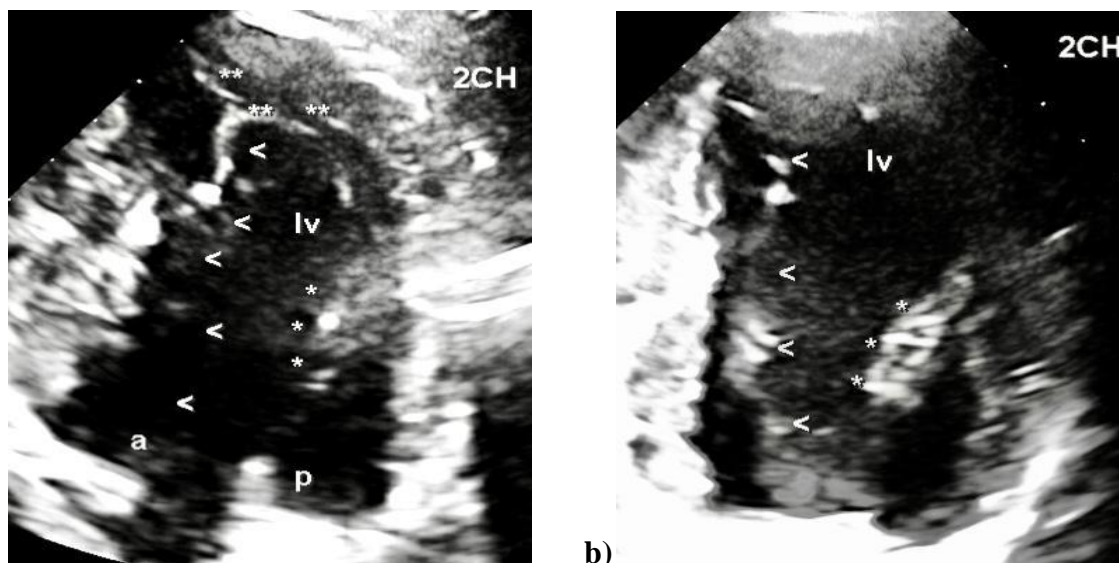


Figure 3: a) 2CH view defines the chordae tendineae attachment to lv apex in a “Y” shaped pattern. Aberrant chordae navigating in a vertical fashion from the lv apex to the base of the heart and finally inserting into anterior mitral leaflet

b) In the 2 CH view the chordal attachment sites are depicted after arising from mitral valve leaflets. Partially they are attached to lv free wall and the rest are attached to lv apex lv, left ventricle, < aberrant chordae traversing the lv, a, anterior mitral leaflet, p, posterior mitral leaflet

Another characteristic finding noteworthy was that the chordae tendineae arising from anterior mitral leaflet were partly attaching themselves to the free wall of LV besides inserting into the LV apex (Figure 4a). Conspicuously laminar, non-turbulent flow was noted in LVOT consistent with no LVOT obstruction (Figure 4b).

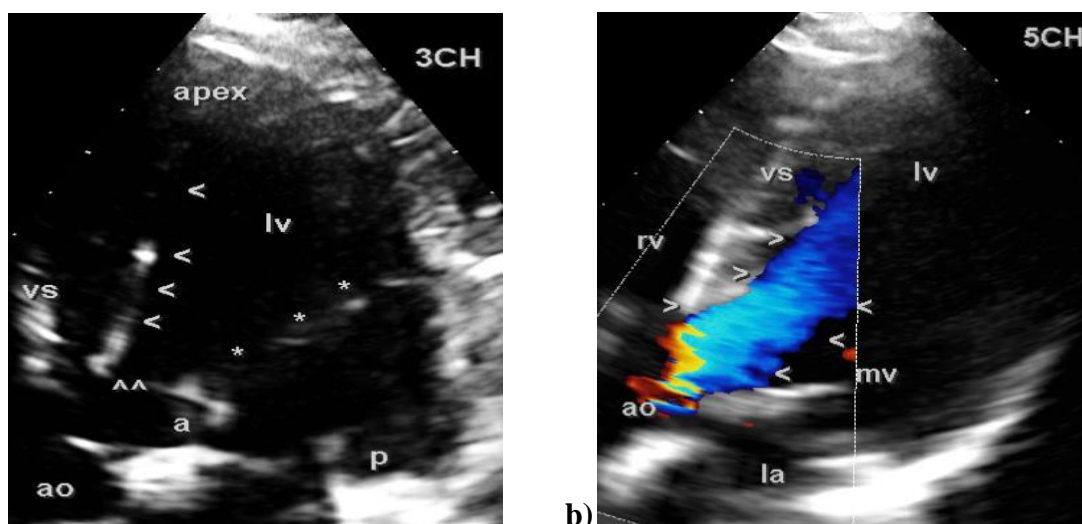


Figure 4: a) In the 3CH view there is a conspicuous insertion of chordal attachment at two sites after arising from anterior mitral leaflet 1. lv free wall and 2. lv apex

b) laminar non turbulent flow is seen in LVOT in 3CH view

< ^ * chordae tendineae, a, anterior mitral leaflet, p, posterior mitral leaflet, AO, aorta, vs, ventricular septum, la, left atrium

Discussion

Anomalous chordal insertion to interventricular septum is a rare anomaly with an incidence of 1 per 26,000 echocardiograms in adults.³ There are some reports regarding aberrant chordae tendineae attached to the Aortic Valve (AV).⁴⁻⁶ These reports show the attachment of the chordae tendineae to various parts of the aorta and the AV cusp, accompanied by AV regurgitation. In one of the reported cases, the chordae tendineae were attached to the aortic cusp and the Valsalva aortic root.⁴ In another case, the chordae tendineae were connected to the aortic cusp and the ascending aorta.⁵ In another patient, the chordae tendineae were attached to the aortic cusp and the Sinotubular junction.⁶ Abdelaziz et al. described a patient in whom transesophageal echocardiography detected a band-like structure attached to the AV without interfering with the AV function. Surgical exploration revealed that this structure was attached to the right coronary cusp and that the other head of this structure was detached from an unknown site in the left ventricle. The histological examination of this structure was compatible with chordae.⁷ Moreover anomalous chordae tendineae insertions into the basal interventricular septum⁸ and LVOT⁹ causing obstruction have also been reported. Thick anomalous chordae exerts strain on ventricular muscles generating atypical cardiac pain. These anomalous insertions may also give rise to arrhythmias, palpitation and syncope. They may even present with sudden cardiac death. These anomalous insertions are associated with congenital anomalies like D-TGA,¹⁰ Ebstein's anomaly and HCM.¹¹ Both leaflets and chordae originate from the cushion tissue¹² and as each stage of embryological development may be abnormal, the different malformations of the mitral valve can be associated with those developmental anomalies. Routine echocardiography can delineate those cases after exhaustive evaluation.¹²

Conclusion

Anomalous insertion of chordae although asymptomatic but, if missed may cause catastrophe like sudden cardiac death. Routine echocardiography can characterise these lesions with accuracy and thus finding an anomalous chordae leads to further echocardiography assessment to cautiously search for associated congenital anomalies. Physicians while evaluating atypical chest pain in outdoor patients must contemplate to perform a TTE, so that these rare cases, although at times associated with lethal outcomes, are not undetected.

To the best of our knowledge we are presenting a first case report of our patient manifesting with aberrant chordae insertion at the LV apex albeit without any cardiac symptomatology or cardiovascular complication.

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