Multimodality imaging in the diagnosis of a large accessory papillary muscle

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ABSTRACT

An accessory papillary muscle is an uncommon congenital anomaly usually found incidentally on routine cardiac imaging. While frequently asymptomatic, it is occasionally associated with mitral regurgitation, left ventricular dynamic outflow obstruction and hypertrophic cardiomyopathy[1] and it is important to differentiate it from other pathological processes including papillary fibroelastoma, left ventricle thrombus, hemangioma, a single papillary muscle with a parachute mitral valve and a left ventricle false tendon.

The clinical implication of these findings varies according to the degree of left ventricular out flow obstruction, location and pathology. We report a case that underscores the importance of multimodality imaging in the diagnosis and differentiation of an accessory papillary muscle from other intracardiac masses.

KEYWORDS: Accessory papillary muscle, cardiac mass, imaging

INTRODUCTION

Papillary muscles (PMs) vary in number, shape, thickness and location, with three PMs located in the right ventricle and two located in the left. Variations in PM distribution are related to incomplete delamination of trabecular ridge in the ventricles [2] and are classified on the numbers of heads (I-III) and type of insertion (A-C).[3] Usually an incidental discovery on echocardiogram imaging, it may occasionally be associated with mitral regurgitation or obstruction of the left ventricular outflow tract.[4] An increase in number or mass of papillary muscles may be seen in patients with hypertrophic cardiomyopathy (HCM) and is considered a variant of HCM.[4] Recognition of this congenital anomaly is fundamental in distinguishing it from other intracardiac pathologies like papillary fibroelastoma, left ventricular thrombus, a single papillary muscle with a parachute mitral valve and a left ventricle false tendon that may warrant a different treatment approach. Here, we present a clinical scenario whereby we diagnose a large accessory papillary muscle using its characteristic radiographic findings in accordance with various imaging modalities.

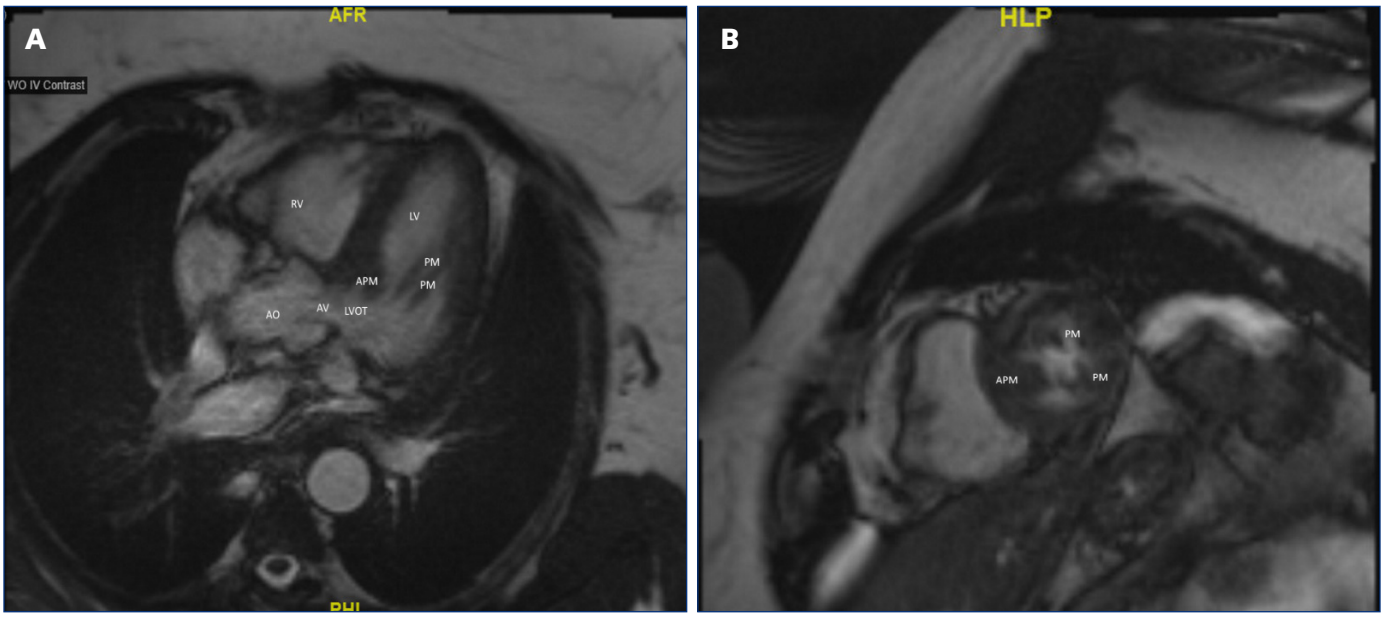
CASE REPORT

A 66-year-old woman with a history of diabetes mellitus, hypertension and obesity was referred to the cardiology outpatient clinic by her primary care physician following an abnormal stress test. She was incidentally found to have an abnormal electrocardiogram, prompting an exercise tolerance test (ETT) where she exercised for six minutes and reached 88% of the maximum predicted heart rate with borderline ST depressions in the inferolateral leads. She denied any history of coronary artery disease or stroke. She exercises twice weekly. Medication regimen included: simvastatin, lisinopril, and metformin. At the time of her outpatient visit blood pressure was 120/60mmHg, pulse of 80, body mass index of 30.20 kg/m^2, and oxygen saturation of 98% on room air. She had unremarkable physical exam, including cardiac exam. An electrocardiogram revealed sinus rhythm with a non-specific intraventricular conduction delay, and laboratory tests showed normal complete blood count, basic metabolic panel, thyroid stimulating hormone and liver function. Due to the abnormal ETT, she had a transthoracic echocardiogram (TTE) ordered which showed normal left ventricular size and systolic function with no regional wall motion abnormalities. An echodense mass was found with independent mobility in the mid to upper septum measuring 1.5 x 1.1 cm without significant left ventricular outflow tract (LVOT) gradient, concerning for fibroelastoma. The decision was made to proceed with a cardiovascular magnetic resonance (CMR) and transesophageal echocardiogram (TEE) for further evaluation of the underlying mass.

A CMR with Gadavist gadolinium-based contrast showed an indeterminate 1.5 x 1.2cm left ventricular mass with elongated morphology which demonstrated similar signal characteristics to the adjacent papillary muscles; however, the motion was different on cine images requiring a TEE for better differentiation and to exclude fibroelastoma. **(Figure 1)**. The TEE demonstrated a contractile echodense mass attached to the interventricular septum on the left ventricular side with no LVOT gradient crossing the left ventricular chamber, unlikely to be a fibroelastoma and likely representing a large accessory papillary muscle **(Figure 2).**

The patient then underwent a nuclear stress test, showing normal myocardial wall thickening with a left ventricular ejection fraction of 60% and no signs of underlying ischemia.

Figure 1. A cardiovascular magnetic resonance with Gadavist Gadolinium-based contrast images
A. Cine steady-state free precession (SSFP) sequence, 3 chamber views showing an accessory papillary muscle (APM) in systole
B. Cine SSFP sequence, short axis view showing an APM during systole.
RV: Right Ventricle, **LV:** Left Ventricle, **LVOT**: Left Ventricular Outflow Tract, **AO**: Aorta,
AV: Aortic Valve, **APM**: Accessory Papillary Muscle. **PM:** Papillary Muscle



DISCUSSION

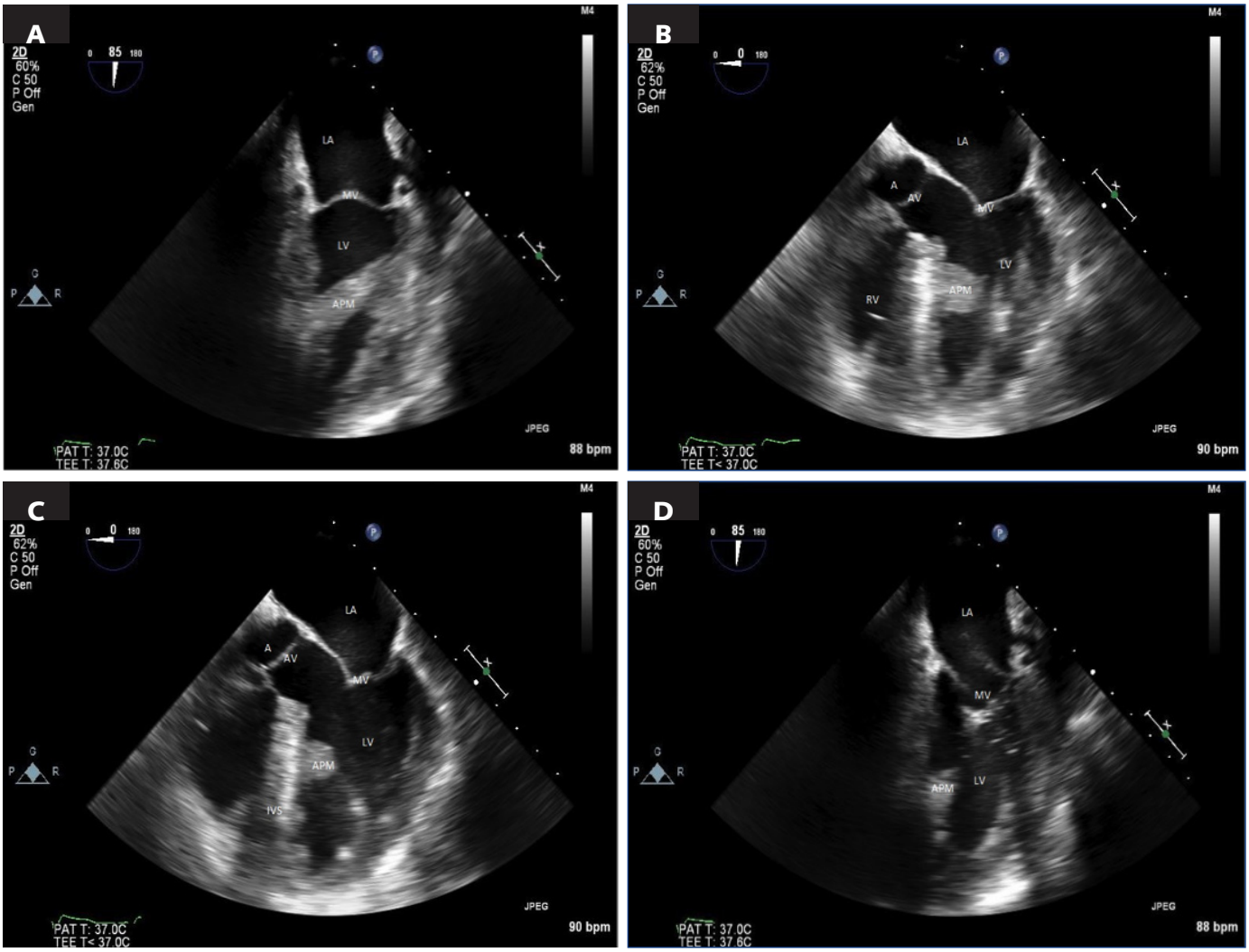
An accessory papillary muscle is an uncommon congenital anomaly, usually found incidentally in asymptomatic patients. However, it can cause mitral valve regurgitation and left ventricular outflow tract obstruction. It is often misinterpreted for a papillary fibroelastoma, left ventricular thrombus, hemangioma, left ventricular false tendon or a single papillary muscle with parachute mitral valve (PMV). A single papillary muscle with PMV is a rare congenital anomaly of the mitral valve apparatus where the chordae tendinea converge to the centrally placed single papillary muscle instead. It is usually associated with aortic stenosis (32%), atrial septal defect (54%) and hypoplastic heart (19%) and when isolated accounts for less than 1% of PMV.[5]

However, left ventricular false tendons are cords that do not attach to the leaflets of the atrioventricular valves and instead connects papillary muscles to each other or to the ventricular wall, interventricular septum, or merely pass between two points on the wall itself. It has been suggested that they might be implicated as a source of idiopathic left ventricular tachycardia.[6] In contrast, a papillary fibroelastoma is usually a small (usually <9 mm), well delineated, pedunculated, non-contractile mass with a characteristic shimmer or vibration at the tumor-blood interface on TEE, ascribed to the finger-like projections of the tumor.[7] On CMR, it appears as a hypointense or intermediate signal in T1-weighted sequences, intermediate signal in T2-weighted sequences and hypointense in the cine-MR steady-state free precession (SSFP) sequences and is not usually enhanced after the administration of contrast.[8] While an accessory papillary muscle will obviously have signal characteristics identical to a normal papillary muscle on CMR, a left ventricle thrombus may include an accompanying regional wall motion abnormality, especially with an apically located thrombus, along with a distinct thrombus margin with jagged edges and movement separate from the underlying endocardium with a higher echodensity reading compared to the myocardium itself.[9]

In this case, an accessory papillary muscle resembled a large fibroelastoma on transthoracic imaging given its independent motion from the other papillary muscles. However, it demonstrated similar characteristic signals to the papillary muscles on cine images of CMR and on TEE. The mass appeared as an echodense mass attached to the interventricular septum with similar morphology to the papillary muscle and without signs of left ventricular obstruction.[1]

While echocardiogram is usually the initial imaging modality, cross sectional imaging with CT or CMR can provide a detailed anatomical picture and is also helpful in the assessment of associated anomalies.[1] A CMR is useful in the evaluation of any physiological consequences of these abnormalities, especially in the context of emerging studies suggesting a hypertrophic cardiomyopathy variant for those who have isolated papillary muscle hypertrophy.[10]

Figure 2. Transesophageal Echocardiogram images.
A. Mid-esophageal long axis view (ME-LAX) showing an accessory papillary muscle (APM) in systole.
B. ME-LAX view showing an APM during diastole.
C. ME- two chamber view showing an APM during systole.
D. ME -two chamber view showing an APM during diastole.
LA: Left Atrium, **MV**: Mitral Valve, **LV**: Left Ventricle, **AO**: Aorta, **AV**: Aortic Valve, **RV**: Right Ventricle, **IVS**: Interventricular Septum, **APM**: Accessory Papillary Muscle.



CONCLUSION

This case illustrates the role of multimodality imaging in the diagnosis of different intracardiac masses and providing insight into future management. There is very scarce data in the literature about long term outcomes in patients with asymptomatic variants of these abnormalities, and future studies are needed to better prognosticate and determine the best course of treatment for these underlying medical conditions.[11]

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