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A Rare Case of Simultaneous Cardiac Papillary Fibroelastoma and Cardiac Myxoma

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ABSTRACT

A 55-year-old female was seen for a cardiac workup due to anxiety and a cardiac tumor was subsequently found on an echocardiogram. The tumor measured 3x2cm in the left atrium and was thought to be cardiac myxoma. Gross and histological findings revealed simultaneous cardiac myxoma and papillary fibroelastoma. The latter caused severe mitral regurgitation when excised requiring a mitral valve replacement. Primary cardiac tumors are rare, and most are benign with over 50% being cardiac myxomas. We experienced an exceptionally rare case of two separate simultaneous cardiac tumors in this patient.

Introduction:

Primary cardiac tumors (PCTs) are relatively rare, and it is even rarer for a patient to have two separate types of PCTs simultaneously. The two most common are cardiac myxoma and papillary fibroelastoma (PFE). There has been debate over the years as to which of these two PCTs is more common. A meta-analysis that investigated incidence and prevalence rates of various cardiac tumors found myxomas to be 58.14% of the PCTs [1]. Given the extremely rare occurrence of combined PCTs, a literature search yielded only eleven such cases on PubMed.

Case:

A 55-year-old female evaluated initially for anxiety, had a cardiac workup completed. The patient felt well overall but had episodes of lightheadedness and dizziness without syncopal episodes. There was no pertinent previous medical history. Patient denied tobacco or alcohol use. Family history reveals fatal paternal myocardial infarction and healthy siblings. On the physical exam, a holosystolic murmur grade 2/6 was heard at the cardiac apex, with the rest being normal.

The patient initially had a stress test followed by a transthoracic echocardiogram (TTE) which revealed a large 3x2cm hyperechoic mass in the left atrium. This mass was thought to be a simple myxoma attached to the atrial septum [Fig 1] and an ejection fraction of 30%. The patient was scheduled for surgery to remove the PCT. During the surgery, the tumor was observed to be in the left atrial fossa ovalis and partially obstructing the mitral valve. The bulk of the tumor was easily removed with ring forceps, but a sticky fibrous material was found tethered to the chordae tendinea of the mitral valve. Once the fibrous material was removed, the

patient developed severe mitral regurgitation. At this point the native mitral valve was replaced using a bioprosthetic valve. A follow up TTE was done a few days later showed a normal left atrium [Fig 2] and no mitral regurgitation. After an uneventful hospital course, the patient was discharged home. Since the patient was not from the area she was lost to follow-up.

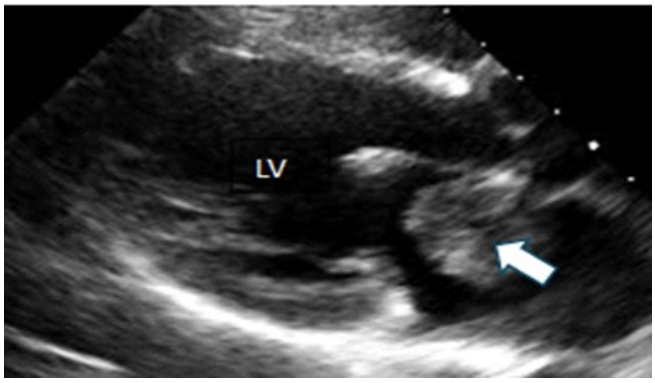


Figure 1: Transthoracic echocardiogram (long axis parasternal view) shows cardiac myxoma in left atrium in close proximity to native mitral valve (arrow).

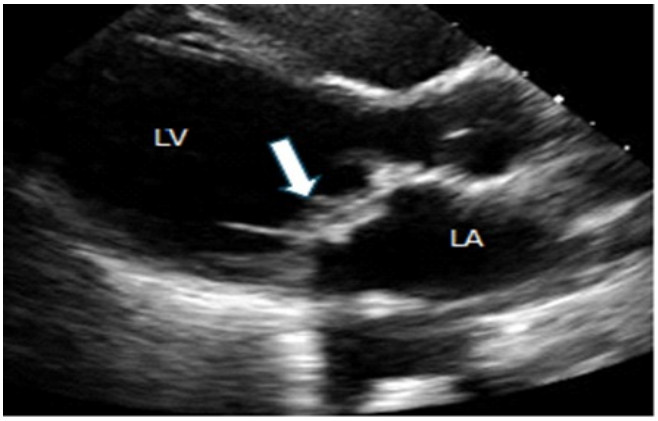
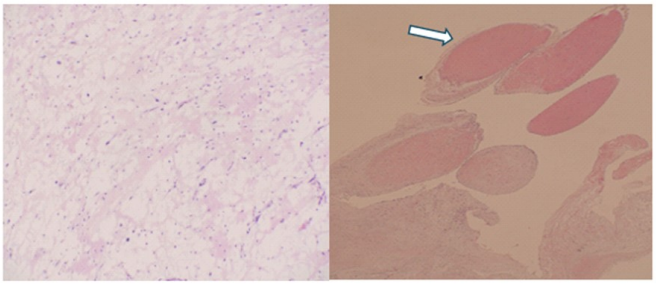
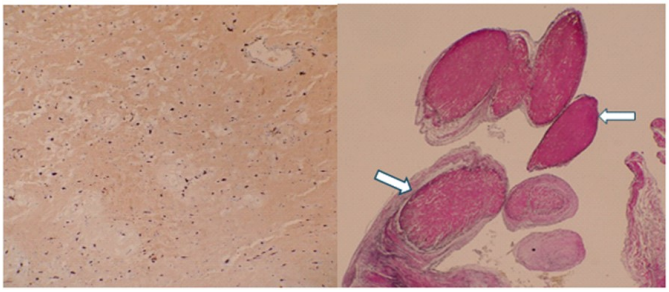


Figure 2: Transthoracic echocardiogram (long axis parasternal view) after removal of cardiac myxoma and associated cardiac papillary fibroelastoma. Bioprosthetic mitral valve replacement (arrow) is installed.

Hematoxylin and eosin staining of a specimen showed two separate histological morphologies. First, there was a hypocellular tumor within a myxoid matrix consistent with myxoma [Fig 3]. Second, there was avascular collagenous component with branching papillae consistent with a PFE [Fige 4]. Immunohistochemical stains, calretinin and elastin, were performed on both the myxoma and PFE and confirmed the diagnosis [Fig 5,6].



Figures 3 and 4: Hematoxylin and eosin stain (40x) shows a hypocellular myxomatous matrix of the cardiac myxoma (left) and branching papillae (arrow) composed of collagen and elastic fibrous tissue of the papillary fibroelastoma (right).



Figures 5 and 6: Calretinin IHC stain showing strong, diffuse cytoplasmic and nuclear staining of Myxoma (left) and elastic stain of the papillary fibroelastoma showing black elastic fibers at the periphery (arrows) of collagenous structures (Right).

Discussion:

Myxomas are typically found in the left atrial fossa ovalis suspended by a stalk about 72%-92% of the time. Patients have a mean age of 50-years-old with females accounting for 70% of the total cases [2,3]. PFE generally originates from endocardial portion of valves commonly on aortic and mitral valves [4,5]. A study ran by the Japanese Association of Thoracic Surgery found the amount of PCTs diagnosed has been increasing yearly [6]. This in-

crease is most likely due to better imaging modalities and higher rates of imaging which may lead to incidental findings.

PCTs are typically diagnosed through either TTE or found incidentally on a thoracic CT scan [3]. If more detail is required, transesophageal echocardiogram (TEE) can provide greater detail [3]. Cardiac Computed Tomography (CCT) or Cardiac Magnetic Resonance (CMR) is sometimes used after an echocardiogram if additional details are required but might not be needed and have their disadvantages [2]. PCTs regardless of type should be excised surgically as there is increased risk for embolization leading to transient ischemic attacks or ischemic strokes [2,4,5].

Conclusion:

At the first evaluation, the patient presented with few symptoms. A tumor was detected only after an echocardiogram ran during a subsequent cardiac workup. During the surgical removal of that tumor, it was discovered that there was more than just a simple cardiac myxoma. The tumor morphology was different, and histology identified two separate tumors. We observed a unique and rare case of simultaneous cardiac myxoma and papillary fibroelastoma.

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