

## CLINICAL VIGNETTE

# Papillary Fibroelastoma of the Mitral Valve: A Source of Acute Stroke

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A 67-year-old male with hypertension, dyslipidemia, type 2 diabetes, and an active smoker presented for cardiology consultation after undergoing an echocardiogram following a stroke 6 months prior with visual field loss. He reported acute loss of vision in his lower right visual field and was diagnosed clinically with an acute stroke by neurology. He was started on a statin but was not started on anti-platelet therapy at the time. He underwent subsequent testing for the source of stroke. Carotid ultrasound showed moderate atherosclerotic plaque at the carotid bulbs and left common carotid artery. Echocardiogram showed normal left ventricular systolic function including normal wall motion, grade 1 diastolic dysfunction and normal filling pressures. Also noted mitral annular calcification with thickened mitral valve leaflets and a small, mobile calcified mass attached to the posterior mitral valve leaflet. A vegetation versus a focal calcification or a papillary fibroelastoma (PFE) could not be excluded.

The patient had no clinical symptoms of systemic infection. His primary care physician obtained bacterial blood cultures to rule out endocarditis, which were negative, and he was referred for cardiac evaluation.

On presentation to cardiology, he was afebrile with a blood pressure of 115/75, pulse of 84/min, respiratory rate 12 breath/min, and 100% oxygen saturation on room air. Cardiac exam revealed a regular rate and rhythm with a normal S1 and S2, no murmur, rubs, or gallops. JVP was normal and lungs were clear to auscultation. Distal pulses were 2+ and he did not have lower extremity edema. EKG showed sinus rhythm at a rate of 76 bpm.

The patient expressed concern about the persistence of his visual symptoms and how further strokes could be prevented. He was started on aspirin, 81 mg daily. His lipid panel was reviewed – LDL 56 mg/dL, HDL 39 mg/dL, TG 227mg/dL, and due to diabetes and an elevated Atherosclerotic Cardiovascular Disease (ASCVD) risk score, he was urged to switch to a higher intensity statin. The patient declined due to fears of side effects. He was also strongly urged to quit smoking and adopt a low-fat diet with increased exercise.

For further evaluation of the mitral leaflet abnormality, a transesophageal echocardiogram was attempted but was unsuccessful due to oxygen desaturation. Twenty-four hour holter monitor revealed paroxysmal atrial fibrillation with an

intermittent rapid ventricular rate and transient episodes of ventricular tachycardia.

CHA<sub>2</sub>DS<sub>2</sub>-VASc Score for Atrial Fibrillation was 6 equating to a nearly 10% annual risk of stroke. He was started on warfarin and metoprolol for rate control. Cardiac MRI confirmed a 3 mm calcified mass on the posterior mitral leaflet consistent with a PFE. An incidental finding of the MRI was a lateral infarct with preserved left ventricular systolic function with ejection fraction of 58%.

### Discussion

Papillary fibroelastoma (PFE) is a primary cardiac tumor most often found on left sided heart valves (aortic 36%, mitral 29%).<sup>2</sup> It appears to have mobile frond-like arms emanating from a central core and is likened to a sea anemones. There is controversy regarding incidence with some studies indicating it is the most common cardiac tumor and others citing it as the second most common cardiac tumor found in adults after left atrial myxoma.<sup>3</sup> There is higher incidence in males with the average age of diagnosis around 60 years old. It is often an incidental finding with no associated symptoms. However, in some instances PFEs can be associated with stroke or transient ischemic attack (TIA) due to embolic tumor fragments or thrombus accumulation within the fronds. Myocardial infarction and sudden cardiac death have also been reported from coronary ostium obstruction by the tumor.<sup>3</sup>

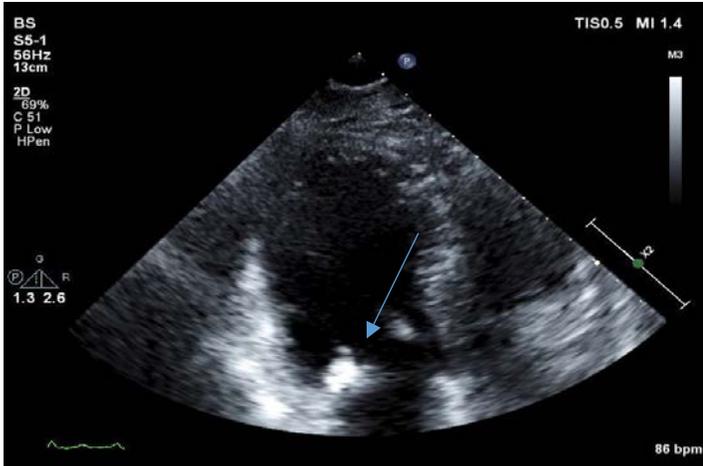
Generally asymptomatic, small non mobile PFEs are left alone with periodic monitoring recommended via echocardiogram. Anticoagulation to prevent systemic embolization of the tumor contents can be recommended. Surgery is strongly advised in the setting of symptoms or with large, pedunculated tumors (>1 cm).<sup>2</sup> Valve sparing shave excisions of PFEs generally have good outcomes in experienced centers.<sup>2</sup> Anticoagulation may be recommended in the interim waiting period as it has been for our patient who remains hesitant regarding surgical intervention.

Our patient's PFE is small (3 mm) and atrial fibrillation remains an important confounding source of his stroke, which supports conservative therapy until his ischemic work up is completed. Should he require open heart surgery, the PFE will be excised. If he opts out of surgical intervention, long-term anticoagulation will be maintained due to atrial fibrillation and

continued potential with risk of subsequent systemic embolization from the PFE.

## Figures

**Figure 1.** 2D Echo image in the apical view of the posterior mitral leaflet. There is moderate mitral annular calcification as well as a small mobile pedunculated calcified mass noted.



**Figure 2.** 24 hour holter montitor strips showing paroxysmal atrial fibrillation (a) and intermittent non-sustained ventricular tachycardia (b).



**Figure 3.** Cardiac MRI image of a 50 year female with a history of chest pressure diagnosed with a papillary fibroelastoma.<sup>1</sup>

## REFERENCES

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