Mitral and tricuspid valve papillary fibroelastomas: A rare cause of multiple intracardiac masses

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A 55-year-old woman with no cardiac history presented with increasing shortness of breath on exertion, worsening cough and orthopnea for three weeks. A transthoracic echocardiogram identified two intracardiac masses. Transesophageal echocardiography demonstrated a 1.8 cm × 1.5 cm mass attached to the base of the posterior tricuspid leaflet (Figure 1A) and a 1.0 cm × 0.5 cm mass attached to the atrial side of the left mitral leaflet (Figure 1B). Both masses displayed indistinct surface features and neither was attached to the atrial wall or interatrial septum. No other intracardiac pathology was identified. The differential diagnosis included primary cardiac tumour, metastatic malignancy, thrombus or vegetation. After an extensive workup, the patient ultimately went on to have surgical excision of these masses. Surgical pathology confirmed the diagnosis: mitral and tricuspid valve cardiac papillary fibroelastomas (CPFs).

CPFs are the second most common primary cardiac tumour in adults. They are benign avascular tumours made of papillary elastin fibrils in a hyaline stroma covered with a single layer of endothelium. These tumours occur most frequently on the aortic and mitral valves, and infrequently on the tricuspid and pulmonic valves. On echocardiography, CPFs are typically small, well-demarcated, highly mobile masses attached to the endocardium by a pedicle. They can have a

highly refractive appearance, often with areas of echolucency and a 'speckled' appearance around the perimeter. Multiple CPFs are an exceedingly rare finding. While typically asymptomatic, CPFs can present with stroke, sudden death, myocardial infarction, heart failure, syncope, pulmonary embolism or peripheral infarction. To date, no recurrences of CPFs after surgical excision have been reported, and the long-term prognosis of these patients appears to be excellent (1-4).

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Figure 1

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