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Cardiac vignette: Giant atrial myopathy

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A 72-year-old male presented to our institution with worsening dyspnea, orthopnea and decreased exercise tolerance. This is on a known background of an idiopathic cardiomyopathy, Atrial Fibrillation (AF) and cardiovascular risk factors. On examination, the patient appeared fluid overloaded. Chest examination revealed coarse crackles to bilateral mid-zones with an audible wheeze. Precordial examination revealed a holosystolic murmur at the apex with radiation to the axillary regions. He had evidence of peripheral to the sacrum. A diagnosis of decompensated Congestive Cardiac Failure (CCF) was made and he was commenced on appropriate inpatient heart failure treatment. Chest radiograph showed gross cardiomegaly with evidence of mild pulmonary venous congestion. A transthoracic echocardiogram revealed severely dilated bi-ventricular sizes with normal left ventricular ejection fraction but severely impaired right ventricular ejection fraction. Strikingly, there was evidence of severely dilated bi-atrial size. Left atrial volume was indexed at 438.3 ml/m2 relative to body surface area. There was also presence of severe mitral regurgitation in the setting of non-coaptation of the mitral valve leaflets. These findings are consistent with a Giant Atrial Myopathy (GAM) in the setting of an idiopathic dilated cardiomyopathy. Giant atrial myopathies are typically associated with rheumatic valve disease, however there are several case reports demonstrating its presence in primary cardiomyopathies. This atrial disease forms a composite substrate for AF, CCF and thrombus formation. Management strategies in this disease are often supportive, with the condition often portending a poor prognosis.

Biography

Shaun Khanna has completed his MBBS from Bond University and is currently pursuing his Master of Medicine at University of Sydney, Australia. He is also working at Blacktown Hospital as a Medical Officer and as a Conjoint Lecturer with the University of Western Sydney. His research interest is in arrhythmias and cardiovascular imaging.

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