Cardiac Vignette: Giant Atrial Myopathy

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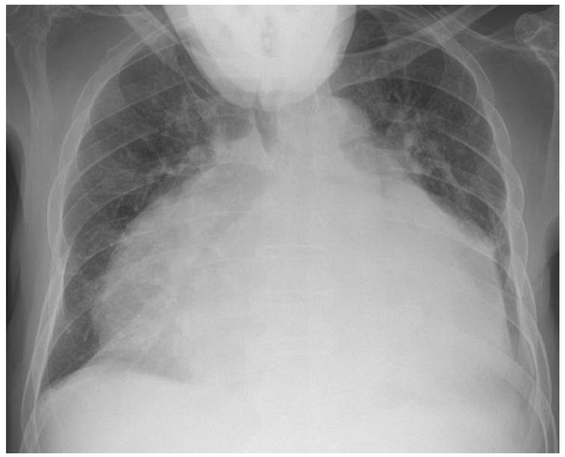
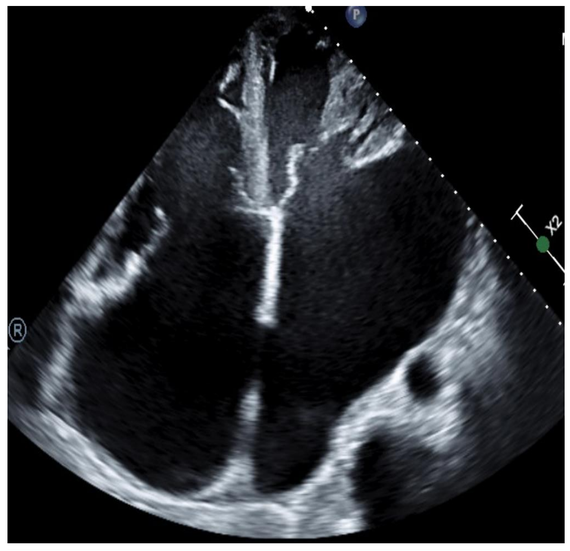
Clinical History

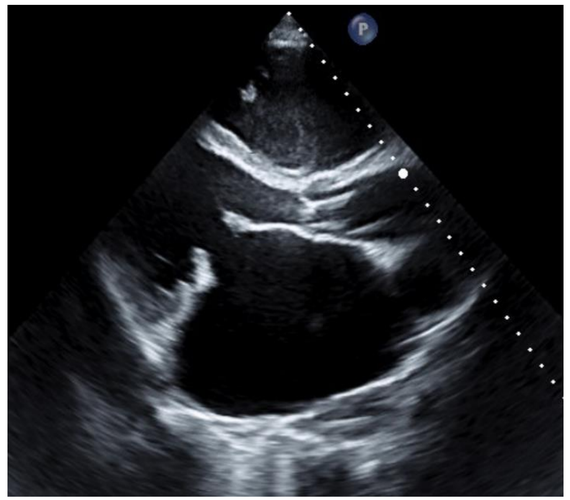
A 72 year old male presented to our institution with worsening dyspnoea, orthopnea and decreased exercise tolerance. This is on a known background of an idiopathic cardiomyopathy, atrial fibrillation (AF), and cardiovascular risk factors.

On examination, the patient appeared in a fluid overload state. Chest examination revealed coarse crackles to bilateral mid-zones with an audible wheeze. Precordial examination revealed a holo-systolic murmur at the apex with radiation to the axillary region. He had evidence of peripheral oedema 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. **See Figure 1.**



A transthoracic echocardiogram revealed severely dilated bi-ventricular sizes with normal left ventricular ejection fraction but with 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/m^2 relative to body surface area. **See Figure 2 & 3.**





Figures 2 & 3: Severely dilated left atrium and right atrium seen in apical 4 chamber and parasternal long views on transthoracic echocardiogram

There was also presence of severe mitral regurgitation in the setting of non-coaptation of the mitral valve leaflets. **See Figure 4.** These findings are consistent with a **Giant Atrial Myopathy** in the setting of an idiopathic dilated cardiomyopathy. During the inpatient stay, he also developed neck pain which was like attributed to the mass effect of the giant left atrium. Cardiothoracics were consulted for valve replacement in light of ongoing decompensated congestive cardiac failure. Unfortunately, the patient developed staphylococcus aureus bacteremia and possible infective endocarditis (IE). He was but unable to be appropriately investigated for IE given his symptomatology and deconditioned state. He passed away during this hospital admission.

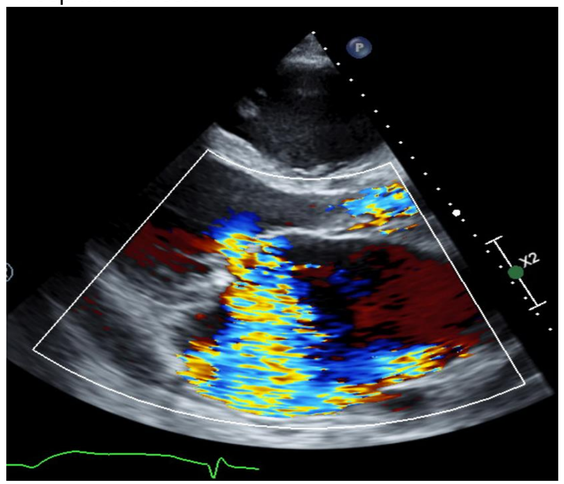


Figure 4: Severe Mitral Regurgitation

Discussion

Giant Atrial Myopathies are rarely seen in the clinical setting, with a reported incidence of 0.3%, typically secondary to rheumatic heart disease with fewer reports of secondary to cardiomyopathies. They carry an overall significant cardiovascular burden with the composite substrate for atrial fibrillation, congestive cardiac failure, atrial thrombus formation and subsequent systemic embolization. [1-2] Whilst intra-cardiac complications are historically monitored, extra-cardiac manifestations should also be anticipated, as a giant left atrium has the ability to cause compression to the posterior mediastinal structures such as the oesophagus. There are several techniques to evaluate atrial myopathies including simple chest radiographs, transthoracic & transesophageal echocardiography and cardiac MRI. [1] There are cases of surgical correction of giant left atria to minimize complications, through mitral valve surgeries and left atrial plications, but with reported minimal long term benefit. [3] For example, *Yuda et al* compared the Maze procedure in patients with giant left atria (Group A) to those without giant left atria (Group B) in restoration of normal atrial electrical activity. At one year, normal sinus rhythm was present in 53% of Group A compared to 75% of Group B. [4] Whilst management is typically aimed at heart failure therapy, anticoagulation, control of tachy-arrhythmias and symptomatic surgical treatment, the condition often portends a poor prognosis.

Conclusion

Giant Atrial Myopathies carry a significant burden of cardiovascular morbidity and mortality. Further large scale studies are required to decipher the most effective management strategy for these patients.

References

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