Case report

Multimodal imaging of an atypical left atrial myxoma

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Case

A 53-year-old man presented with a 12-month history of increasing shortness of breath, decreasing exercise tolerance, peripheral oedema, abdominal distension and paroxysmal nocturnal dyspnoea.

He was a previously fit and well man with no co-morbidities. His only cardiac risk factor was of previous tobacco use.

On examination, he had signs consistent with biventricular heart failure and a pansystolic murmur. His Brain Natriuretic Peptide (BNP) was raised at 4800.

His ECG showed sinus tachycardia, bifid-peaked P waves and incomplete right bundle branch block.

Given the history, examination and the raised BNP, he was referred directly for echocardiogram by his general practitioner and a 6\( \times \)5 cm mass was found in his left atrium.

The homogenous mass was seen to almost completely fill the left atrial cavity. It obstructed the mitral valve in diastole causing at least moderate mitral regurgitation in systole. There was secondary pulmonary hypertension with an estimated right ventricular systolic pressure of 65 mmHg.

The mass was further interrogated by the complimentary imaging modalities of transoesophageal echo (both 2- and 3D), cardiac MRI (CMR, Siemens Avanto 1.5-T, Erlangen, Germany) and coronary angiography.

Transoesophageal echocardiography (TOE) delineated an atypical anatomical location of a clefted-shaped mass, which was clearly attached by a stalk to the posterior wall of the left atrium (Figures 1 and 2).

Given that the mass was atypical, a CMR was done to rule out an infiltrative process.

Accurate volume and function assessment, in addition to mass size and location, was outlined using cine CMR. Left Ventricular (LV) function was moderately impaired with an estimated ejection fraction of 38%.

CMR tissue characterization using delayed contrast enhancement following the administration of gadolinium demonstrated a heterogenous composition of the mass, typical of a myxoma, with no myocardial infiltration (Figure 4).

Coronary angiography demonstrated the typical ‘myocardial blush’, with vascularity arising from the left circumflex artery (Figure 3).

Inpatient surgery was carried out. The mass was confirmed to be attached to the posterior wall of the left atrium just above the posterior mitral valve leaflet. The valve itself was unaffected and functioned normally. A 5.1\( \times \)4.4\( \times \)3.0 cm pedunculated gelatinous mass weighing 43 g was resected with the stalk shaved down to the endocardium (Figure 4). Histology confirmed an atrial myxoma.

Discussion

Primary tumours of the heart are extremely rare with a reported autopsy prevalence of only 0.001–0.03%.\(^1\) Of these, the majority is benign myxomas.\(^4\)

Myxomas are typically benign tumours and cause symptoms through cardiac obstruction (cavity, valve or outflow tracts), embolization and constitutional upset.
The majority of myxomas arise from the interatrial septum (80%). Attachment to the posterior wall is rare and should prompt consideration of an alternative diagnosis particularly of malignancy. Imaging is obviously the mainstay for diagnosis of cardiac masses. Traditionally, transthoracic and 2-D TOE were the initial and best modalities for mass characterization and differentiation. 3D TOE and CMR have now surpassed these to become the modalities of choice as they provide superior quantitative and qualitative information on the mass location, tissue composition and myocardial and valvular function, facilitating precise operative planning by the surgeon, especially in this case where the location was atypical.

Figure 1. TOE with colour flow mapping demonstrating moderately severe functional Mitral regurgitation (2-D without CFM image shown for comparison).

Figure 2. 3D TOE image demonstrating the clefted mass, its attachment to the posterior wall of the left atrium and prolapse into the left ventricle during diastole.
Conclusion

Primary tumours of the heart are rare and the majority is benign myxomas. In the assessment of an atypical cardiac mass, novel cardiac imaging modalities are complementary and each contributes uniquely towards the diagnosis and surgical planning.

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Conflict of interest: None declared.

References


Figure 3. Coronary angiography demonstrating neovascular supply to the myxoma via the left circumflex artery (‘Tumour Blush’).

Figure 4. Post-gadolinium mid-short axis CMR view showing heterogenous enhancement of the myxoma but normal myocardium enhancement. The insert at the bottom right shows the gelatinous-clefted appearance of the resected myxoma.