Novel contrast echocardiographic features of cardiac myxoma with cystic degeneration: a case report

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Case presentation

A 63-year-old lady presented to our emergency department with a 6-month history of chronic cough and progressive shortness of breath on exertion with New York Heart Association class III heart failure. She also reported episodic non-exertional chest pain and bilateral ankle swelling. On examination, she was afebrile, with blood pressure 121/77 mm Hg, heart rate 97 beats/min and respiratory rate 16 breaths/min. Cardiac examination revealed an elevated jugular venous pressure at 4 cm above the sternal notch with a right parasternal heave. A loud pulmonary second heart sound and a diastolic murmur were detected. The murmur was variable with posture and was best heard at the apex in the right lateral position on end expiration. Respiratory examination showed clear lung fields, and mild pitting ankle oedema was noted. Chest X-ray revealed enlarged bilateral pulmonary trunks. Electrocardiogram showed sinus rhythm of 94 beats/min, right axis deviation and tall right precordial R waves. Inflammatory markers were within normal range. However, levels of highly sensitive troponin I and N-terminal prohormone of brain natriuretic peptide were elevated (85 ng/L and 1786 ng/L, respectively). Bedside transthoracic echocardiography screening in the emergency department revealed a large left atrial mass. The clinical impression was pulmonary hypertension secondary to a left-sided cardiac tumour.

The patient had a history of left parotid pleomorphic adenoma for which she had undergone partial excision 15 years ago. She had no history of cardiopulmonary disease and no family history of cardiac tumours.

Differential diagnosis

The top differential diagnosis of primary cardiac tumour was cardiac myxoma but other primary malignant tumours such as sarcoma and secondary cardiac tumours were possible. Given the clinical history, absence of fever and normal inflammatory markers, an infective process was deemed unlikely.

Investigations

A detailed transthoracic echocardiography examination demonstrated a double-barrel–shaped left atrial mass with central echolucent space of 5.8 cm × 2.7 cm in size (Fig 1a). The mass was seen attached to a thick echogenic base at the interatrial septum and was prolapsing through the mitral valve into the left ventricle during each diastolic phase, causing a mild degree of mitral regurgitation and significant mitral inflow obstruction. The average mean gradient across the mitral valve was 15 mm Hg (Fig 1b). Bi-atrial enlargement was seen. The right ventricle was dilated with preserved systolic function. There was a significant degree of pulmonary hypertension and right ventricular systolic pressure was 87 mm Hg. Left ventricular size and systolic function were normal. A contrast echocardiographic study was performed by intravenous administration of SonoVue (Bracco Diagnostics Inc, Milan, Italy) and revealed an initial contrast enhancement at the base of the stalk (Fig 1c), followed by delayed contrast enhancement of the central echolucent space (Fig 1d and 1e) and subsequent early contrast washout prior to the cardiac chambers (Fig 1f).

Urgent in-house cardiothoracic surgical consultation was obtained and immediate open-heart excision of the cardiac tumour was performed. Intraoperative findings showed a large left atrial cystic mass with an internal solid component and wide-based stalk attaching to the interatrial septum. The mitral valve appeared normal. The left atrial mass, along with the stalk and part of the involved interatrial septum, was resected en bloc. No significant mitral regurgitation was detected with saline testing and there was no interatrial flow detected on intraoperative transoesophageal echocardiography following resection. The patient was successfully decannulated and the sternum was closed uneventfully.

Gross examination of the specimen showed a solid cystic tumour measuring 5.5 cm × 4.5 cm × 2.5 cm with a base measuring 2.5 cm × 1.5 cm (Fig 2a). The tumour was carefully cut open to

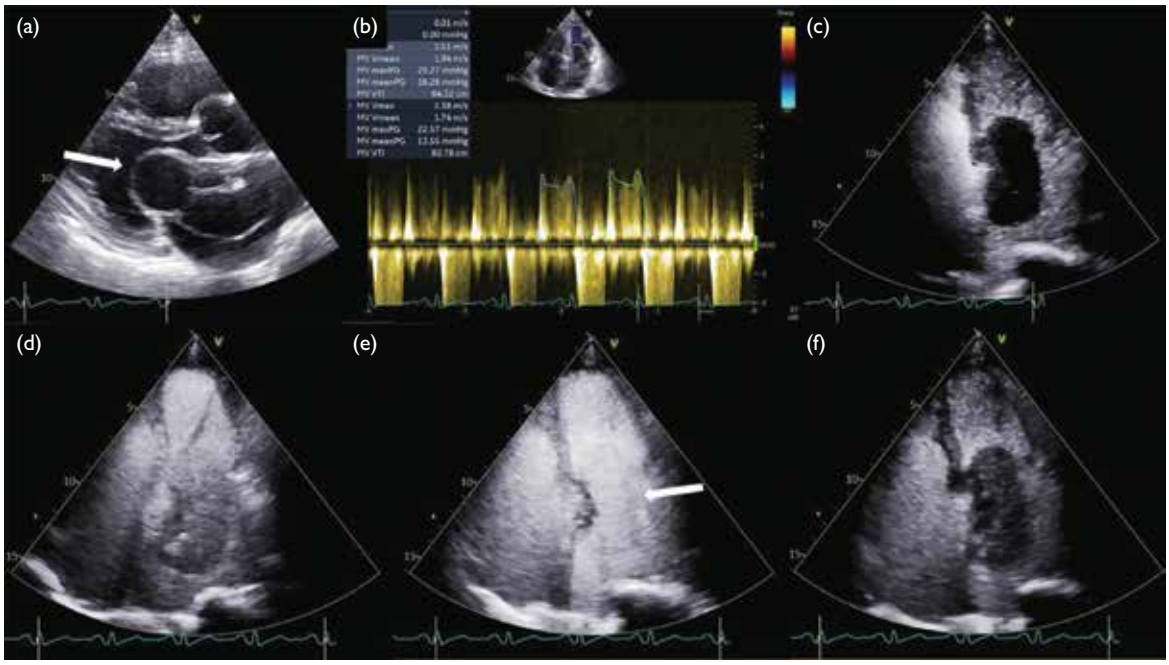

FIG 1. Two-dimensional transthoracic echocardiographic examination of the cardiac mass of the patient. (a) Double-barrel–shaped left atrial myxoma with central echolucent space (arrow). (b) Doppler mitral inflow showing significant gradient across mitral valve. (c) Early contrast phase showing enhancement of the stalk of cardiac myxoma. (d) Delayed contrast enhancement of echolucent space. (e) Homogeneous opacification of previous echolucent space of the cardiac myxoma (arrow). (f) Early contrast washout prior to the cardiac chambers

reveal multilocular surfaces with small gelatinous semitranslucent areas. Serially sectioned specimens showed prominent cystic change with solid myxoid areas adjacent to the base (Fig 2b). Microscopic examination showed thick-walled blood vessels at the stalk of the mass (Fig 2c). The tumour cells exhibited a classic concentric arrangement around capillary and a halo of matrix around cellular clusters (Fig 2d). Sampling from the cystic area revealed morphology similar to the rest of the tumour. Immunostaining for calretinin showed positive nuclear staining in tumour cells (Fig 2e). Overall histopathological features were compatible with cardiac myxoma.

Management

The patient made an uneventful postoperative recovery and pre-discharge echocardiogram showed no residual mass, significant mitral regurgitation or pericardial effusion. She did not report any chest pain or shortness of breath. She was discharged uneventfully on postoperative day 7.

Follow-up

The patient was followed up in the outpatient clinic 2 months after discharge. Her exercise tolerance had improved and ankle swelling resolved. She did not complain of any chest pain or shortness of breath. Her chronic cough had subsided.

Discussion

Cystic degeneration of cardiac myxoma is rare. It is caused by foci of myxoma stromal liquefaction resulting in a cyst-like structure with clear fluid content. The stalk of cardiac myxoma is typically dominated by the presence of large, thick-walled and occasionally dysplastic arteries giving rise to the described 'tumour blush' occasionally observed at coronary angiography.[1] Previous histopathological studies of cardiac myxomas have shown that these tumours produce vascular endothelial growth factor that likely induces angiogenesis for tumour growth.[2,3] On microscopic tissue examination of our patient, we also found these thick-walled blood vessels at the stalk of cardiac myxoma (Fig 2c). Vascular communication between the stalk and the fluid content of cardiac myxoma with cystic degeneration has not been described before.

The use of contrast agent in echocardiography is particularly helpful in assessing vascular communications, vascularity of cardiac masses and to differentiate masses from intracardiac thrombi.[4] Compared with contrast computed tomography, contrast echocardiography offers the advantage of capturing the extended real-time contrast enhancement sequence. To the best of our knowledge, only one case report has discussed the feature of cystic degeneration of cardiac myxoma on contrast echocardiography.[5] We have described a novel feature on contrast echocardiography of cystic degeneration of myxoma: initial enhancement of the base of stalk (Fig 1c), followed by delayed contrast enhancement of the central echolucent space (Fig 1d and 1e) and

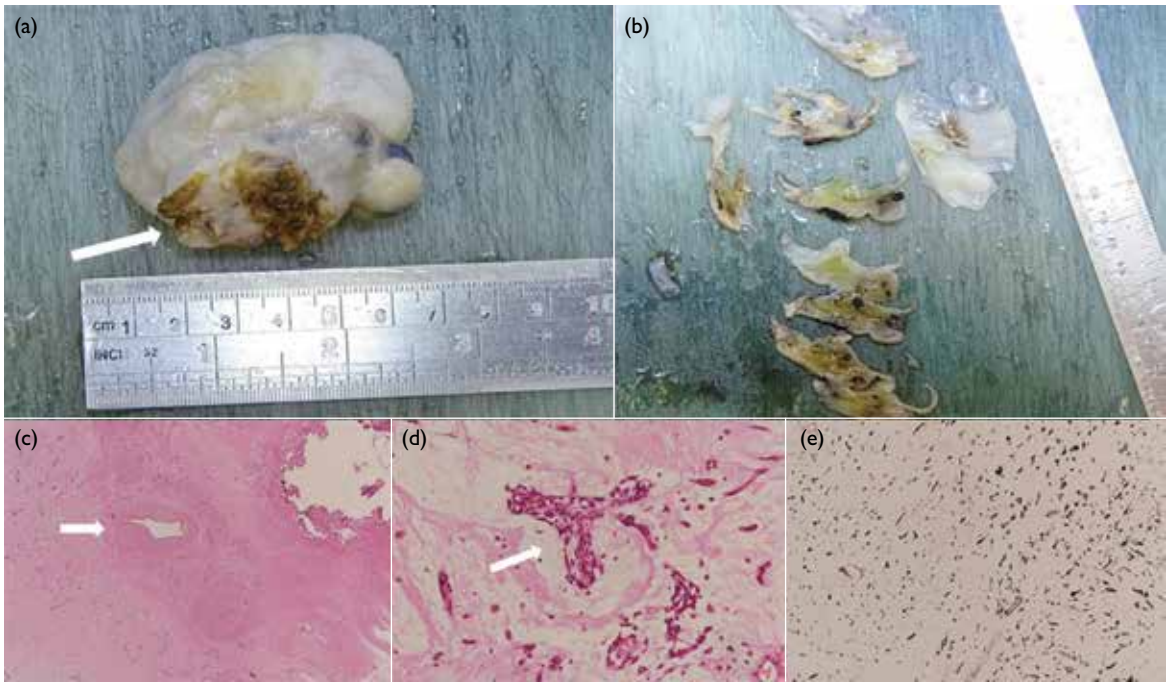

FIG 2. Gross (a and b) and microscopic (c to e) examination of excised cardiac mass of the patient. (a) Gross examination of intact specimen with cauterised base (arrow). (b) Serially sectioned specimen showing prominent cystic change with solid myxoid areas near the base. (c) Thick-walled blood vessels (arrow) at the base/stalk of the lesion (haematoxylin and eosin staining, ×4). (d) Tumour cells showing classic concentric arrangement around capillary (arrow) and classic 'halo of matrix' around cellular clusters (haematoxylin and eosin staining, ×20). (e) Immunostaining for calretinin showing positive nuclear staining in tumour cells (immunohistochemical stain with calretinin, ×4)

early contrast washout prior to the cardiac chambers (Fig 1f). This suggested the presence of some degree of vascularity at the contrast-enhanced base of stalk with vascular communication between the stalk and the echolucent cystic space of the mass.

Conclusion

Left atrial tumour is a rare cause of pulmonary hypertension. Variable diastolic murmur may be detected on physical examination. Cystic degeneration of cardiac myxoma is rare with specific features on contrast echocardiography. Histopathological examination of the cardiac mass provided a histological basis for the unique contrast enhancement pattern on echocardiography.

Author contributions

Concept or design: DPH Lee.
Acquisition of data: All authors.
Analysis or interpretation of data: DPH Lee.
Drafting of the manuscript: DPH Lee.
Critical revision of the manuscript for important intellectual content: All authors.

All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

All authors have disclosed no conflicts of interest.

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Ethics approval

The patient was treated in accordance with the Declaration of Helsinki and has provided consent for all procedures and publication.

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