Case Report

Calcified right ventricular fibroma in an adult
Huanhuan Gao, Shuai Yuan, Zhiqiang Hu, Zhelan Zheng, Yanli Wang, Shengjun Wu

Abstract

**Background:** Cardiac fibromas are benign tumours of the heart and are composed of fibroblasts and collagen. They are one of the most common cardiac tumours encountered in children and adolescents but are rare in adults.

**Case report:** The patient was a 57-year-old man who presented with a two-year history of chest tightness and nausea and was hospitalised in The First Affiliated Hospital of Zhejiang University, Hangzhou, China, in December 2020. The patient was in a good overall condition and denied palpitations, syncope, chest pain and dyspnoea. His blood pressure, heart rate, electrocardiogram and laboratory investigations were within the normal range. However, calcification of the right cardiac border was found incidentally on a chest radiograph.

Transthoracic echocardiography revealed a 4.3 × 0.7-cm area of calcification within the myocardium at the anterior wall of the right ventricle near the apex, but normal right ventricular wall motion (Fig. 1A). The diagnosis of a benign pericardial tumour was confirmed by computed tomography (Fig. 1B), and coronary angiography showed normal coronary arteries (Fig. 1C). Intra-operative transoesophageal echocardiography identified a 3 × 2-cm hyperechoic mass in the apical interventricular groove, stretching to the anterior wall of the right ventricle (Fig. 1D).

**Conclusions:** Pre-operative diagnosis using various imaging modalities and early surgery are key to optimising the prognosis of patients with a cardiac fibroma.

**Keywords:** cardiac fibroma, tumours, transthoracic echocardiography, calcified mass

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Cardiac fibroma is a rare benign tumour in the myocardium and is composed of fibroblasts and collagen. It is the second most common benign cardiac tumour after rhabdomyoma in children but is rarely seen in adults. Cardiac fibroma is usually located in a ventricle or the interventricular septum and typically presents as central calcification, which distinguishes it from a rhabdomyoma. The clinical manifestations of cardiac fibroma are non-specific but include stethalgia, arrhythmia, dyspnoea and even sudden death, depending on its position and size. However, cardiac fibroma may be symptomless and an incidental finding on imaging. Early surgery is key to a favourable outcome in a symptomatic patient.

The study was approved by the Institutional Review Board at The First Affiliated Hospital of Zhejiang University, Hangzhou, China. The procedures were conducted according to the principles of the Helsinki Declaration.

**Case report**

The patient was a 57-year-old man who presented with a two-year history of chest tightness and nausea and was hospitalised in The First Affiliated Hospital of Zhejiang University, Hangzhou, China, in December 2020. The patient was in a good overall condition and denied palpitations, syncope, chest pain and dyspnoea. His blood pressure, heart rate, electrocardiogram and laboratory investigations were within the normal range. However, calcification of the right cardiac border was found incidentally on a chest radiograph.

Transthoracic echocardiography revealed a 4.3 × 0.7-cm area of calcification within the myocardium at the anterior wall of the right ventricle near the apex, but normal right ventricular wall motion (Fig. 1A). The diagnosis of a benign pericardial tumour was confirmed by computed tomography (Fig. 1B), and coronary angiography showed normal coronary arteries (Fig. 1C). Intra-operative transoesophageal echocardiography identified a 3 × 2-cm hyperechoic mass in the apical interventricular groove, stretching to the anterior wall of the right ventricle (Fig. 1D).

Based on the diagnosis, the tumour was scheduled to be resected via standard median sternotomy without cardiopulmonary bypass. Cardiopulmonary bypass is required if invasion into the ventricular wall or the coronary artery is suspected intra-operatively or in the event of any intra-operative haemodynamic deterioration.

At surgery a protruding firm, yellowish mass (3 × 3 × 2 cm) was discovered close to the left anterior descending artery (Fig. 2A). Given that the tumour was growing on the surface of the right ventricular apex and not protruding into the cavity of the right ventricle, the border between the tumour and the superficial fatty layer of the myocardium was sharply dissected after local fixation of the tumour, with preservation of the left anterior descending artery. The tumour was carefully and completely resected with off-pump surgery without damaging the left anterior descending artery (Fig. 2B).
Histopathological examination showed that the mass was a fibroma, which presented as a tumour-like hyperplasia of the fibrous tissue with abundant collagen fibres. There was no evidence of mitotic figures or malignancy (Fig. 3).
The patient recovered well after surgery and was discharged on postoperative day eight. Echocardiography performed at three-month intervals postoperatively did not reveal any evidence of recurrence and the patient remains asymptomatic.

Discussion

Cardiac fibromas are uncommon, benign, solitary tumours that occur in children and adults. These tumours account for 12–16% of primary cardiac tumours in children but for less than 5% of all cardiac tumours in adults. The majority of cardiac fibromas are found in the left (60%) or right ventricle (30%), with a minority in the right atrium (10%).

In the gross specimen, a cardiac fibroma is firm, well restricted but not encapsulated, grey-white in colour, and typically does not grow to a large size. Calcification of the central portion is an important characteristic of a cardiac fibroma that distinguishes this tumour from a rhabdomyoma and indicates a poor blood supply. The myocardium can interdigitate with a cardiac fibroma at the tumour border and replace functioning muscle, leading to intractable congestive heart failure.

Histopathological examination of a cardiac fibroma reveals fibroblasts embedded in collagen and varying numbers of elastic fibres. Sometimes, necrosis and cystic degeneration may be observed. In our case, the cardiac fibroma originated from the myocardium of the right ventricle and received its blood supply via the distal left anterior descending artery. The tumour was calcified and firm with a smooth, yellowish surface.

Echocardiography is the preferred imaging method in view of its ready accessibility and non-invasive nature. Furthermore, computed tomography and magnetic resonance imaging (MRI) can localise the tumour and characterise the tissue and surrounding structure. MRI can also provide additional functional data, reveal the underlying pathological mechanisms, and evaluate the haemodynamic effects of this tumour.

The clinical manifestations of cardiac fibroma vary in severity according to the position and size of the tumour. Although many patients with this condition are asymptomatic, about 50% present with symptoms, including inflow and outflow obstruction, angina, arrhythmias and conduction disorders. Heart failure is the most common clinical manifestation, followed by chest pain, syncope and sudden death. Our patient presented with chest tightness and nausea because the calcified mass was compressing the myocardium and causing cardiac ischaemia.

Cardiac fibroma rarely resolves spontaneously and surgical resection should be considered in symptomatic patients. Asymptomatic individuals require long-term follow up and may need surgery to avoid complications. Complete removal is the first goal of surgery for a cardiac fibroma. However, although this tumour was benign and had an easily distinguishable excision plane, partial excision might have been preferable if complete resection was not possible, because of a high risk of damage to essential cardiac structures, despite the higher risk of recurrence. The option of heart transplantation remains available if irreparable cardiac damage is suspected during tumour resection.

Cardiopulmonary bypass is usually required intra-operatively to remove a cardiac fibroma without causing unnecessary injury to surrounding structures when the tumour is located within a ventricle. In our patient, who was symptomatic, cardiopulmonary bypass was not required for complete excision of the tumour because it was located on the surface of the right ventricular wall and there was a clear border between the tumour and the myocardial tissue, and the right ventricular wall was preserved intact.

Conclusions

This report describes an unusual calcified fibroma in the right ventricle in an adult. Surgical excision is an effective way of improving the prognosis in a symptomatic patient.

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