Cardiology in the Young

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Surgical treatment of left ventricular fibroma in adult: case report and literature review

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Original Article

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Abstract

Cardiac fibroma is a rare benign primary tumour of the heart. In the paediatric population, it has been reported as the second most common benign cardiac tumour following rhabdomyoma. However, the prevalence of cardiac fibroma is rarely reported in the adult population. Signs and symptoms are nonspecific, including palpitations, cardiac murmur, arrhythmias, dyspnoea, cyanosis, chest pain, and sudden mortality, whereas, a number of patients with cardiac fibroma are asymptomatic. Surgical resection should be considered as the best option in symptomatic patients. This study reported four surgical cases of adults with cardiac fibroma arising from the left ventricle and a literature review regarding the clinical and pathological features, diagnostic modalities, therapeutic aspects, and prognosis of this rare entity.

Cardiac fibroma is a rare benign primary tumour of the heart. In the paediatric population, it has been reported as the second most common benign cardiac tumour following rhabdomyoma,[1] even though which may not present clinically until adulthood. However, the prevalence of cardiac fibroma is rarely reported in the adult population. Signs and symptoms are nonspecific, including palpitations, cardiac murmur, arrhythmias, dyspnoea, cyanosis, chest pain, and sudden mortality,[2] whereas a number of patients with cardiac fibroma are asymptomatic.[3] Surgical resection should be considered as the best option in symptomatic patients. This study reported four surgical cases of adults with cardiac fibroma arising from the left ventricle and a literature review regarding the clinical and pathological features, diagnostic modalities, therapeutic aspects, and prognosis of this rare entity.

Cases report

Between July 2016 and May 2019, four adults (four females) diagnosed with primary intramural tumours arising from the left ventricle underwent a complete surgical resection. Their ages ranged from 15 to 56 years, height ranged from 152 to 178 cm, and weight ranged from 59 to 77 kg. Echocardiography and cardiac CT or/and MRI confirmed the diagnosis of the cardiac mass located at the wall of left ventricle in all the patients (Fig 1). Basal characteristics of the patients are shown in Table 1.

Surgery was performed through a median sternotomy with cardiopulmonary bypass and cold cardioplegic arrest. The pericardium was opened, and a tumour mass was evident arising from the wall of left ventricle. Dissection was started from the surface of left ventricle. The tumour mass was dissected free of the surrounding myocardium using sharp dissection until to the normal myocardium. Then, the tumour mass was completely removed with or without entering the ventricular cavity, leaving a defect in the wall of left ventricle. For the endocardium remained intact, the defect of left ventricle wall was closed with a running 2/0 Prolene suture. For the defective endocardium, it was reinforced with a polytetrafluoroethylene or pericardium patch firstly and then closed with a running 3/0 Prolene suture. The surgical information of the patients is shown in Table 2.

There are no hospital and late deaths and no major complications. Post-operative electrocardiogram of all the patients showed sinus rhythm and echocardiography showed normal left ventricular ejection fraction with no residual mass. Histopathological examination conducted by the Pathology Department of Fuwai hospital confirmed the tumour mass to be the fibroma. All patients remained symptom-free without episodes of arrhythmias during the follow-up period. The echocardiography revealed normal left ventricular size and left ventricular ejection fraction of each patient without tumour recurrence.

Discussion

Primary cardiac tumours are extremely rare, with a reported incidence ranging from 0.0017 to 0.019%.[4] Cardiac fibroma is the second most common primary cardiac tumour in infants and young children after rhabdomyoma. Although cardiac fibroma is a benign and solitary tumour

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Table 1. Clinical characteristics of the four patients in this study

Patient number	Age (years)	Gender	Height (cm)	Weight (kg)	Symptom	Cardiothoracic ratio in X-ray	Electrocardiogram	Pre-operative workup
1	15	Female	171	77	Arrhythmias	0.5	Ventricular premature beat	Echo (LVEF 64%, LVEDD 45 mm), MRI
2	56	Female	152	61	Chest pain and dyspnoea	0.48	SR	Echo (LVEF 65%, LVEDD 45 mm), CT
3	29	Female	158	59	Asymptomatic	0.44	SR	Echo (LVEF 65%, LVEDD 43 mm), CT, MRI
4	17	Female	178	70	Dyspnoea	0.51	SR, ST segment change	Echo (LVEF 70%, LVEDD 52 mm), CT, MRI

Echo = Echocardiogram; LVEDD = left ventricular end-diastolic diameter; LVEF = Left ventricular ejection fraction; SR = sinus rhythm.

Table 2. The surgical information of the patients

Patient number	Tumour location	Surgery	Tumour sizes (mm)	Histology	Post-operative hospital stay	Follow-up duration (months)	Current status
1	Apex and posterolateral wall of LV	Complete tumour resection	50 × 35 × 18	Fibroma	8	16	Well, SR, ST segment change, no recurrence (LVEF 64%, LVEDD 43 mm)
2	Apex and anterior wall of LV	Complete tumour resection	50 × 35 × 18	Fibroma	7	5.5	Well, SR, no recurrence (LVEF 60%, LVEDD 39 mm)
3	Posterolateral wall of LV	Complete tumour resection	65 × 35 × 30	Fibroma	10	12	Well, SR, no recurrence (LVEF 63%, LVEDD 40 mm)
4	Apex of LV	Complete tumour resection	60 × 40 × 40	Fibroma	7	19	Well, SR, no recurrence (LVEF 60%, LVEDD 49 mm)

LV = left ventricle; LVEDD = left ventricular end-diastolic diameter; LVEF = left ventricular ejection fraction; SR = sinus rhythm.

composed of fibroblasts and collagen, it is clinically important as it may present with symptoms such as inflow and outflow obstruction, conduction system disease, and sudden death. In contrast to cardiac rhabdomyoma, cardiac fibroma rarely regresses spontaneously and surgical removal in a symptomatic case should be considered.

Although the incidence is low, the patients should be found as early as possible, even though they are asymptomatic. Echocardiography is non-invasive, fast, and does not involve the use of radiation. It is generally the initial diagnosis modality for evaluating cardiac fibroma. Supplementary diagnostic techniques include CT or MRI. CT and MRI can provide the location of the tumour, as well as identifying its surrounding structures and haemodynamic effects. In addition, MRI can provide additional functional data. Therefore, cardiac MRI is the modality of choice for further evaluation of cardiac fibroma.[1,3]

Torimitsu et al[5] found that first, the high ratio of tumour-to-heart size may generate low cardiac output and therefore lead to a poorer prognosis. Second, the ratio of the sites of cardiac fibroma occurrence corresponds with the ratio of the muscular weight of the cardiac chamber. Third, septal involvement is a significant poor prognostic factor for cardiac fibroma regardless of tumour diameter relative to heart size, since cardiac fibroma involving the interventricular septum more frequently induces conduction system disease.

Surgery is required as an effective management strategy in the adults when cardiac fibroma cause ventricular inflow/outflow tract obstruction, valve dysfunction, heart failure, and arrhythmia. The tumour size in relationship to the left ventricular cavity has often been considered a limiting factor for complete excision of fibromas, because of leaving a large defect in the wall of left ventricle. In addition, it was difficult to see the demarcation zone between the tumour and the normal tissue in the presence of cold cardioplegia and remove the tumour mass completely without damage to others, such as the chordae, coronary vessels. Therefore, an experienced surgeon can complete the operation better through the trans-oesophageal echocardiography.

In the present study, one patient presented with frequent ventricular premature beats. The onset of ventricular premature beat for displacement or compression of the conduction system is often life-threatening and difficult to control, which can result in sudden death. The ventricular premature beat was eliminated after the patient underwent complete fibroma excision, suggesting that significant debulking of the tumour mass might return to normal motion of left ventricle, which leaded to sinus rhythm.

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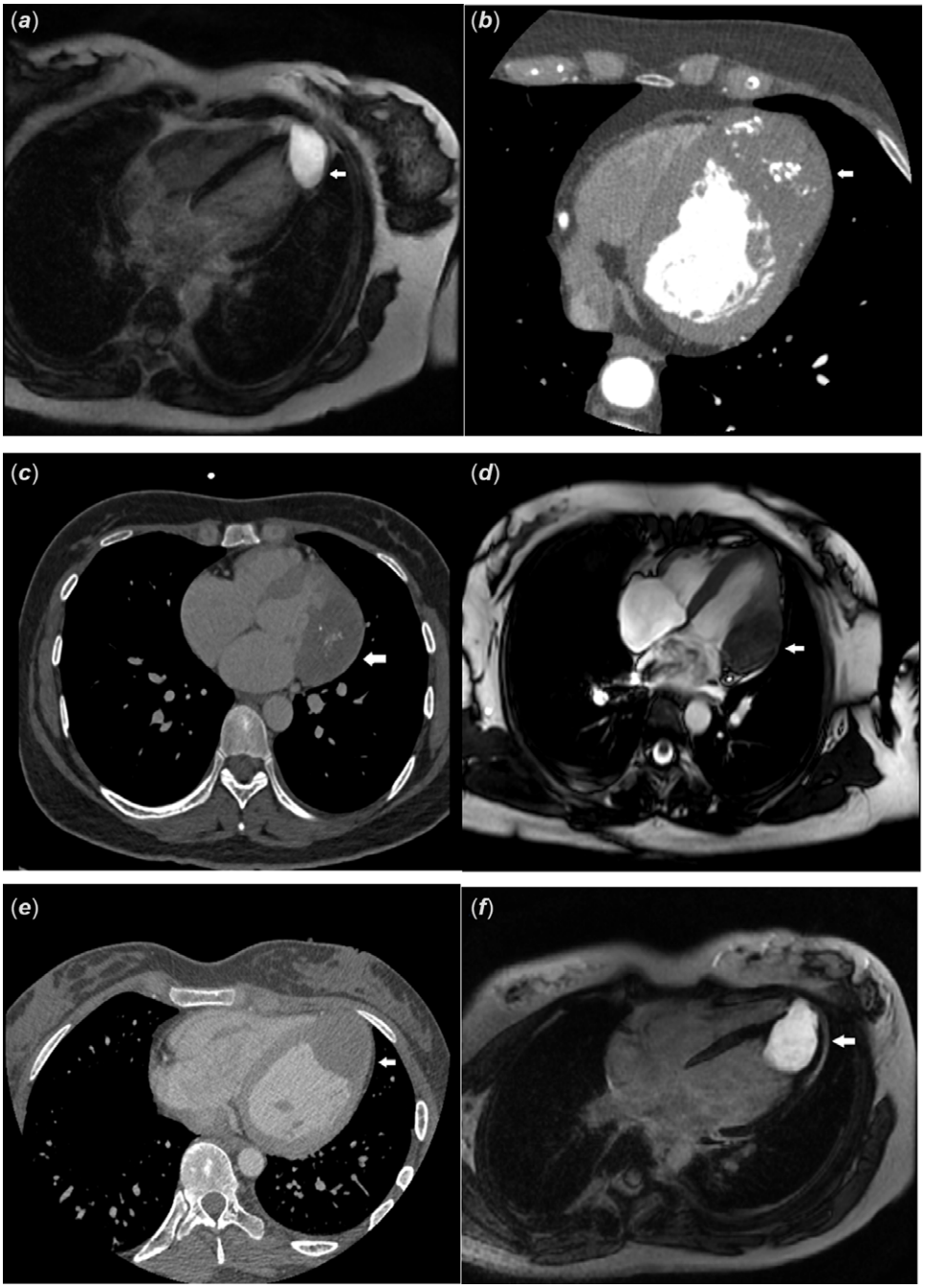

Figure 1. (***a***) MRI of patient 1. (***b***) CT of patient 2. (***c***) CT of patient 3. (***d***) MRI of patient 3. (***e***) CT of patient 4. (***f***) MRI of patient 4.

In this study, we found that the tumour mass grew far away from the left ventricular cavity, and there is no significant change in the diameter of the left ventricular end-diastolic tract before and after surgery. Those symptoms may be caused by a huge tumour mass occupying the left ventricular cavity which is limited. However, this hypothesis needs further study.

Conclusion

Cardiac fibroma is very rare in adults. Echocardiogram, CT, and MRI can provide valuable findings. Surgical excision is a reliable and effective method for treatment. The cardiac function at short-term to medium-term follow-up is excellent, with a resolution of symptoms and arrhythmic vents.

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kind in any product, service and/or company that could be construed as influencing the position presented in, or the review of, the manuscript entitled.

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