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CASE REPORT

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Left ventricular fibromas in pediatric patients: a case series and review of the literature

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Abstract

Cardiac fibromas are rare benign primary tumors of the heart; nearly one-third of affected patients are less than 1 year old, and only 15% of patients develop this disease in adulthood. Here, we report three cases of cardiac fibroma in our hospital. In all the patients, the cardiac mass was diagnosed using cardiac imaging, namely CT and MRI scans. The tumors were surgically resected under cardiopulmonary bypass, and the fibroma was diagnosed by histopathology and microscopy. The cardiac function of the three pediatric patients recovered after the operation, and the patients were discharged successfully.

Keywords Pediatric, Cardiac surgery, Cardiac fibroma, Cardiopulmonary bypass

Introduction

Cardiac fibromas are rare benign primary cardiac tumors and account for 12–16% of all pediatric primary heart tumors, making them second only to rhabdomyomas [1]. The average age at onset is 13-years; nearly one-third of patients are less than 1-year old, and only 15% of cases develop in adulthood [2]. Cardiac fibromas typically present as isolated masses with an average diameter of 5 cm. Additionally, these fibromas lack a capsule and are capable of infiltrating normal myocardium. Fibromas predominantly affect the left ventricular (LV) free wall (58% of cases), followed by the right ventricle (28%), the interventricular septum (17%), and the atrium [3]. Symptoms primarily include arrhythmias, dyspnea, chest pain, and sudden death, but approximately one-third of patients have no significant symptoms. Echocardiography usually serves as the initial diagnostic tool, followed by magnetic resonance imaging (MRI), which can delineate the extent of myocardial infiltration by the fibroma. In addition, computed tomography (CT) can reveal the mass and may reveal central calcification within the fibroma. When the tumor invades the interventricular septum and conduction system, there is a risk of fatal arrhythmias. Surgical resection is an effective method for treating cardiac fibroma.

Recently, three pediatric patients were treated for cardiac fibroma in our institution. The specific conditions of the three cases are summarized in Table 1.

Case 1

A female pediatric patient aged 1 year and 9 months presented to the clinic weighing 11.5 kg. During the hospital admission examination, there was no syncope or hemoptysis. The preoperative electrocardiogram revealed sinus rhythm with a rightward electrical axis. Preoperative echocardiography revealed an enlarged left ventricle

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Table 1 The essential information of the three cases

	Case1	Case2	Case3
Year	1 9/12 year	4 month	8 month
Weight (kg)	11.5	5.5	9.5
Gender	Male	Male	Male
Diagnosis	Ventricular mass	Ventricular mass	Ventricular mass
Pre-op LVEF	70%	32.4%	71%
Pre-op ECG	Sinus rhythm, right-axis deviation	Sinus rhythm, right-axis deviation ST deviation	Sinus rhythm ST deviation
Tumor size(mm)	34×35	52×54	42×25
pathology results	Fibroma	Fibroma	Fibroma
Cardioplegic	HTK	HTK	HTK
CPB time (min)	108	144	143
ACC time(min)	79	119	116
Post-op LVEF	55%	65%	40%
Discharge LVEF	61%	60%	62%

and a tumor-like protrusion of the entire left ventricular lateral wall apex, with a large intramyocardial space-occupying lesion measuring approximately 41×26 mm, a relatively uniform echo pattern, and an unclear boundary with the myocardial tissue (Fig. 1a). No significant blood flow signal was detected within the space-occupying mass, which had a regular shape. Contractile motion was observed in the outer layer of the ventricular wall, whereas the thickness of the remaining left ventricular wall was normal. A fibroma was considered likely. Cardiac-enhanced MRI revealed a space-occupying lesion at the apex of the left ventricular lateral wall and that the lesion did not exhibit a significant change in shape with contraction. The lesion measured approximately 34×35 mm and had a blurry boundary with the myocardium, showing iso-intense T1 and slightly high T2 signals (Fig. 1b). These findings indicate a high likelihood of a benign fibroma. Left ventricular function (LVEF) was as follows: LVEF 57%, CO 2.2 L/min, EDV 41 ml, and EDVi 76 ml/m^2.

After radiological confirmation, a decision was made to proceed with cardiac tumor resection under cardiopulmonary bypass (CPB). After establishing extracorporeal circulation through the ascending aorta and superior and inferior vena cava, the entire left ventricular lateral wall apex, resembling a protruding tumor, could be seen with a large intramyocardial space-occupying lesion measuring approximately 50×30×30 mm with unclear boundaries with heart muscle tissue (Fig. 1c). After aortic clamping, 600 ml of Custodiol HTK-Solution solution were perfused. An incision was made at the apex of the left ventricle, and the tumor, located in the myocardial layer of the left ventricle, was completely peeled off the left ventricular wall. A bovine pericardial patch was sutured with 5–0 Prolene sutures to reinforce the

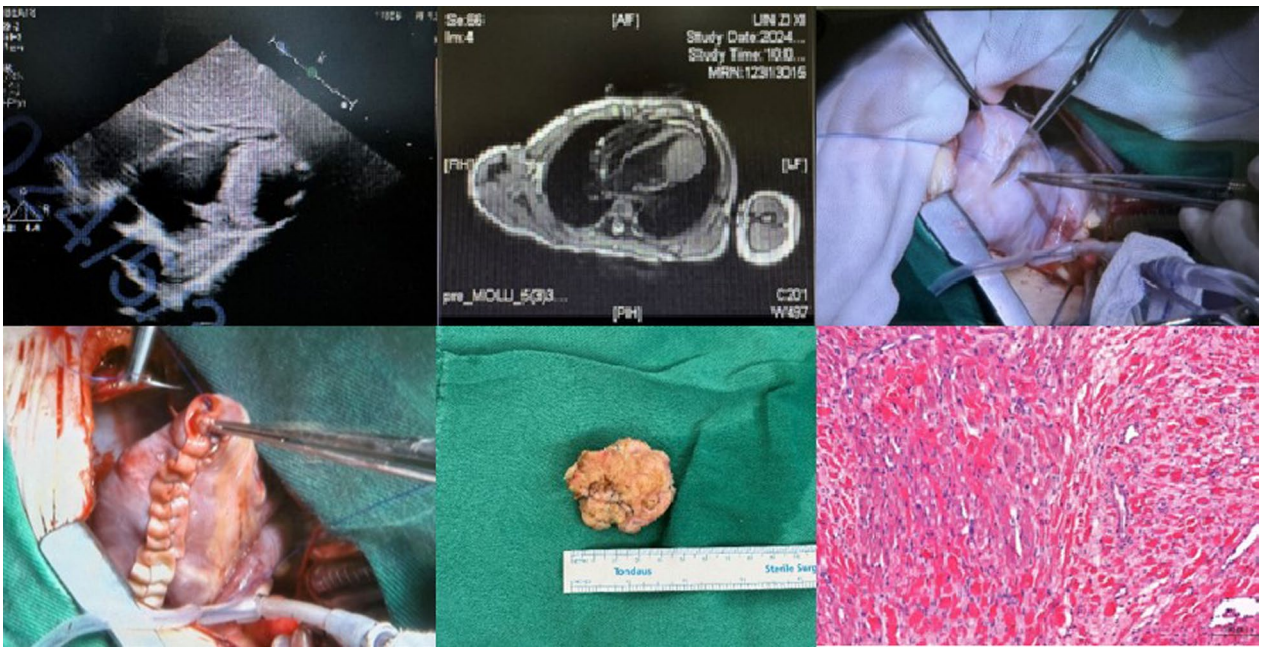

Fig. 1 **a** Transthoracic echocardiography revealed the mass LV. **b** CMR LGE imaging showed tumor morphology. **c** The tumor is seen from the surface of the ventricle. **d** Suture closure of the left ventricular incision. **e** Measured the size of the tumor. **f** Hematoxylin–eosin staining of tumor tissue

left ventricular sidewall endocardium, and 4–0 Prolene sutures were continuously threaded through bovine pericardial patch to close the left ventricular apex incision (Fig. 1d). The removed fibroma was sent for pathological examination, and the tumor body is shown in Fig. 1e. After release of aortic cross-clamp in 79 min, the heart resumed beating spontaneously. The hemodynamics were stable, and the patient was successfully weaned from CPB after 16 min. Intraoperative transesophageal echocardiography revealed a slightly small left ventricular inner diameter, normal thickness of the interventricular septum and left and right ventricular walls, slightly low movement amplitude of the lateral wall near the apex of the heart, and minor regurgitation of the tricuspid and mitral valves. Pathological examination revealed that the tumor was composed of spindle cells with few cells, no mitotic figures, and interstitial collagen fibers or myxoid material, and the tumor partially invaded the myocardium (Fig. 1f). The patient was transferred back to the pediatric intensive care unit (PICU), weaned from the ventilator within 4 h, discharged from the PICU within 40 h, and successfully discharged from the hospital 5 days after surgery. The predischarge electrocardiogram (ECG) revealed sinus rhythm, abnormal Q waves, and ST-T changes. Predischarge echocardiography revealed an LVEF of 61%, minor mitral regurgitation, and trace tricuspid regurgitation.

Case 2

A four-month-old male infant weighing 5.5 kg presented to the clinic. Echocardiography during a postnatal examination revealed a solid, echogenic nodule in the left ventricular wall, suggesting a rhabdomyoma, with a small amount of pericardial effusion. Two months prior to admission, cardiac MRI suggested "space-occupying lesions within the myocardium of the left ventricular posterior wall, lateral wall, anterior wall, and interventricular septum, considering fibroma". After admission, the child has no symptoms of dyspnea or respiratory distress and can lie flat. Echocardiography suggested an enlarged left ventricle with a generally reduced contraction amplitude. A large mass filling the entire ventricular cavity (high likelihood of fibroma) adhered to the interventricular septum, with a mass diameter of approximately 52×54 mm, echo intensity, and uniform texture. Only the posterior apex of the left ventricle was involved in ventricular cavity function (Fig. 2a). There was functional mitral regurgitation, moderate to severe regurgitation, and an LVEF of 32.4%. Preoperative CT revealed a large wall-thickening lesion in the anterior wall adjacent to the anterior interventricular septum and anterolateral wall, with a lower density than the myocardium in the arterial and delayed phases, adjoining the aortic sinus portion (left coronary sinus) and the apical part of the left ventricle

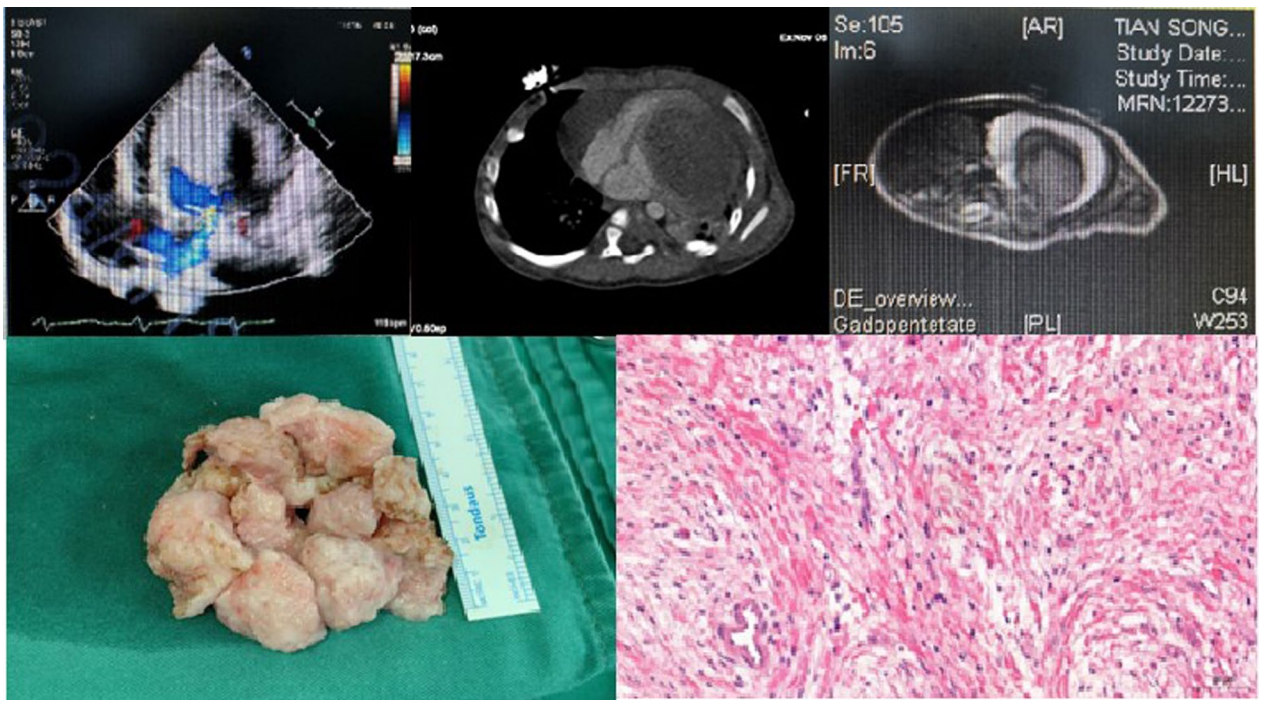

Fig. 2 **a** Transthoracic echocardiography revealed the mass LV. **b** Tumor morphology under CT. **c** CMR LGE imaging showed tumor morphology. **d** Measured the size of the tumor. **e** Hematoxylin–eosin staining of tumor tissue

(Fig. 2b). This lesion protruded into the left ventricular cavity, causing chamber shrinkage, and had a clear boundary as well as a small right ventricular chamber. Surgical resection of the cardiac tumor was planned. Preoperative MRI revealed a space-occupying lesion measuring approximately 57 × 40 × 43 mm in the anterior wall of the left ventricle adjacent to the anterior septum and the anterior lateral wall (Fig. 2c). The proximal end of the space-occupying lesion was close to the aortic sinus, and the distal end was close to the apex of the left ventricle. The lesion showed expansive growth, and the convex left ventricle significantly decreased the diameters of the upper and lower sections of the heart cavity. Early filling of the space-occupying lesion was not observed during overperfusion. Enhanced scanning revealed that the left ventricular space was progressively enhanced from the edge of the lesion, and multiple unenhanced areas were observed in the center. Preoperative electrocardiogram revealed sinus rhythm, a rightward electrical axis, and ST-segment changes. Extracorporeal circulation was established through the superior and inferior vena cava to the aorta, and 250 ml of Custodiol HTK-Solution solution was infused after aortic clamping, achieving satisfactory cardiac arrest. The tumor tissue was incised from the left ventricular wall; the tumor was tough, nonencapsulated, and infiltrated the myocardium. The tumor was excised, measuring approximately 70 × 70 × 20 mm, and then sent for pathological examination (Fig. 2d). Bovine pericardial patch was used, the cavity was closed with 5/0 Prolene sutures, and the left ventricular incision was sutured with 6/0 Prolene sutures. Left ventricular venting was performed, and after release of aortic cross-clamp in 119 min, the heart resumed spontaneous beating in sinus rhythm, and the patient was successfully weaned from CPB after 18 min of assistance. Intraoperative echocardiography revealed that after excision of the tumor from the anterior wall and interventricular septum, the local ventricular wall was thinner, with a reduced motion amplitude. After the ventricular surface was repaired, a liquid dark area of approximately 30 × 12 mm was visible, which was filled with tiny communication holes approximately 3 mm apart from the left and right ventricular chambers. The remaining wall thickness was normal, the contraction amplitude was satisfactory, and the LVEF was 55%. Pathological examination revealed that the tumor cells were long and spindle shaped, with slightly larger nuclei, oval shapes, and no mitotic figures. More mucoid substances exist between cells (Fig. 2e). There were more myocardial cells interspersed among the tumor cells. The immunohistochemistry results were as follows: Calretinin (−), Desmin (T), and Ki67 (1%). The child was weaned from the ventilator 69 h after surgery and discharged from the PICU within 7 days.

Three weeks after surgery, the patient experienced shortness of breath and pericardial effusion was suspected. The emergency ultrasound examination revealed pericardial effusion. So sternotomy was performed for hemostasis. But 36 h after sternotomy, ultrasound examination revealed pericardial effusion again. Then left ventricular angiography revealed communication between the residual cystic cavity and both ventricles as well as leakage into the pericardial cavity, and ventricular cyst repair surgery was performed under CPB. The aortic root was perfused with Custodiol HTK-Solution solution, the interventricular septal cyst wall was opened, and the interventricular septal defects were closed with a bovine pericardial patch. The child was discharged from the PICU 6 days after the operation and was discharged home 15 days after the operation. Three months after surgery, ECG revealed sinus rhythm, an LVEF of 75%, increased echogenicity and uneven echogenicity of the myocardium of the interventricular septum and anterior wall of the left ventricle. The communication between the original left ventricular wall cystic cavity and the pericardial cavity ceased, and no free-liquid dark areas were observed in the pericardial cavity.

Case 3

An eight-month-old male infant weighing 9.5 kg presented to the clinic. A heart murmur was identified during physical examination after birth, and a cardiac mass, which was detected at a local hospital, was not treated with systemic medications. The infant was brought to our institution for further treatment. Upon admission, echocardiography revealed a full left atrium, a small left ventricular chamber, and a large space-occupying lesion extending from the basal segment to the apical segment of the interventricular septal wall (Fig. 3a). The largest dimension of the lesion was 51 mm, and its thickest part, approximately 32 mm, was located at the midsection of the interventricular septum. The boundary with the interventricular septal myocardial tissue was unclear, and the mass protruded into the left ventricular outflow tract, causing mild narrowing. Preoperative CT revealed a soft-tissue-density mass in the muscle wall of the interventricular septum with an irregular shape (Fig. 3b). The largest cross-section was approximately 42 × 25 mm, with mild uneven enhancement and delayed enhancement. The CT values in the plain, enhanced, and delayed phases were approximately 42 HU, 48 HU, and 46 HU, respectively. The boundary with surrounding tissues was unclear, and the mass exhibited expansive growth, protruding into both the left and right ventricular cavities. The mass involved the apical and inferior myocardium

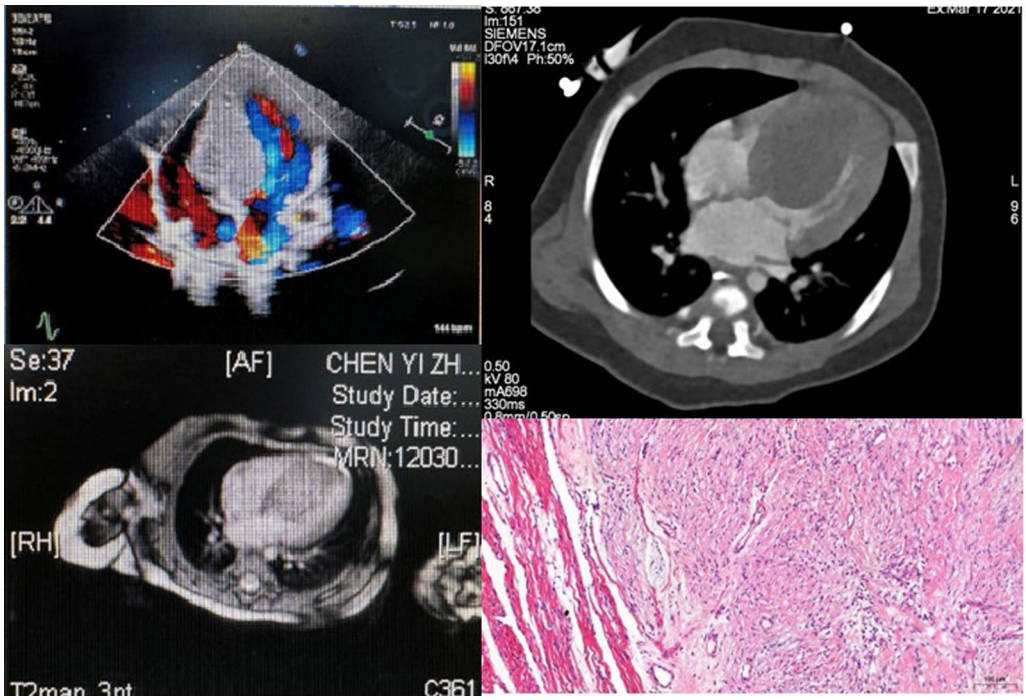

Fig. 3 **a** Transthoracic echocardiography revealed the mass LV. **b** Tumor morphology under CT. **c** CMR LGE imaging showed tumor morphology. **d** Hematoxylin–eosin staining of tumor tissue

of the left ventricle, leading to a reduction in the size of the left ventricular chamber, compression and shrinkage of the right ventricle, and narrowing of the outflow tracts of both ventricles. The left ventricular papillary muscles and tendinous cords were not clearly visible. Preoperative MRI revealed a large space-occupying lesion in the interventricular septum, extending from the proximal end of the septum to the apex, measuring approximately 50 × 26 × 39 mm (left-to-right anterior-to-posterior superior-to-inferior), with the thickest part of the midsection of the septum measuring approximately 30 mm (Fig. 3c). The mass protruded into both the left and right ventricles and displayed slightly long T1 and T2 signals.

Both ventricular chambers were compressed, leading to mild narrowing of the left ventricular outflow tract. Left ventricular function assessment revealed an LVEF of 53.7%, CO of 1.19 L/min, EDV of 18.6 ml, and EDVi of 22.2 ml/m^2. Myocardial first-pass perfusion indicated no significant enhancement in the early phase, with no significant reduction in perfusion or filling defects in the remaining myocardium.

Delayed enhancement scanning: the periphery of the mass showed clear enhancement, whereas the center was less enhanced, showing a progressive increase in enhancement over time. No significant abnormal enhancement was detected in the myocardium. Under CPB, tumor resection surgery was performed, CPB was established between the superior and inferior vena cava and the ascending aorta, and 400 ml of Custodiol HTK-Solution solution was perfused. An incision was made parallel to the left anterior descending artery at the apex of the heart, revealing a large tumor mass occupying the interventricular septum, approximately 8 cm in diameter. The mass was peeled off from the interventricular septum in blocks in 9 pieces, leaving the septum thin after tumor removal. The interventricular septal incision was closed after the septum was reinforced with a piece of pericardium. After the ascending aorta was opened, the heart resumed spontaneous beating, and intracardiac function was good, as assessed by transesophageal echocardiography. Support was successfully withdrawn after 18 min, with a CPB time of 143 min and an ACC of 116 min. Pathological examination revealed gray–white tissue measuring 65.6 × 25 mm and weighing 29.1 g, with the cut surface appearing gray–white and tough. Microscopically, the tumor was composed mostly of spindle cells and collagen fibers, with some spiral patterns of spindle cells, a matrix showing increased mucoid substances, some areas showing interspersed growth of myocardial cells, and no significant atypia.

The pathological diagnosis was fibroma, and the immunohistochemical results were as follows: desmin (−), Ki67 (2%), myogenin (−), myoglobin (sparse +), SMA (−), vimentin (+), CD99 (+), and CD34 (−) (Fig. 3d). The child

was extubated 45 h after surgery, transferred to the PICU after 6 days, and discharged 16 days after surgery. Predischarge echocardiography revealed that the mass in the mid-upper segment of the interventricular septum had disappeared, revealing an irregular anechoic area measuring 40×9 mm, with a breach on the left ventricular side of approximately 2.7 mm. The right ventricular side echo continuity was intact, the interventricular septum contraction amplitude was reduced, the thickness and contraction amplitude of the remaining left ventricular wall was normal, and the outflow tracts of both ventricles were unobstructed. There was a slight reduction in left ventricular systolic function. The LVEF was 50%, and the predischarge electrocardiogram revealed sinus rhythm. One and a half years after surgery, follow-up echocardiography revealed that the intraseptal tumor had disappeared, forming a part of an anechoic cystic cavity, with unobstructed outflow tracts from both ventricles and an LVEF of 67%. An electrocardiogram revealed sinus arrhythmia with abnormal Q waves.

Discussion

Cardiac fibroma is a rare benign cardiac tumor that primarily occurs in infants and children. It is the second most common type of cardiac tumor in children, following rhabdomyoma [3]. This tumor is usually solitary, firm in texture, and has clear boundaries but lacks a true capsule. It is mainly composed of fibroblasts and collagen fibers, with calcification often present in the central part. Since 1976, only approximately 100 cases have been reported [4]. Nearly one-third of young patients have asymptomatic cardiac fibromas, which are often diagnosed later in life [5]. All recent cases in our center were pathologically confirmed fibromas.

Infant cardiac fibromas do not regress or disappear on their own, and they can easily invade the interventricular septum and conduction system, leading to intractable arrhythmias or even sudden death. Christina et al. reported that cardiac fibromas can invade the ventricular conduction system and lead to ventricular arrhythmias at a rate of up to 64% [6]. A multicenter retrospective study conducted in Shanghai Jiao Tong University Affiliated Children's Hospital revealed that among 166 cases of pediatric cardiac tumors diagnosed between 1998 and 2014, only 21 cases of fibroma were reported [7]. Among the 21 patients with cardiac fibromas, four had preoperative arrhythmias, including premature ventricular contractions and paroxysmal supraventricular tachycardia. However, none of the patients in our case series presented with arrhythmias before surgery.

Cardiac fibromas typically manifest as a single nonconvex growth toward the heart chamber, usually located at the anterior wall and interventricular septum. Larger fibromas can obstruct intracavities or valve orifices and lead to severe congestive heart failure. In our first case, the fibroma didn't obstruct intracavity. But echocardiography revealed an enlarged left ventricle and a tumor-like protrusion of the entire left ventricular lateral wall apex. After abnormal findings on preoperative echocardiography, MRI is the preferred method for further assessment and confirmation of cardiac fibromas. The MRI features of fibromas include an intramyocardial location, clear boundaries, iso-intense on T1-weighted imaging and low-intensity on T2-weighted imaging, along with delayed gadolinium enhancement [8]. In our three cases, we performed echocardiography and MRI examinations for all the patients.

Given that cardiac fibromas do not spontaneously resolve, surgical resection is advised upon diagnosis for symptomatic patients and those without symptoms but with large tumor volumes [9]. Thorough removal of the tumor can yield excellent therapeutic outcomes. Partial resection may provide symptom relief for inoperable tumors, which generally have a good prognosis [10]. A single-center, retrospective cohort study from Boston children's hospital metained that large ventricular fibromas can be resected safely with appropriate surgical planning and an emphasis on debulking [11]. For young patients with large tumors accompanied by severe heart failure, heart transplantation may be considered [12]. According to the Mayo Clinic, 18 cardiac fibromas were resected, resulting in one death, one subtotal resection, and 16 complete resections. With over 30 years of follow-up, no tumor recurrence or increase in size was observed in cases of subtotal resection. According to the Mayo Clinic, the resection of fibromas is recommended even in asymptomatic patients because of the risk of fatal ventricular arrhythmias [7]. However, the decision to treat and the timing and method of treatment for asymptomatic cardiac fibroma remain unclear [13, 14]. In our center, the preferred approach is surgery once a diagnosis is made. One of the three surgical pediatric patients in our center presented with a large tumor deep in the myocardium, and postoperative complications included a ventricular–pericardial fistula and septal perforation, requiring another operation for repair. The patient was successfully discharged 15 days after surgery, and a checkup at six months revealed normal heart function and sinus rhythm.

As fibromas generally invade the myocardium, resecting them under cardiopulmonary bypass can be difficult, and the myocardial protectant of choice is generally Custodiol HTK-Solution solution, which ensures adequate clamping time in a single perfusion. Furthermore, perfusing an ample volume of the cardioplegia solution, preferably 40–60 ml/kg, is essential. Additionally, since the Custodiol HTK-Solution requires 5–8 min for Na^+ ions to equilibrate, an adequate perfusion time must be ensured. During clamping, ice-cold compresses

are regularly placed in the pericardial cavity to provide myocardial protection. After the ascending aorta clamp is released, heart function is assessed, and adequate support is provided. Under good cardiac function confirmed by transoesophageal echocardiography, the anesthesiologist can determine the optimal time for weaning from bypass. In patients with large tumors that deeply invade the myocardium and significantly impact heart function, if cardiac function does not recover after prolonged support, it might be necessary to consider the transition to extracorporeal membrane oxygenation (ECMO) support as a bridge therapy for endpoint treatment or heart transplantation [4, 15].

Limitation

There are some limitations. First, we have only summarized three recent cases of cardiac fibroma patients, which is not a large enough sample size. Second, since these are recent cases, we have not yet conducted high-frequency follow-ups. We will continue to follow up with the patients in the future.

Conclusions

In conclusion, cardiac fibromas are relatively rare cardiac tumors that require close attention during routine echocardiography and CT examinations. Most centers recommend immediate surgical resection upon diagnosis to prevent further tumor growth, which could impair ventricular function and rhythm Myocardial protection during surgery is highly important; therefore, choosing the appropriate cardioplegia solution and adequate perfusion volume is essential. After cardiac resuscitation, heart function is evaluated and machine support is provided and adjusted slowly, and ECMO assistance can be considered if heart function is poor.

Abbreviations

LV	Left ventricular
MRI	Magnetic resonance imaging
CT	Computed tomography
LVEF	Left ventricular function
CPB	Cardiopulmonary bypass
PICU	Pediatric intensive care unit
ECG	Electrocardiogram
ACC	Aortic clamping
ECMO	Extracorporeal membrane oxygenation

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s13019-025-03382-1.

Additional file 1.

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Author contributions

CZ Conceptualization; Formal analysis; Funding acquisition; Investigation; Resources; Software; Writing original draft. JL Resources; Software. SL Writing Review and Editing. HW Resources; Investigation. YJ Writing Review. JL Writing Review and Editing.

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Availability of data and materials

No datasets were generated or analysed during the current study.

Consent for publication

Not applicable.

Competing interests

The authors declare no competing interests.

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