

Images in Congenital Cardiac Disease

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Multiple papillary fibroelastoma presenting with mitral, tricuspid, and pulmonary valve involvement and surgical treatment: case report

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Abstract

Approximately 90% of primary paediatric cardiac tumours are benign lesions. Depending on their location and size, benign cardiac tumours may cause inflow and outflow obstructions, cyanosis, valvular insufficiencies, myocardial ischaemia, associated dysfunction, systemic embolisation, arrhythmias, and even sudden death. Decision-making and timing for surgery can be challenging in children. Herein, we present an asymptomatic 11-year-old girl with papillary fibroelastoma in the mitral, tricuspid, and pulmonary valves, discussing the decision-making process and successful surgical management.

Introduction

Approximately 90% of primary paediatric cardiac tumours are benign lesions. Depending on their location and size, benign cardiac tumours may cause inflow and outflow obstructions, cyanosis, valvular insufficiencies, myocardial ischaemia, associated dysfunction, systemic embolisation, arrhythmias, and even sudden death. Decision-making and timing for surgery can be challenging in children. Herein, we present an asymptomatic 11-year-old girl with papillary fibroelastoma in the mitral, tricuspid, and pulmonary valves, discussing the decision-making process and successful surgical management.

Case report

An asymptomatic 11-year-old patient, who had previously been investigated for a murmur at the age of 2 months, presented to our unit. Echocardiography revealed multiple mobile masses around 10 mm located in the mitral, tricuspid, and pulmonary valves. There was mild to moderate regurgitation in the mitral and tricuspid valves and trivial regurgitation in the pulmonary valves (Figure 1; Movie 1A, 1B, 1C in the Data Supplement). To better delineate the masses, we performed cardiac MRI and transoesophageal echocardiography. Cardiac MRI showed dysplastic nodular lesions with haemorrhagic content (Figure 2, Movie 3A, 3B, 3C in the Data Supplement), while multinodular lesions were seen in transoesophageal echocardiography (Figure 3, Movie 2A, 2B, 2C in the Data Supplement). Despite the patient being asymptomatic, a decision for surgical removal was made to prevent embolisation and confirm the pathological diagnosis of the masses. During the operation, multinodular lesions were excised from the tricuspid, mitral, and pulmonary valves with great care taken to preserve the leaflets. Since valve insufficiency was not hemodynamically significant, valve repair was not deemed necessary (Figure 4). After the masses were removed, the valves were tested with saline, and mild regurgitation was noted. The patient was weaned off cardiopulmonary bypass easily, and post-cardiopulmonary bypass echocardiography showed good biventricular function with mild to moderate tricuspid, mitral, and trivial pulmonary valve regurgitation, with no masses seen (Figure 5, Movie 5A, 5B). The degree of valve insufficiency did not change before and after the operation. The postoperative course was uneventful, and the patient was discharged on the 7th postoperative day. Histopathology confirmed the diagnosis of papillary fibroelastoma. The patient, who was still asymptomatic at the 8th postoperative month follow-up, stated that her exercise capacity had also increased.

Discussion

Benign cardiac lesions can be diagnosed through a murmur and cyanosis or can be detected incidentally during echocardiographic examination. The main determining factors for the decision to surgically remove lesions are their location and size. Lesions in the outflow tract

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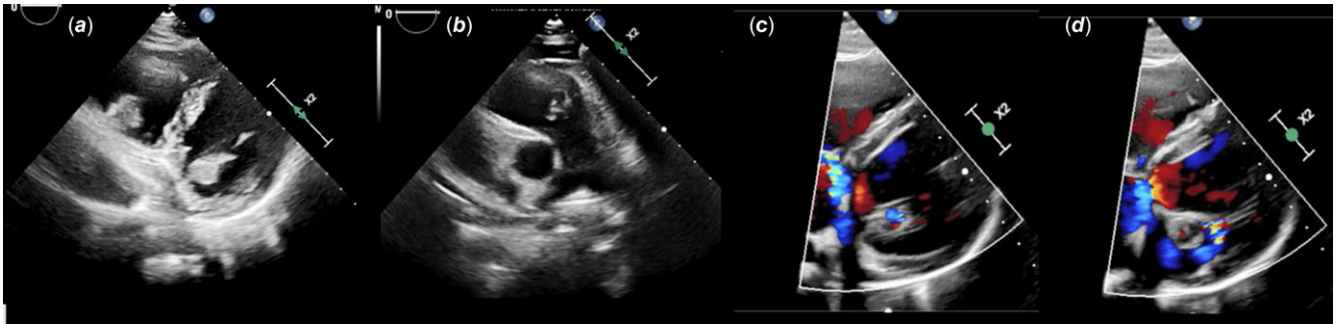


Figure 1. (a) Large lesions in the mitral and tricuspid valves. (b) Medium-sized lesion in the pulmonary valve. (c-d) Mild to moderate mitral regurgitation and tricuspid regurgitation jet (preoperative period images).

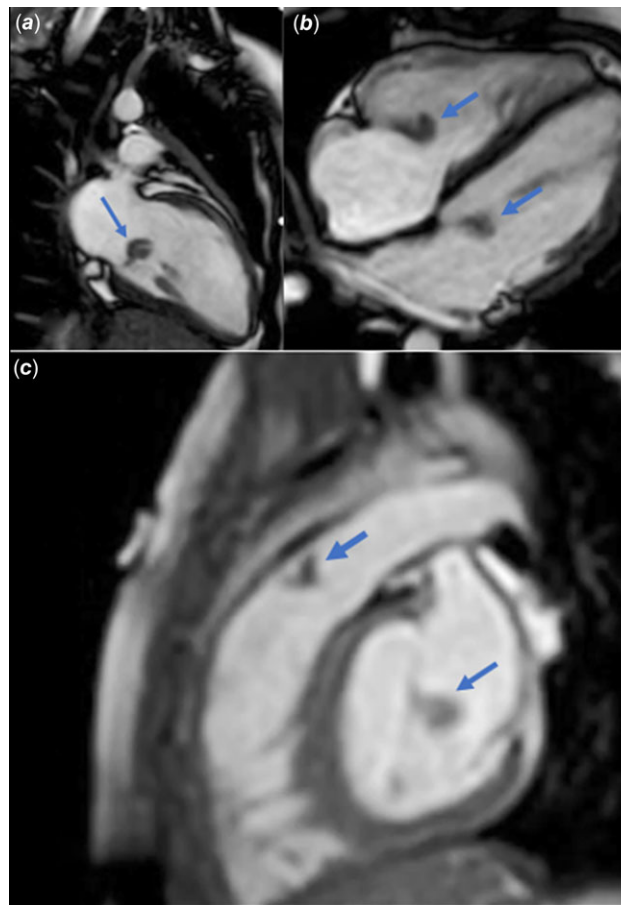


Figure 2. Images of lesions on cardiac MRI. (a) Coronal section modified two-chamber MRI image of the lesion in the mitral valve. (b) Sagittal section four-chamber MRI image of the mitral valve lesion and tricuspid valve lesion. (c) Short-axis MRI image of the lesion in the mitral valve and the lesion of pulmonary valve in the right ventricular outflow tract.

can result in obstruction, embolism, and myocardial ischaemia, while larger lesions in the inflow can disrupt the coaptation of the valves and cause problems related to serious valve insufficiency.¹ Benign cardiac lesions with valvular involvement include papillary fibroelastoma, blood cysts, thrombus, and rarely myxoma. The differential diagnosis of congenital tumours characterised by multiple valve involvement is even more difficult, with only a few case reports available.² Furthermore, noninvasive diagnostic methods may not provide clear diagnostic information in most patients. As our patient did not have any underlying

conditions causing thrombus formation, thrombus was not considered a primary cause. Cardiac MRI did not offer more diagnostic information than echocardiography. Despite the difficulty in deciding whether to perform surgery in an asymptomatic patient with a cardiac mass, we opted for surgical excision due to the large size and multivalvular involvement. Our goals were to determine the tissue diagnosis, reduce the risk of embolisation, and prevent potential valvular problems.

Papillary fibroelastoma constitutes a significant portion of childhood valve lesions. Cases of fibroelastoma characterised by

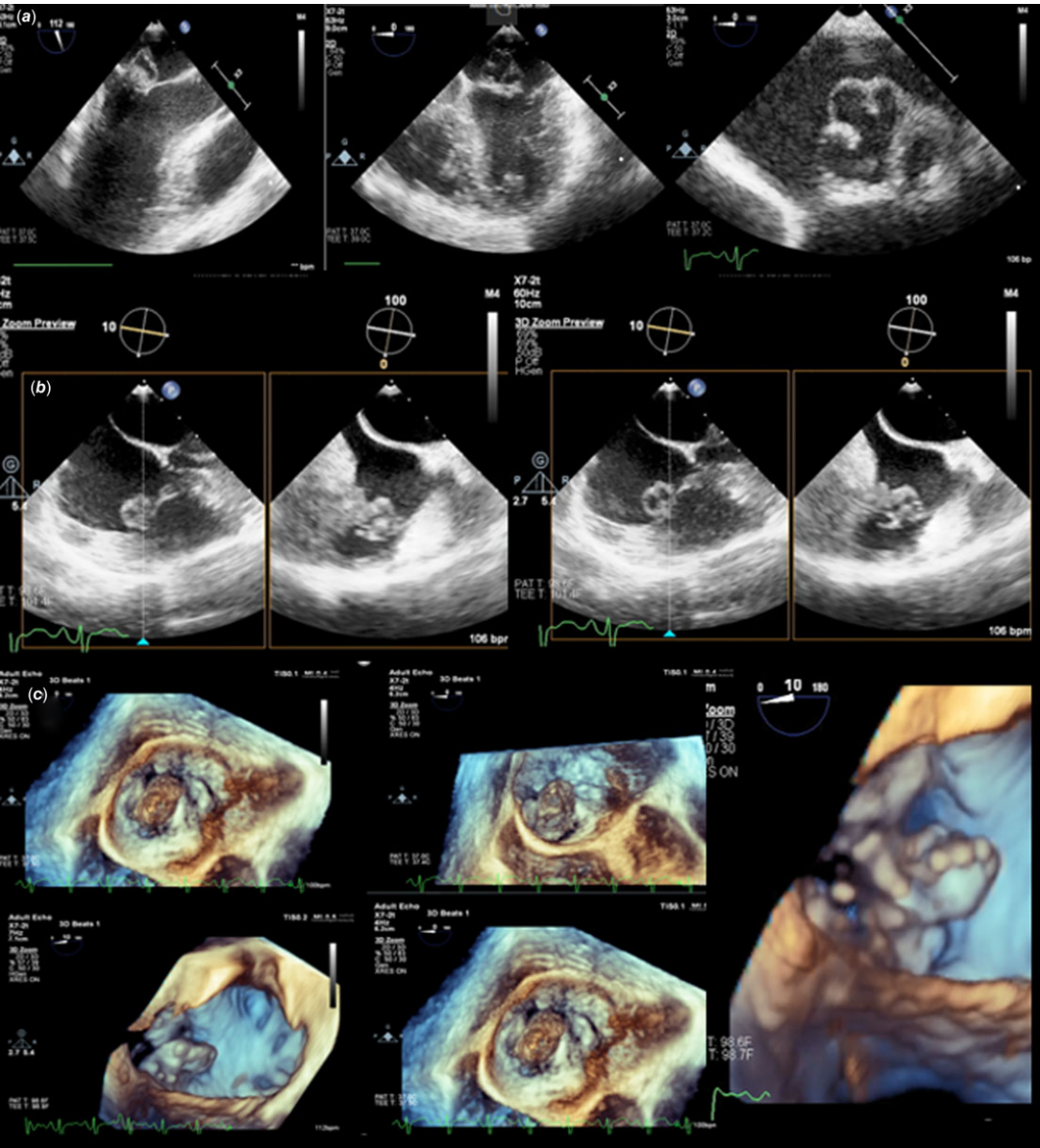


Figure 3. The transesophageal echocardiography images. (a) Cauliflower-like images of the lesion in the mitral valve. (b) Cauliflower-like images of the lesion in the tricuspid valve. (c) 3D image of the lesion in the mitral valve as nodular and raised from the valve.



Figure 4. Multinodular lesions excised from mitral valve (a) tricuspid valve (b) pulmonary valve (c).

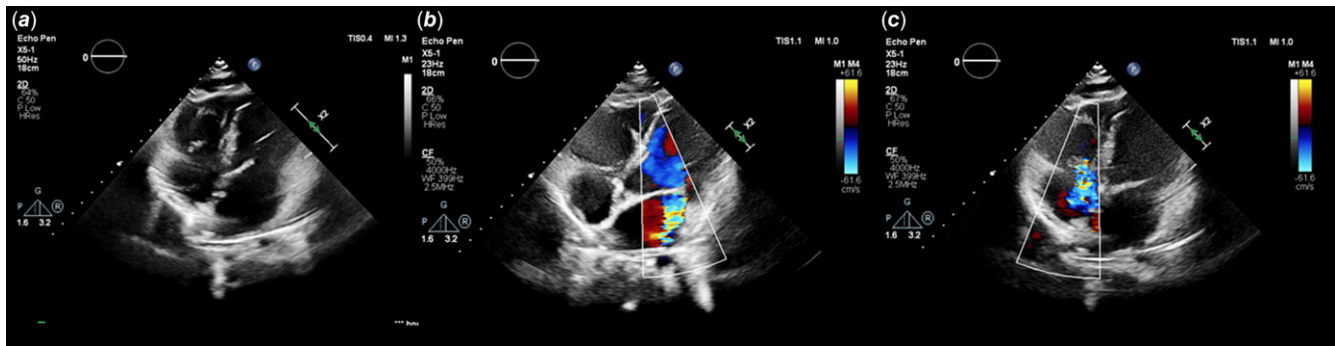


Figure 5. (a) 2D images of the mitral and tricuspid valves after the excising operation of large multiple papillary fibroelastoma lesions. (b) Doppler images of mild to moderate mitral regurgitation jet. (c) Doppler images of mild to moderate tricuspid regurgitation jet.

multiple valve involvement are limited in the literature. In the available literature, 7–10% of patients diagnosed with papillary fibroelastoma present with multiple valve involvement. It is stated that previous surgery and/or the presence of hypertrophic cardiomyopathy may play a role in its pathophysiology, suggesting an aberrant reactive response to trauma.^{3,4} However, papillary fibroelastoma cases characterised by multiple valve involvement can also occur de novo, as seen in our patient.⁵ Surgery has been recommended for lesions such as fibroelastoma, with a suggested risk of life-threatening events if the lesion is greater than 1 cm. Surgery may also be required in blood cysts, depending on the size of the lesion.^{6,7} In asymptomatic patients, it is challenging to decide between surgery and follow-up without surgery, considering the surgical risk. The location and size of the lesion provide predictions for clinical situations that can have serious long-term consequences. However, when making the decision, the ease of surgical technique and possible serious consequences should also be taken into account.⁵ Although the mass contributed to the valve insufficiency in our patient, a decrease in valve regurgitation after excision was expected. However, the degree of valve insufficiency did not change postoperatively. Since the primary goal was to prevent the mass and potential life-threatening complications, the procedure was considered successful. While rare, recurrence of papillary fibroelastoma after surgical excision can occur, particularly in cases of incomplete excision or complications. One study reported a recurrence rate of approximately 1.6%, emphasising the importance of thorough excision to reduce the risk of recurrence.⁸ Therefore, patients should be monitored for recurrence, even though it is rare.

Supplementary material. The supplementary material for this article can be found at <https://doi.org/10.1017/S1047951124036503>.

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