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Brief Report

Giant fibroma of the right ventricle

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Abstract Cardiac fibromas are rare benign tumours of connective tissue that occur most frequently in children within the left ventricle. Spontaneous regression has not been observed, and surgical intervention is usually required. We have successfully treated a 1-year old girl with a giant fibroma of the right ventricle using the principles of the Batista procedure. The diagnosis was primarily made using transthoracic echocardiography.

Keywords: Cardiac tumours; echocardiography; Batista procedure; ventricular reduction

PRIMARY TUMOURS OF THE HEART ARE RARE IN ALL age groups, occurring with a frequency from 0.0017% to 0.027%. The cardiac fibroma is a benign tumour of connective tissue that mainly affects children, and has also been detected in the fetus.[1] Most fibromas occur within the left ventricle. They are symptomless in about one-third of patients, and consequently many are discovered incidentally. When clinical manifestations occur, these can be murmurs, disturbances of conduction, congestive heart failure, atypical chest pain, obstruction of the ventricular inflow or outflow tracts, or sudden death.[2,3] The tumours do not usually regress, and therefore need surgical resection.[2–5] We describe our experience here with a giant fibroma of the right ventricle, which was removed following the principles of the Batista procedure.

Case report

A 1-year-old girl was referred to the Latvian State Cardiology Centre for Children because of cardiac arrhythmias. The child was asymptomatic, and her development was in the normal range. On examination, she had a systolic murmur and an arrhythmia. The chest X-ray revealed an enlarged heart, and the electrocardiogram showed right axis deviation, slight right ventricular hypertrophy, and atrial ectopic beats.

Transthoracic cross-sectional echocardiography revealed a giant mass that occupied the right ventricular cavity without producing obstruction of the outflow tract. The tumour was localised within the myocardium of the anterior ventricular wall, measuring 40 mm × 30 mm, and had a volume of 110 mm^2. It was dense and circumscribed, not encapsulated, but incorporated hyperechogenic islands of calcified tissue (Fig. 1a). The left ventricular cavity was depressed from the right ventricular side. A pericardial effusion of about 5 mm was present. Spiral computed tomography confirmed the echocardiographic diagnosis. The decision to perform surgical treatment was made after the diagnostic procedures confirmed the presence of a giant fibroma of the right ventricle.

After performing a thoracotomy, the pericardium was opened and the external appearance of the heart was examined. The anterior wall of the right ventricle was rigid from the base to the apex of the heart. A zone measuring 30 mm by 70 mm of the anterior surface of epicardium of right ventricle was grey in colour, but the coronary arteries appeared normal, albeit that the area showed markedly decrease contractions.

Inspection through a right atriotomy revealed immobile leaflets of the tricuspid valve, and showed the right ventricular cavity to be filled by the mass of the tumour. Excision of the tumor through the tricuspid valve was not possible, so it was necessary to open the anterior wall of the right ventricle from

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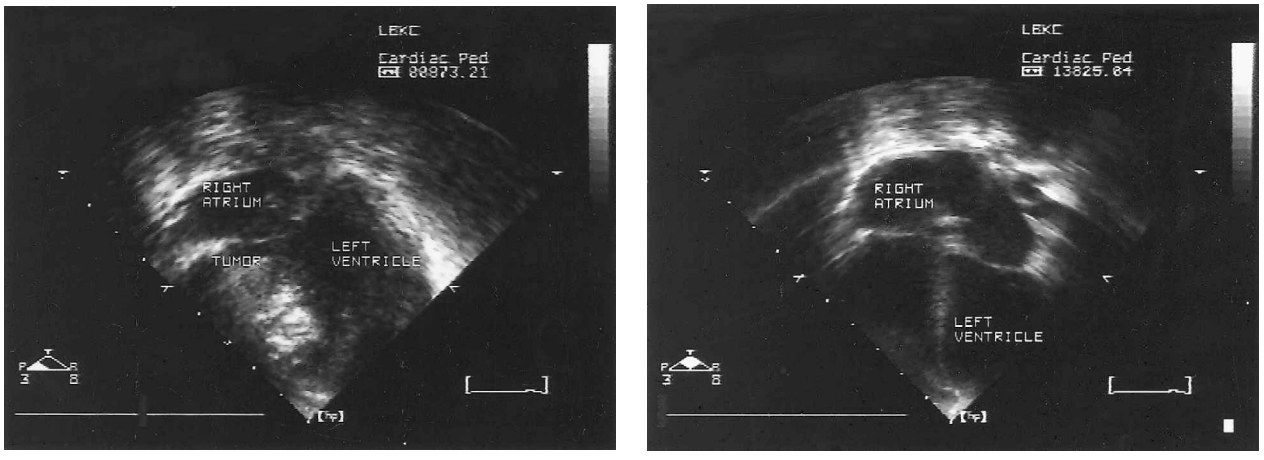

(a) (b)

Figure 1.
Preoperative transthoracic echocardiography in the apical four-chamber view (a) shows a giant tumour in the right ventricular cavity. Subsequent to removal of the tumour, comparable echocardiographic imaging (b) shows a normal right ventricular cavity.

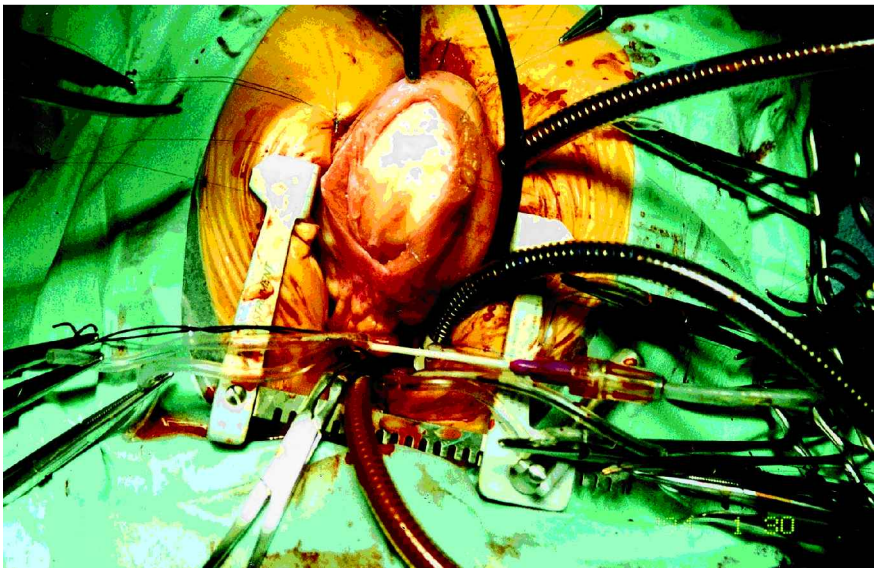

Figure 2.
The view of the tumour obtained by the surgeon at operation. It measured 75 mm by 55 mm by 30 mm.

the base to the apex of the heart. This showed the tumour to be rounded, firm, yellow- white- grey in colour, with extension from the supraventricular crest to the apex of the right ventricle, leaving only a small space between the tricuspid valve and the right ventricular outflow tract.

Sharp dissection permitted removal of the tumour, which measured 75 mm by 55 mm by 30 mm, leaving a bare area of approximately 30 mm by 70 mm in the anterior wall of the right ventricle (Fig. 2).

The papillary muscles of the tricuspid valve were normally developed, but had been displaced by the tumour. Approximately half of the remaining anterior wall of the right ventricle was visibly thinned after excision of tumour. Because of these findings, we anticipated that the contractility and ejection potential of right ventricle would be decreased. We decided, therefore, to follow the Batista principle, itself based on the law of Laplace, and reduced the size of the right ventricle, thus increasing its contractility, by partial ventriculectomy.[6,7]

A potentially akinetic zone making up half of the right ventricular anterior wall was resected, and the incision was sutured. Following this, the patient was weaned from bypass uneventfully. Surgical success was confirmed with postoperative transoesophageal echocardiography, which showed a normal right ventricular cavity with good contractility, but with trivial tricuspid valvar regurgitation. The subsequent postoperative course was uncomplicated, and the patient was extubated five hours after the operation with no need for inotropic support. Recovery was quick, and girl was discharged on the 6th day after operation, requiring only diuretic medication.

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Pathological examination confirmed the diagnosis of fibroma

In a post-operative follow-up, a 24-hour dynamic electrocardiogram and transthoracic echocardiography (Fig. 1b) proved that the patient was free from arrhythmic episodes, and had normal right and left ventricular function. Follow-up at one-year showed the child to be doing well, with no evidence of recurrence of the tumour.

Discussion

The exact nature of the cardiac fibroma is unclear, and there is ongoing debate regarding whether it represents benign neoplasia or is a hamartoma.[3] The tumour usually grows in the left ventricle, and involvement of the right ventricular free wall and cavity, and the atriums, is less frequent. Cardiac fibromas average 50 mm in diameter. Large lesions are known to obstruct the outflow tracts and compress the cardiac chambers.[2,3] Their growth is confined to the myocardial mass, but they can incorporate proximal segments of coronary arteries, this feature precluding complete surgical removal. Spontaneous regression has not thus far been observed, and surgical intervention is usually required. The tumour, however, often proves unresectable. Cardiac transplantation is then the only option, and has been performed in both children and adults.[3,5]

The tumour can usually be recognised using echocardiography.[4] Improvements in echocardiographic technology[3] now permit exact delineation of the location and extent of the tumour, its tissue characterisation, hemodynamic significance, and recognition of any associated pericardial effusion.

In our case, we approached the tumour through a right ventriculotomy. Due to thinning and akinesia of the anterior wall after excision of the tumour, we carried out partial ventriculectomy in order to reduce the size of the right ventricle and preserve its contractility. This approach is comparable to the one proposed by Batista for critically ill patients with dilated cardiomyopathy, in these cases using partial left ventriculectomy as the alternative to cardiac transplantation.[6,7] In our case, the principle has successfully been used for right ventricular problems.

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