

## Case Report

## Primary Intramural Fibroma of the Left Ventricle with a Late Apparent Symptomatology

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We report a case of a 13-year-old girl who was admitted to the emergency department with syncope. She had been diagnosed in infancy with a large intramural fibroma of the left ventricle, but had been free of any clinical symptoms during the last decade. The patient underwent a thorough and detailed clinical and imaging study. We discuss the contribution of new non-invasive imaging modalities to the diagnosis of primary cardiac tumours and focus on the dilemmas of the paediatrician concerning the management of cardiac fibromas.

Key words:

**Primary cardiac tumours, fibroma, childhood.**

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**P**rimarily cardiac tumours in childhood are rare, but they are extremely problematic as regards their diagnosis, differential diagnosis and therapeutic management. The most common types are rhabdomyoma, fibroma and myxoma. Fibromas are histologically benign, but their prognosis may be influenced by their location and size and by episodes of arrhythmias. The predominant picture may be one of obstructive phenomena (obstructive cardiomyopathy type), or heart failure (dilated cardiomyopathy type), or arrhythmias, which may be life-threatening. The surgical removal of fibromas is not always feasible.<sup>1,2</sup>

We present the case of a fibroma in the left ventricle with severe ventricular tachyarrhythmias in infancy, but with slow subsequent development and reappearance of symptoms in the early teens.

### Case description

A 13-year-old girl was admitted to the emergency department with dizziness, a tendency towards fainting and vomiting, blurred vision and tachycardia. When she was 4 years old, during investigation of a murmur, echocardiographic and histological examinations

had detected a large (3.5 × 5.5 cm) fibroma in the left ventricle, with no reduction in left ventricular systolic function. Fifteen months later she had a severe episode of ventricular tachycardia that was cardioverted by electrical defibrillation. During follow up the tumour showed slow intramural growth (maximum dimensions 7.2 × 7.7 cm), while on the clinical side the patient experienced sporadic episodes of tachycardia and only during the last two years she became easily tired.

Her growth and psychomotor development were normal. On clinical examination the patient was pale, she appeared unwell and had sinus tachycardia with occasional ectopic systoles, slight swelling of the liver and a systolic pulmonary ejection murmur. Haematological and biochemical examinations were normal.

Electrocardiographic monitoring showed sinus tachycardia with occasional ventricular extrasystoles while awake and sinus bradycardia with very rare ventricular extrasystoles during sleep. The chest X-ray showed left ventricular hypertrophy with an abnormal left cardiac border (Figure 1).

Echocardiography visualised a non-homogeneous mass with large dimensions located mainly on the lateral wall of the left



**Figure 1.** Chest radiography showing cardiomegaly with abnormal left cardiac border.

ventricle with extension to the interventricular septum. On admission there were signs of systolic and diastolic left ventricular dysfunction that disappeared after a few hours.

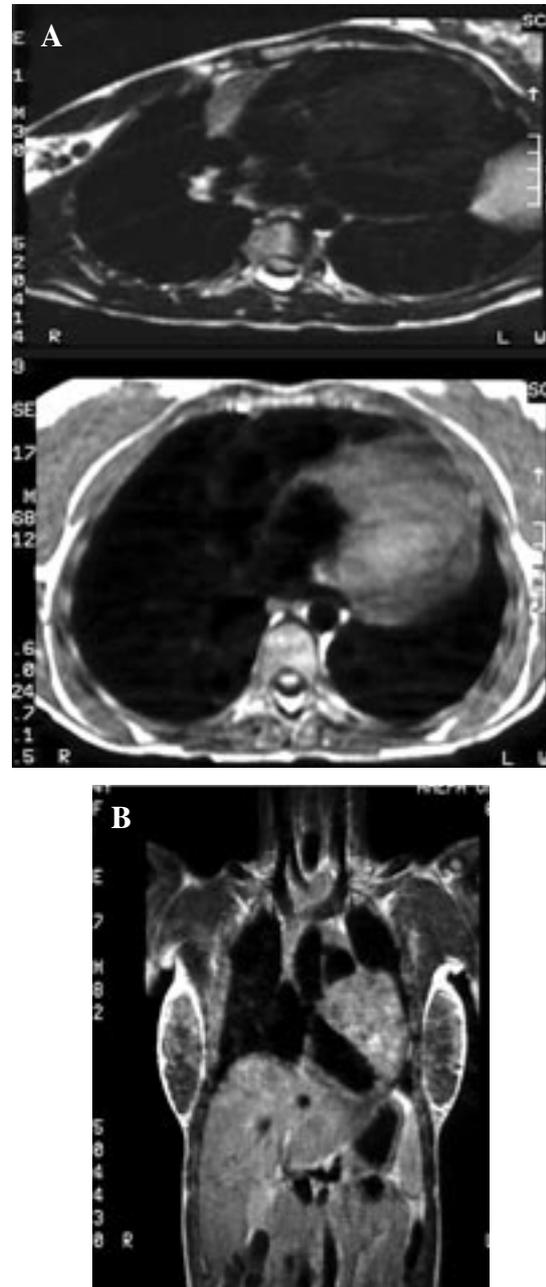
Magnetic resonance imaging confirmed the existence and the size of the tumour, which arose from the anterolateral wall of the left ventricle, extending to the interventricular septum and compressing the left atrium (Figure 2).

Cardiac catheterisation and coronary angiography showed displacement of the anterior descending branch by a non-vascularised intramural mass, normal trabeculation of the right ventricle and satisfactory systolic function of both right and left ventricles (Figure 3).

Under strict bed rest and medication with vasodilatory agents (captopril) and b-blockers from the next day, the patient showed a gradual improvement in her general condition, with restoration of a normal heart rhythm and rate. The clinical, electrocardiographic and echocardiographic follow up 6 and 12 months later showed no further change. Based on the rationale that this was an arrhythmogenic fibroma and that the syncope episode was probably due to a short episode of ventricular tachycardia, we recommended an implantable cardioverter/defibrillator, but the parents refused.

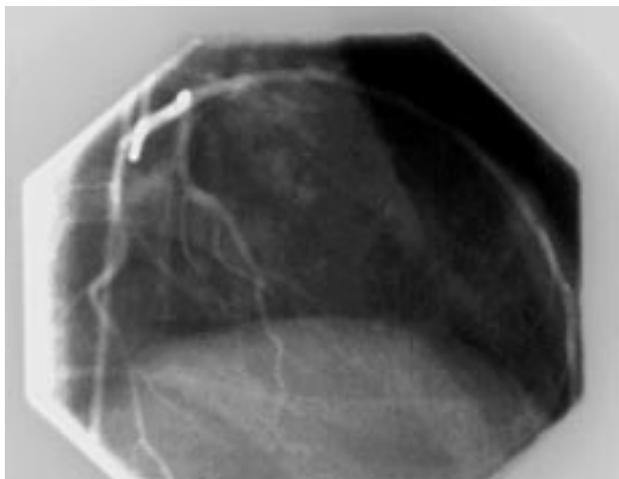
## Discussion

Primary cardiac tumours are rare at all ages, and especially so in childhood. Their incidence has been reported to range from 0.001-0.08% in hospitalised children or post mortem series (Table 1).<sup>1-5</sup> The case de-



**Figure 2.** A: Transverse magnetic resonance imaging sections in T1 and T2 sequences. A large mass arises from the anterolateral wall of the left ventricle, extends to the interventricular septum and compresses the left atrium. B: Coronal section of the magnetic resonance in T1 sequence after the injection of Gd-DTPA, showing the large size of the mass, its extensions and the weak enhancement.

scribed here was the third primary cardiac tumour and the first fibroma in a total of around 2500 cases of heart disease that required an echocardiographic examination in our Paediatric Cardiology Diagnostic Laboratory over a five-year period. Despite their rarity, primary



**Figure 3.** Coronary angiography showing displacement of the anterior descending branch by a non-vascularised intramural mass, normal trabeculation of the right ventricle and satisfactory systolic function of both right and left ventricles.

cardiac tumours are extremely problematic for the clinical physician<sup>1,3</sup> as regards their diagnosis and differential diagnosis,<sup>1-3,5</sup> their therapeutic management and their long-term prognosis.<sup>1-6</sup>

Our case concerned a histologically confirmed “arrhythmogenic” fibroma, whose existence was known from the patient’s infancy and which progressively increased in size without causing obstructive or embolic phenomena. After ten years without serious symp-

toms the patient showed manifestations of heart failure.

Cardiac fibroma is a congenital neoplasm that typically appears during childhood, in fact during infancy in one third of cases, and is usually discovered by chance, because of cardiomegaly or during the investigation of a murmur, as in our case. It is extremely common (14%) in Gorlin syndrome (basal cell nevus)<sup>5</sup> and is the most frequent primary cardiac tumour to be surgically removed during childhood.<sup>2</sup>

It manifests clinically as heart failure or arrhythmia, or it may be discovered during a post mortem as the cause of sudden death, especially in cases where the tumour invades the stimulus conduction system.<sup>7,8</sup> In a third of cases the fibroma is asymptomatic. Embolic phenomena have not been associated with fibroma.<sup>7-10</sup>

The fibroma is usually lone, with clear boundaries and a diameter that reaches 2-10 cm. Calcifications, isolated or multiple, are common, while cysts and haemorrhage are rare. In our case, at the time that arrhythmias appeared (mainly ventricular tachycardia) after 1-2 years, the fibroma had almost doubled in size.

The characteristic radiological picture of a fibroma is cardiomegaly, the “strange” cardiac silhouette, as in the present case, which is due to the tumour occupying the left ventricular free wall. In 25% of cases calcifications may be discovered radiologically.<sup>7,8</sup> The diagnosis may be confirmed rather easily by echocardiography or magnetic resonance imaging, which show high reliabili-

**Table 1.** Classification, frequency and prognosis of the primary cardiac tumors in childhood.

Tumour	Frequency	Prognosis	Surgical excision
Rhabdomyoma	Most common: 25%*	78% deaths <1 year	Problematic
Fibroma	15%*	Left ventricular arrhythmias, obstruction	Problematic
Teratoma	Intracardiac extremely rare	Extracardiac events	Possible
Haemangioma-angioma	Extremely rare		Possible
Myxoma	Most common in adults: 30%*	Embolic events, sudden death	Possible
Mesothelioma of the atrioventricular node		Arrhythmias	Not reported
Purkinje hamartoma	Very rare	Sustained tachycardia	Possible
Primary malignant tumours: • malignant teratoma • rhabdomyosarcoma • fibrosarcoma • neurogenic sarcoma	Rare	Tendency to take over the myocardium, invasion of the cavities	Problematic

\*Percentage of total primary cardiac tumours.

ty.<sup>11-16</sup> For intramural fibromas the occupied region of the myocardium is clearly hypokinetic.<sup>7,11-12</sup> The hypokinesia of the occupied region in our patient, as shown by all imaging methods, in combination with the likelihood of episodes of ventricular tachycardia (though not shown on repeated Holter monitoring) can explain our patient's clinical findings.

Whereas the detection of a fibroma with the echocardiogram is mainly morphological, magnetic resonance imaging describes in detail the volume, shape, and texture, with the characteristic images of fibrous tissue that are recorded.<sup>7,13-15</sup> With the use of contrast medium it is also possible to study the vascularisation of the tumour, which is usually poor.<sup>13</sup> In our case we performed cardiac catheterisation and coronary angiography when the diagnosis and functional consequences of the tumour were already certain, mainly in order to study the coronary arteries.<sup>11</sup>

In cases where the fibroma extends within the left ventricular cavity, its development is likely to mimic obstructive cardiomyopathy.<sup>1,8</sup> The clinical signs in our patient were probably due to sudden transient episodes of ventricular tachycardia, as stated above, and/or to a deterioration in the diastolic function of the ventricles, of which there were strong indications during the emergency echocardiographic examination.<sup>1,2,16</sup>

The main therapeutic aim is the treatment of the arrhythmias using antiarrhythmic medication tailored to the individual case, and this usually has satisfactory results. In the case of frequent arrhythmias the implantation of a defibrillator is recommended. In the present case the arrhythmia converted spontaneously and defibrillator implantation was recommended, rather than medication. Treatment of the fibroma entails its complete or partial surgical removal, when feasible, usually with beneficial results.<sup>7,17-19</sup> The site and extent of the fibroma in our patient did not permit its surgical removal, as in the past. In similar cases heart transplantation has been proposed and has been carried out with great success, but so far the survival rate does not seem to be very good.<sup>20</sup>

The potentially malignant behaviour of these tumours, because of the occurrence of heart failure, obstruction and arrhythmias, which do not rule out long-term survival, calls for constant alertness and treatment with medication, defibrillator or surgical means whenever feasible.

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