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Brief Report

Cardiac rhabdomyoma presenting as infective endocarditis: a case report

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Abstract We report a case of cardiac rhabdomyomas in an infant who presented with right ventricular tachycardia, and a clinical picture of infective endocarditis. Typical features of tuberous sclerosis developed subsequently. To the best of our knowledge, cardiac rhabdomyoma has not been reported previously in association with infective endocarditis.

Keywords: Rhabdomyoma; infective endocarditis; tuberous sclerosis

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Case report

A 3-week-old, weighing 4 kg male infant developed prolonged and vague febrile illness, accompanied with pallor. Subsequently, at the age of 2 months, he was presented to our emergency department with signs of compensated shock. His rhythm on the cardio-respiratory monitor revealed wide complex tachycardia at a rate of 300/minute. Twelve-lead electrocardiogram revealed a wide QRS tachycardia with left bundle branch block and inferior axis with 1:1 atrioventricular relationship. The tachycardia responded to cardioversion. He developed two other attacks of right ventricular tachycardia during the first 24 hours and a third attack on day 9 after admission. All attacks failed to respond to adenosine or amiodarone and responded to cardioversion.

Physical examination, when in sinus cardiac rhythm (100/minute), revealed that he was an acyanotic infant (oxygen saturation: 97% at room air). He was febrile (39°C) but had no respiratory distress. He had normal heart sounds and no audible murmur. The liver was 4 cm below the right costal margin and the spleen was not palpable.

Chest radiograph showed a cardiothoracic ratio of 0.6 and normal lung vascularity. The electrocardiogram when in sinus rhythm showed a frontal QRS axis of 75°, normal PR-interval (0.12 seconds), no Delta waves, no ST-abnormalities, and QTc equalled 0.36 seconds.

The echocardiogram revealed normal cardiac anatomy and contractility. Four homogenous, hyperechoic masses were present in the right ventricle: the largest mass measured 4.2 mm^2 and was located beneath the pulmonary valve; another oscillating, 2.4 mm^2 mass on the anterior tricuspid valve leaflet; and two masses measuring 3.2 and 2.1 mm^2 at the right ventricular apex. An intramural 4.5 mm^2 mass was seen at the apical interventricular septum (Fig 1) and an area of thickened myocardium was seen at the lateral left ventricular wall. Doppler evaluation revealed left to right shunt through a patent oval foramen and moderate mitral and moderate tricuspid valve regurgitation. No inflow obstructions and no pressure gradient across the right ventricular outflow tract in systole were observed.

Laboratory analysis revealed leucocytosis (19.100/mm^3), normocytic anaemia (haemoglobin 9.5 g/dl), and normal platelets count (431,000/mm^3). The urine was positive for microscopic hematuria. Four blood cultures were obtained, and all confirmed persistent staphylococcus epidermidis bacteremia, which was sensitive to vancomycin.

On the basis of the clinical and laboratory findings, and especially that this infant had a previous umbilical venous catheterisation at the neonatal unit, we considered the diagnosis of neonatal infective endocarditis. The patient was treated with

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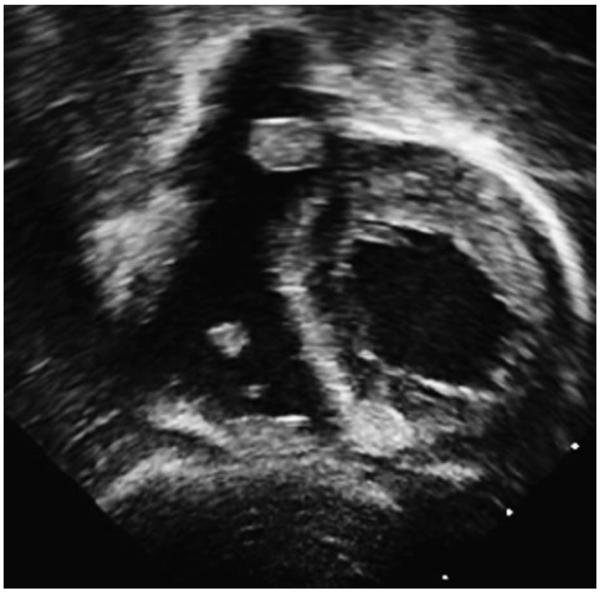


Figure 1.
Subcostal coronal view of the right ventricle showing the subpulmonic mass (4.2 mm^2), the tricuspid valve mass, and the intramural mass at apical interventricular septum.

vancomycin and Rifampicin, besides anticoagulation therapy in form of low molecular heparin.

Following the initiation of vancomycin after 15 days, he became afebrile and the blood culture was negative. A repeat echocardiogram showed a decrease in the grade of mitral and tricuspid valve regurgitation from grade two to grade one, and a decrease in the size of the subpulmonic mass from 4.2 to 2.7 mm^2 (Fig 2). The tricuspid valve mass decreased from 2.4 to 1 mm^2, and one of the right ventricular–apical masses had completely disappeared, the other decreased from 3.2 to 1.2 mm^2.

On the 3rd week after admission, he developed recurrent focal seizures that were controlled by anticonvulsant medications. The electroencephalogram revealed focal epileptic electrical activity, and the magnetic resonance imaging of the brain described subcortical and cortical wedge-shaped lesions, associated with multiple subependymal nodules; these findings are consistent with the diagnosis of tuberous sclerosis. Renal ultrasound displayed multiple renal cysts, and progressive systemic evaluations yielded the development of multiple, cutaneous hypomelanotic macules.

Following completion of 4 weeks of antimicrobial therapy, the echocardiogram showed no further change in the size of the cardiac masses. Doppler evaluation, though, confirmed mitral and tricuspid valve competence. The blood culture was negative for the pathogens following the discontinuation of antibiotics and the patient was discharged home in good health.

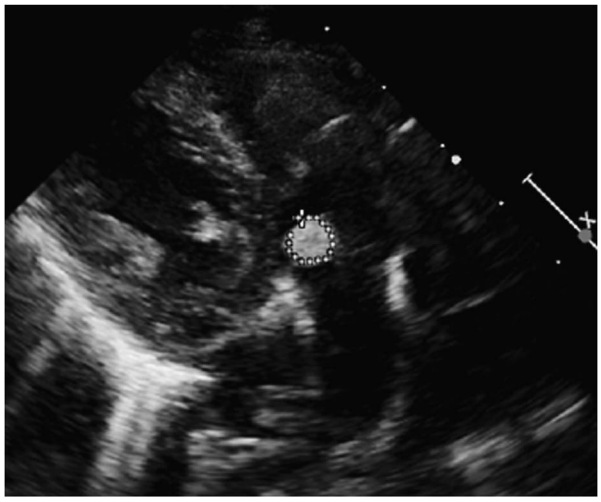


Figure 2.
Parasternal long axis through the right ventricular outflow tract showing the subpulmonic mass (2.7 mm^2).

He was followed up till the age of 18 months, he has been in normal sinus rhythm, and does not have any cardiac complication. However, spontaneous regression in the size of the subpulmonic cardiac tumour was first noted at the age of 14 months.

Discussion

Intracardiac masses can be tumours, vegetations, or thrombi. They are differentiated from one another on several grounds, the most important of which is the associated clinical and laboratory features. The clinical picture in our patient included diagnostic major and minor criteria for tuberous sclerosis. Cardiac rhabdomyomas frequently occur in association with tuberous sclerosis. Arrhythmia was reported in 16% of the paediatric patients with cardiac rhabdomyomas[1]; of these, 6% presented with ventricular tachycardia.[1]

Infective endocarditis has frequently been reported in association with cardiac tumours including cardiac myxoma[2–4] and fibroelastoma,[5,6] and the diagnosis of infected cardiac tumour was confirmed by histological examination of the surgically resected masses.[4–6]

Other clinical features in our case are prolonged fever associated with persistent staphylococcus bacteremia after umbilical venous catheterisation. In newborns and young infants, infective endocarditis is usually right-sided, and particularly in this age group, the classic manifestation of infective endocarditis may be few or absent. Thus, persistent staphylococcus bacteremia after a brief right-heart catheterisation requires considering the diagnosis of infective endocarditis.[7]

In our case, the resolution of mitral and tricuspid valve regurgitation supports the diagnosis

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of endocarditis. We believe that the intracardiac masses in our patient are not merely rhabdomyomas. The clinical response to antimicrobial and anticoagulation therapy along with the resolution of one and the decrease in the size of the other cardiac masses suggest that some of these masses were virtually infected thrombi, whereas the others were infected tumours. Owing to the fact that the natural history of cardiac rhabdomyoma is one of spontaneous regression and surgical intervention is usually not indicated,[8] the definite prove for infected rhabdomyoma by histopathological study could not be obtained in our case.

A variety of organisms have been implicated as a cause of infected cardiac tumours.[4–6] Review of the literature showed only one reported case of pulmonic valve myxoma, complicated by bacterial endocarditis because of staphylococcus epidermidis.[9] The similarity between that case and ours is that, in both, staphylococcus epidermidis endocarditis complicated the right-heart tumour that involved the pulmonary valve. To the best of our knowledge, this report is the first of cardiac rhabdomyoma in association with infective endocarditis. Thus, it suggests that, like other cardiac tumours, cardiac rhabdomyomas may get infected, and although this is limited to a case, it may have more favourable prognosis than other infected cardiac tumours.

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Conflicts of Interest

None.

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