CASE SERIES

# Prenatally diagnosed large intrapericardial rhabdomyoma without haemodynamic compromise

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## Abstract

Cardiac rhabdomyoma is the most common primary cardiac tumour and is considered to be a hamartoma of developing embryonic cardiac myocytes. It is commonly seen in tuberous sclerosis. The size and location of the tumour is the surrogate determinant of the risk of haemodynamic compromise. Pericardial rhabdomyoma is very rare and tends to follow the inherent natural history of spontaneous regression. We present cases of two foetuses diagnosed with large pericardial rhabdomyoma and no haemodynamic consequences.

**Keywords:** Foetus cardiac tumors, intrapericardial rhabdomyoma, prenatal diagnosis.

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## Introduction

Rhabdomyoma of the heart is one of the most common tumours of the heart and is considered to be a hamartoma of developing embryonic cardiac myocytes.[1] Around 60 to 80% of cases are associated with tuberous sclerosis.[2] It is commonly located intramurally and can be diagnosed prenatally by foetal echocardiography. We present two cases whose foetuses had large pericardial rhabdomyoma, diagnosed prenatally on echocardiography and their clinical outcome post-natally.

## Case 1

A 28 year old woman with a gestational age of 32 weeks, diagnosed with a cardiac abnormality on routine antenatal scanning was referred for echocardiogram on 3rd December 2019 to Liaquat National Hospital, Karachi. There was no family history of neuro-developmental abnormalities or seizure disorder. The foetal echocardiogram showed a huge tumour inside the pericardium measuring 33mmx18mm with trace pericardial effusion (Figure 1). The systolic function of both ventricles was qualitatively normal without valve regurgitation. The heart rhythm was normal with 1:1 conduction on pulse-wave Doppler. There was no evidence of structural heart defect or hydrops foetalis. Weekly follow up foetal echocardiograms did not show any significant change in the cardiovascular status.

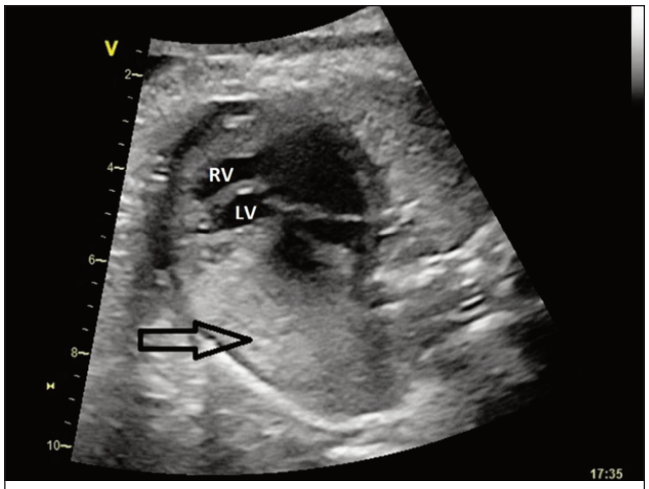


**Figure-1:** Case 1 Large pericardial rhabdomyoma (arrow). LV: left ventricle, RV: right ventricle.

The foetus was delivered at 35 weeks of gestation by Caesarean section. Cardiovascular examination was unremarkable. Postnatal echocardiogram showed a large tumour inside the pericardium with a few hyperechoeic intramural tumour masses in the left ventricular myocardium and interventricular septum. None of the intramural tumour masses or the pericardial tumour produced haemodynamic abnormalities. There was no pericardial effusion detected. Management was conservative with an echocardiographic follow up. There was a significant regression in the pericardial tumour by one year of age. Pathological confirmation was not sought. Magnetic resonance imaging (MRI) of the brain revealed tubers in the brain for which the child is being followed up by the neurologist.

## Case 2

A 21 year old woman with a gestational age of 26 weeks was referred on 21st March 2020 to Liaquat National Hospital, Karachi, for foetal echocardiography with the diagnosis of an extracardiac mass on routine antenatal ultrasound. Foetal echocardiogram showed a mass in the pericardium measuring 28x20mm, adjacent to the left ventricle, with no evidence of cardiac compression (Figure 2) or hydrops foetalis. Vascularity of the mass was not increased on colour flow mapping. There was one

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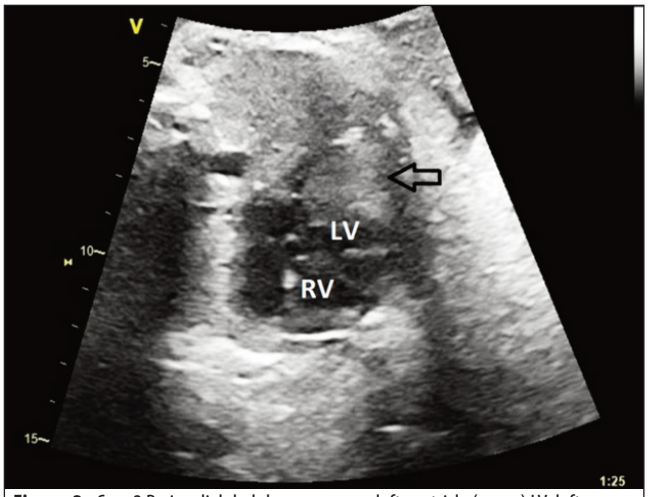


**Figure-2:** Case 2 Pericardial rhabdomyoma near left ventricle (arrow) LV: left ventricle, RV: right ventricle.

hyperechoeic area within the interventricular septum. The heart was structurally normal and there was no hydrops foetalis. The diagnosis was multiple rhabdomyoma, intrapericardial and small intramural, since the appearance was homogenous as opposed to the non-homogeneous teratoma.

The foetus was followed by frequent echocardiogram which was initially done weekly then fortnightly for hydrops foetalis or cardiac haemodynamic compromise. The foetus was delivered at term by Caesarean section and post natal echocardiogram confirmed the pericardial tumour of about the same size as in-utero.

At seven months of age, the size of the pericardial rhabdomyoma was reduced to 14x14mm and the intramural density was not seen. Pathological studies were not carried out as the tumour had regressed substantially. Screening for tuberous sclerosis was done by neurologist. His neuroimaging was significant for multiple tubers scattered all over cerebrum. The child had seizures twice and has been placed on antiepileptic medicines.

## Discussion

Primary tumours of the heart are rare and an intrapericardial location is even more uncommon.[3] Teratoma is considered to be the most common intrapericardial tumour and is easily diagnosed because it contains calcified foci and small cysts.

In a meta-analysis of 266 foetuses with cardiac rhabdomyoma, only one had it in the pericardium.[4] In that review, in 57% of mothers, the major indication for cardiac evaluation was the finding of a foetal cardiac mass during routine obstetric ultrasound. In 43%, foetal echocardiography was done because of either foetal dysrhythmia, hydrops foetalis or a family history of tuberous sclerosis.

Around 50-80% of the diagnosed cardiac rhabdomyomas have evidence of tuberous sclerosis. There may be either a positive family history or a neurological abnormality or an abnormal cerebral imaging.[2,4] The most common location of rhabdomyoma is within the ventricular myocardium, rarely in the atria, pericardium or epicardium, or cavo-atrial junction.[5] The differential diagnosis includes teratoma, fibroma, myxoma and haemangioma, in decreasing frequency, each having distinctive sonological features. Presence of multiple tumours particularly involving ventricular myocardium is highly suggestive of rhabdomyoma,[1] as was in case 1.

Complications include compression of cardiovascular or pulmonary structures or intracardiac conduction tissue, producing haemodynamic compromise. These complications best correlate with the size, and more importantly, with the location of the tumour.[5] Ventricular inflow or outflow tract obstruction would lead to decrease in cardiac output and increase in pressure within the atria and vena cava. This may lead to rise in hydrostatic pressures resulting in pericardial effusion, ascites or hydrops foetalis.[1,5] This invariably results in dismal outcome. Obstruction can be seen at any time during the gestational period and therefore warrants repeated follow-up ultrasound examinations till the foetus is born. Associated cardiac structural anomalies found include hypoplastic left heart, Fallot's tetralogy and endocardial fibro-elastosis which may be incidental or the result of outflow tract obstruction.[4] The natural history of cardiac rhabdomyoma, intramural or pericardial, is spontaneous regression.

Prenatal MRI is an advanced additional diagnostic modality which helps in detecting tubers in brain, renal or any other organ[4] but may not be easily available. However, absence of cerebral involvement does not indicate better postnatal outcome.

## Conclusion

Primary tumours of the heart are rare and pericardial tumours are even rarer. Rhabdomyoma happens to be the most common one with pathognomonic distinctive ultrasonography features. Since there is a strong association of foetal rhabdomyoma with tuberous sclerosis, foetal echocardiography is a useful modality for indirect early diagnosis of tuberous sclerosis. This will help in parental counseling as well as in making a post natal management plan to improve outcome of tuberous sclerosis.

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**Consent:** Both patients consented verbally for being published as case report.

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