

Cardiac rhabdomyoma leading to an early neonatal death

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Introduction

Primary cardiac tumours are rare. Over 75% of them are benign¹, the commonest being rhabdomyoma. Cardiac rhabdomyomas are usually multiple (92%) and often arise in the septum, ventricular wall or papillary muscles of infants and young children¹. Here we describe a cardiac rhabdomyoma in a newborn, which occurred in the interventricular septum.

Case report

A 37 year old woman, in her second pregnancy, was found to have polyhydramnios. Antenatal ultra-sound scan revealed an enlarged fetal heart with a 3.6×3.2 cm hyperechoic area in the interventricular septum of the heart. A live, non-asphyxiated baby girl, weighing 3.6 kg, was delivered by vacuum extraction at term. Apgar score was 10 at birth. The baby's breathing became progressively difficult from two hours after birth and she died 25 hours after birth in spite of adequate resuscitation.

The baby had no dysmorphic features. The heart was enlarged (Figure 1) and weighed 100g. The great vessels were normally distributed. There was a large lobulated tumour originating from the interventricular septum and projecting into the right ventricle. The tumour markedly reduced the volume of the right ventricular cavity. There were no congenital cardiac malformations. The tumour was lobulated and tan white in colour. It measured $5 \times 4.5 \times 3$ cm and weighed 70g (Figure 2).

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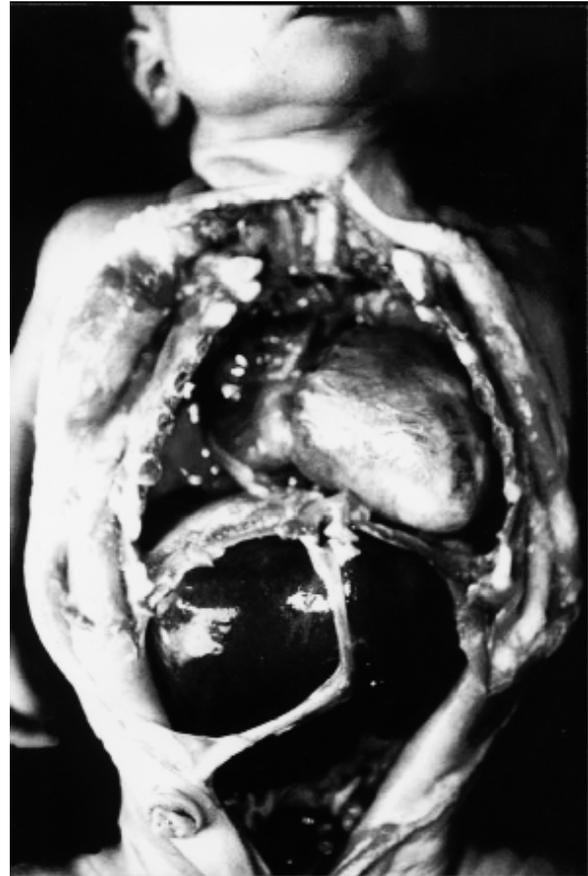


Figure 1. Enlarged heart almost occupying the entire thoracic cavity.

Histology confirmed the presence of a rhabdomyoma composed of tightly packed, round to polygonal cells separated from one another by thin fibrous septae and narrow vascular channels. Many of the cells contained vacuolated cytoplasm (Figure 3). Mitoses were absent. There were no cerebral features suggestive of tuberous sclerosis. The sibling of this infant had no features suggestive of tuberous sclerosis.



Figure 2. Cut section of the rhabdomyoma.

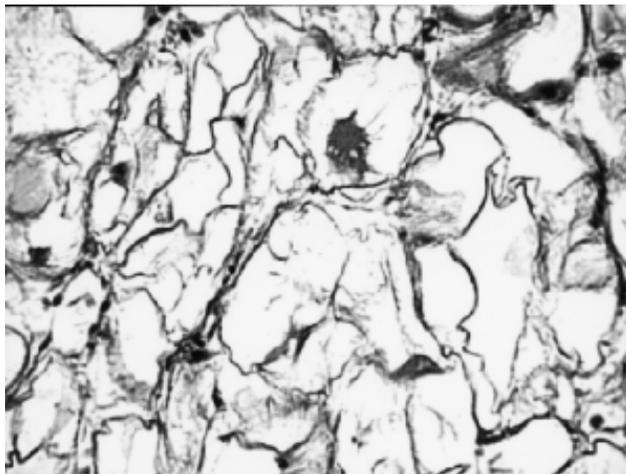


Figure 3. Vacuolated cells of rhabdomyoma

Discussion

Cardiac rhabdomyomas are currently regarded as hamartomas, most likely originating as a result of arrested cardiocyte maturation¹. Various studies have shown that they are slow growing in-utero and show no or minimal postnatal growth. They often occur as multiple intramural lesions¹. Although this patient did not have other features, in 30-80% of the cases it is associated with tuberous sclerosis¹.

Clinical features of cardiac rhabdomyoma depend on the size, site and the number of tumours. The presenting features may be tachycardia, arrhythmia^{2,3}, cardiac murmur, ventricular outflow obstruction⁴, complete or variable atrio-ventricular block, pericardial effusion, cardiomegaly, cardiac failure, Wolff-Parkinson-White syndrome⁵, or even sudden death. In the majority symptoms occur at early age or

even before birth^{2,6}. Cardiac dysrhythmias and non-immune hydrops indicate poor prognosis.

Chest X-Ray is usually normal or non-specific. In large tumours distortion or focal engorgement of the cardiac contour is seen. It may also produce an apparent mediastinal mass. Ultrasound scan can be used antenatally to diagnose cardiac rhabdomyoma.

Echocardiography (transthoracic/transoesophageal) shows a variable sized tumour in the myocardium/cardiac masses. It is useful in determining the size, number, location and evaluation of haemodynamic consequences of tumours. Serial studies are useful and safe in monitoring tumour size⁷. The optimal technique for displaying intra and extra cardiac masses is the MRI. It can depict intracavity tumours, myocardial tumours and extension beyond the heart. Cardiac angiography and spiral CT can be used in the detailed evaluation of the cardiac lesion.

Multiple postnatal management options exist. If asymptomatic, a conservative management is advised. When cardiac outflow obstruction, persistent arrhythmias, cardiac failure or cardiogenic emboli are present, surgical resection may be considered. Rarely heart transplantation is indicated⁸.

Partial or spontaneous regression has been reported in 54% of cases. Regression may take place in a period as short as three weeks¹. The chance of spontaneous regression does not depend on the initial size, number or location of the tumour. DiMario et al found that most lesions in patients followed with serial echocardiograms were undetectable by the age of 6 years⁹. Family screening for tuberous sclerosis is recommended.

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