

CASE REPORT

Dystrophic calcification of the fetal myocardium

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Abstract

Intramural cardiac masses were detected antenatally in three fetuses by echocardiography. The masses were initially thought to be rhabdomyomas. All three pregnancies were terminated and histology showed dystrophic calcification in all, with no evidence of tumour. Therefore, dystrophic calcification of the fetal myocardium may have a similar appearance to single or multiple rhabdomyomas. This should be considered when counselling parents after detection of masses in the fetal heart, particularly when considering the risk of associated tuberous sclerosis.

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Keywords: fetal myocardium; dystrophic calcification; rhabdomyomas; tuberous sclerosis

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Figure 1 Fetal echocardiogram of case 1. There are two apparently separate masses within the ventricular septum (arrowhead). (rv, right ventricle; lv, left ventricle.)

Tumours in the fetal heart are most commonly rhabdomyomas and, particularly when they are multiple, have a strong association with tuberous sclerosis.¹ After an antenatal diagnosis of cardiac rhabdomyoma parental counselling must include some assessment of the risk of severe disability associated with tuberous sclerosis.² This report concerns three fetuses with apparent intramyocardial masses, initially thought to have the features of rhabdomyomas. Each pregnancy was terminated, principally because of the possibility of these masses being markers of tuberous sclerosis in the fetus.

Case 1

Routine antenatal ultrasound at 23 weeks' gestation detected two echogenic intramural cardiac masses in the interventricular septum (fig 1). Both were 6 mm in maximum diameter but were not causing any obstruction to flow. Rhabdomyoma was thought to be the most likely cause and the possibility of tuberous sclerosis was raised with the parents who chose to terminate the pregnancy. Necropsy revealed extensive dystrophic calcification associated with fibrous scars in the interventricular septum but there was no evidence of rhabdomyoma. There were no other abnormalities and there was no evidence of any generalised fetal ischaemia.

Case 2

At routine ultrasound at 22 weeks' gestation there was a single large echogenic mass in the interventricular septum involving at least a

quarter of the entire septum. There was no evidence of any haemodynamic disturbance. Following parental counselling, the pregnancy was terminated for similar reasons to case 1. Necropsy showed a large scar associated with extensive dystrophic calcification in the ventricular septum. The rest of the examination failed to detect other sites of ischaemic damage or any other disease. Figure 2 shows the histology of the septal myocardium.

Case 3

An ultrasound scan was performed at about 16 weeks' gestation because of persistent vaginal bleeding. Extensive intrauterine haematoma was detected and the heart appeared abnormal. Detailed echocardiography at 24 weeks' gestation showed a large mass of variable density, which appeared lobulated, affecting at least 40% the ventricular septum and posterior wall of the heart. The mass appeared to cause obstruction to both inflow and outflow of the left ventricle and the left ventricular cavity was small. The pregnancy was terminated and necropsy showed a large fibrous scar with dystrophic calcification within the ventricular septum. The left ventricle, mitral valve, and aortic valve were hypoplastic and the myocardium generally had a mottled appearance. The rest of the examination failed to reveal evidence of ischaemic damage to other organs, and placental examination was unremarkable.

Discussion

Rhabdomyomas frequently arise in the interventricular septum, and are said to have particular echocardiographic features.³ Lobulation of a tumour or multiple lesions are said to be almost diagnostic of rhabdomyoma.³ In two of our cases these typical features were seen, but in the third there was a single, elongated mass involving an extensive area of myocardium. Our provisional diagnosis of rhabdomyoma led us to inform the parents of the possibility of an association with tuberous sclerosis and its potentially associated handicap,² and it was this particular aspect of counselling that appeared to be the major factor in the parental choice to terminate the pregnancy in each case. Despite our provisional diagnosis of rhabdomyoma being wrong in these cases, it seems likely that such extensive areas of myocardial necrosis would have led to some degree of compromise of cardiac function had the pregnancies progressed.

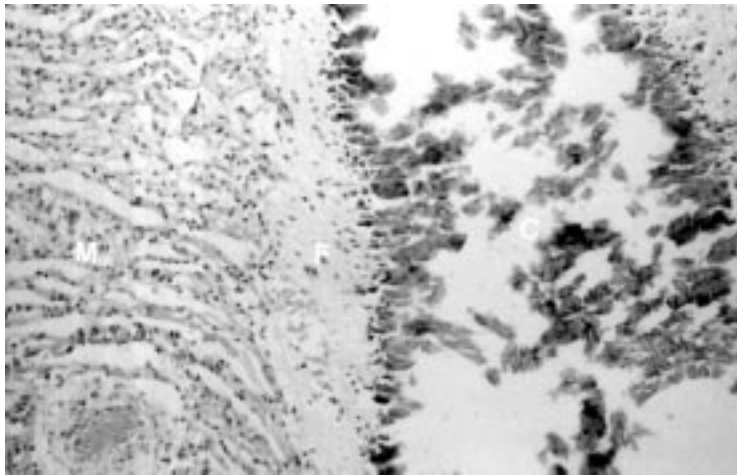


Figure 2 Histology of the septal myocardium in case 2. (M, normal myocardium; F, fibrosis; C, dystrophic calcification.)

Dystrophic calcification of the fetal myocardium is reported to occur in 12% of stillbirths,⁴ it occurs at sites of myocardial injury, and is probably just one aspect of a severe generalised ischaemic insult. Myocardial injury may occur in association with chorioamnionitis (for instance with chickenpox, cytomegalovirus or coxsackie B virus), with maternal cocaine abuse, placental abruption, and placental infarction.⁵ There is usually evidence of a generalised fetal insult. In case 3 there was clear evidence of earlier placental abruption, but necropsy failed to demonstrate any site of ischaemia apart from the myocardium. In our other two cases we also failed to find evidence of generalised fetal ischaemia, suggesting a regional cause of injury with subsequent myo-

cardial necrosis and calcification, although the underlying cause of this injury remains unclear. Similar mechanisms of injury may play a role in causing papillary muscle mineralisation in some fetuses with myocardial echogenic foci, although these foci are generally thought to be benign. The significance of dystrophic calcification of the fetal myocardium is uncertain, and its incidence in uncomplicated pregnancies is unknown, although in our own service catchment population of over 5 million it seems very rare. Cardiac rhabdomyoma is the most common intracardiac tumour both antenatally and in infancy with up to 86% of cases eventually found to be associated with tuberous sclerosis.^{6,7} Dystrophic calcification of the myocardium may closely mimic the ultrasound appearances of rhabdomyoma but may have very different implications for fetal outcome

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