

Isolated Fetal Cardiac Rhabdomyoma: A Case Report

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Citation

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Abstract

Foetal cardiac rhabdomyomas are the most common prenatally diagnosed cardiac tumours. They are known for their association with the tuberous sclerosis complex. We describe a case of an isolated cardiac rhabdomyoma which was picked up on a routine obstetrical ultrasound.

Cardiac rhabdomyomas are known for their association with tuberous sclerosis complex, an autosomal dominant multisystemic disorder. Therefore when diagnosed antenatally, attempt should be made to look for other stigmata associated with this complex.

We describe the case of an isolated fetal cardiac rhabdomyoma diagnosed on routine antenatal scan, done at 34 weeks' gestation. The tumor had an uneventful course in the prenatal period. At birth, the diagnosis of cardiac rhabdomyoma was confirmed. There were, however, no other stigmata of tuberous sclerosis in the neonate. The parents and others in the family were screened for presence of tuberous sclerosis and were found to be negative. The neonate had no evidence of hemodynamic compromise at birth, hence was kept under expectant observation for tumor regression. Serial evaluation of the tumor using echocardiography, showed no increase in the size at three and six months of life, with no deterioration of the cardiovascular status.

CASE REPORT

A 23 year old, primigravida was referred to our tertiary care hospital at 34 weeks' gestation, following a routine obstetrical ultrasound done elsewhere, which revealed a solid tumour in the foetal heart. She had no known antenatal risk factors.

Ultrasound done here, at the time of evaluation, showed no evidence of intra uterine growth restriction. Four chamber view of the foetal heart revealed a homogenously echogenic, solid mass, measuring one by one cm, arising from interventricular septum and abutting into the left ventricle. There was no evidence of pericardial effusion. The findings were consistent with that of cardiac rhabdomyoma. Umbilical artery Doppler studies were normal. There were no other gross anomalies on survey of foetal anatomy.

Foetal Echocardiography confirmed the presence of rhabdomyoma, away from the outflow and inflow tracts.

The patient was counselled regarding the possibility of tuberous sclerosis in the foetus. Parents and their first degree

relatives were screened for the presence of tuberous sclerosis with history and clinical examination and were found to be negative. Foetal growth was monitored closely. Serial ultrasounds were done to look for onset of foetal growth restriction, size of the tumour and development of hydrops. The size of the tumour remained same till delivery and afterwards.

Labour was induced at 40 weeks' gestation and she delivered a healthy male baby, weighing 2.7 kgs, by LSCS. Thorough physical and radiological examination of the neonate, however, did not reveal any other stigmata of tuberous sclerosis. There was no increase in the size of the tumour on neonatal echo done on third day and six months of life. The infant continues to be in regular follow up at our hospital.

DISCUSSION

The incidence of congenital cardiac tumours is 1-2 :10,000, and 90% of them are benign.¹ Of these, rhabdomyomas are the most commonly diagnosed in utero, followed by teratomas and fibromas. The reported prevalence is 0.25% in

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autopsies and 0.08% among the live born. 1,2

Rhabdomyomas are hamartomas derived from embryonal myoblasts. They may be solitary or multiple. Interventricular septum is the most common site of origin.

The prognosis of tumours detected prenatally is favourable, with most of them regressing spontaneously after birth. The risk of foetal demise is 4-6%.³ Poor prognostic indicators include inflow or outflow tract obstruction, leading to hydrops; atrioventricular valve dysfunction, leading to valvular incompetence and presence of dysrhythmias.

Cardiac rhabdomyomas are known for their association with tuberous sclerosis complex, an autosomal dominant multisystemic disorder. The risk of association is 30-50% , in case of a solitary tumour, and 70% with multiple lesions.⁴ When a cardiac rhabdomyoma is diagnosed prenatally, attempt to look for other features of this complex, should be made .

Standard obstetrical management is appropriate for uncomplicated cases. Delivery in a tertiary care centre with paediatric cardiology and surgery facilities is recommended.

After delivery, in the absence of haemodynamic compromise, expectant management is advised. Regular follow up with serial echocardiograms is the mainstay of management.

Figure 1

Obstetrical ultrasound depicting the transverse view of fetal thorax, at the level of the heart. The arrow mark showing the cardiac rhabdomyoma, which appears like a solid, homogeneously echogenic mass



Figure 2

Fetal echocardiography confirming the diagnosis of cardiac rhabdomyoma, seen arising from the interventricular septum



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