

Perinatal Management of Cardiac Tumors: A Case Series

A Amelia, MD, MB Mohd Nizam, MBBCh

Pediatrician, Pediatric Department, Hospital Sultanah Aminah Johor Bahru, Johor DT

SUMMARY

Primary cardiac tumours in the fetuses and neonates are uncommon. Foetuses with cardiac tumour have risk for heart failure and hydrops fetalis. Therefore, an early decision for delivery should be made in the evidence of foetal compromise. Early neonatal care varies on tumour size, type, location and obstructive features. Antenatal detection of foetal cardiac tumours ensures better prenatal and postnatal management. We describe our 5-year experience in managing 5 cases of primary cardiac tumours from 1st January 2006 to 31st December 2010

KEY WORDS:

Cardiac tumours; Rhabdomyomas; Atrial myxoma; Tuberos sclerosis; prostaglandin

INTRODUCTION

Primary cardiac tumours in the unborn foetus and neonate are rare with a reported incidence of 0.01 to 0.28 %¹⁻⁴. Most of these tumours are benign. However, some may cause heart failure and circulatory collapse leading to foetal demise or neonatal death. Fortunately, advances in foetal and neonatal imaging allow early detection of cardiac tumours during prenatal period and improve neonatal outcome^{2,4}. We describe our successful management of primary cardiac tumours diagnosed antenatally over 5 years duration.

CASE REPORT

A total of 5 cases of cardiac tumors diagnosed antenatally and confirmed immediately after delivery over a 5-year period, from 2006 to 2010. Table I summarized the demographic data, presentations, and outcome of patients. Four patients were detected in the later part of pregnancy which was between 32-38 weeks of gestations. None showed signs and symptoms of fetal heart failure. All had lower segment caesarian section (LSCS) (4 elective LSCS and 1 emergency LSCS for fetal distress).

3 newborns were symptomatic and cyanosed at birth requiring prostaglandin infusion to maintain their pulmonary circulation. Of these 3 patients, 2 had surgical resection. In Patient-2, the tumour was located in the right atrium (figure 1) with features of atrial myxoma and was confirmed histologically. Post operatively; he remains well with no tumour recurrence. Patient-4 has a single homogenous right ventricular mass which was obstructing the right ventricular inflow tract. Surgical intervention was later chosen for this patient as the compressive symptoms and signs persisted. The tumour was resected at day 14 of

life. He was discharged well at one week post-surgery. Histological examination result was consistent with rhabdomyoma. Patient-1, on the other hand, has multiple homogenous cardiac tumours on echocardiography findings. He was clinically treated as rhabdomyoma and required 9 days of prostaglandin infusion. The tumours later regressed and he was discharged home.

None of the neonates has family history of tuberous sclerosis (TS). However, further investigation showed TS in 2 patients (Patient-1 and Patient-3). Patient-1 has multiple ash leaf spots and subependymal tuber. At 4 years old, the tumours regressed completely and his development remains normal. Unfortunately, he developed a second-degree heart block (Morbitz type 1). As for Patient-3, he has subependymal hamartoma, bilateral renal cyst and ash leaf spots. At 13 months old, he has developmental delay and epilepsy. His cardiac tumour regressed to a measurement of 11 x 16 mm.

DISCUSSION

In our case series, antenatal detection of cardiac tumours is feasible. We found that serial assessments throughout pregnancy are important to assess the haemodynamic effects that the tumour has on the heart. Foetuses with cardiac tumour have risk for heart failure and subsequently foetal hydrop if cardiovascular flow obstruction develops. An early decision for delivery should be made if there is evidence of foetal compromise in order to prevent stillbirth¹⁻⁴.

We have observed that clinical manifestation of cardiac tumours varies with tumour size, its location and its potential growth. Tumours in the right atrium or tumours interfering with the conduction system may cause arrhythmias. Heart failure symptoms occur in obstructive cardiac tumours. The obstruction simulates a severe or critical valve stenosis^{2,4}. The clinical presentations include respiratory distress and haemodynamic compromise. The usage of prostaglandin infusion in this condition has been widely accepted². Prostaglandin infusion maintains Patent Ductus Arteriosus patency. This ensures adequate blood flow in pulmonary or systemic circulation while awaiting for further intervention or tumour regression². There are no data or guidelines available on timing of tumour regression or on the duration of prostaglandin infusion. Therefore, the decision is mainly based on serial echocardiography surveillance and physician own clinical judgment.

In our case series, 4 patients have cardiac rhabdomyomas and one with cardiac myxoma. Rhabdomyoma is the commonest type of primary cardiac tumours. The incidence of rhabdomyomas in primary cardiac tumours

This article was accepted: 22 February 2013

Corresponding Author: Amelia Alias, Pediatrician, Pediatric Department, Hospital Sultanah Aminah Johor Bahru, Johor DT
Email: ameaz76@yahoo.com

Table 1. The characteristic, presentation and outcome of neonate with cardiac tumour diagnosed antenatally

Patient	Gestational age	Mode of delivery	Clinical presentation at birth	Medical management	Type of cardiac tumours and location	Obstruction	Surgery	Family history of TS	TS feature	Current status
1	37 weeks	Emergency LSCS	Cyanosed at birth without respiratory distress	Prostaglandin infusion for a total 9 days	Rhabdomyomas: RA, RV, IVS and LV	Yes Partially block RVOT flow	No	No	Yes	4 yrs, Regress with secondary heart block
2	18 weeks	Elective LSCS	Cyanosed with respiratory distress	Prostaglandin till surgical removal	Atrial myxoma, RA cavity (Figure 1)	Yes Block RV inflow	Yes	No	No	3 years, Asymptomatic, no recurrence
3	32 weeks	Elective LSCS	Asymptomatic	Conservative	Rhabdomyomas: LV cavity and IVS (Figure 2)	Yes Partially block LVOT flow	No	No	Yes	13 months, Epilepsy, developmental delay, Tumour regress
4	36 weeks	Elective LSCS	Heart failure and cyanosis, ventilated	Prostaglandin infusion until surgical removal	Rhabdomyomas: RV cavity and RV free wall	Obstruction in the RV inflow and outflow	Yes	No	No	7 month, asymptomatic No recurrence of tumour
5	34 weeks	Elective LSCS	Asymptomatic	Conservative	Rhabdomyomas: LV cavity and LV free wall	No LVOT obstruction	No	No	No	2 month, asymptomatic and tumour regress in size

Abbreviations:

LSCS: Lower Segment Caesarian section, **RA:** Right Atrium, **RV:** Right Ventricle, **RVOT:** Right Ventricular Outflow Tract, **LVOT:** Left Ventricular Outflow Tract, **IVS:** Interventricular septum,

TS: tuberous sclerosis

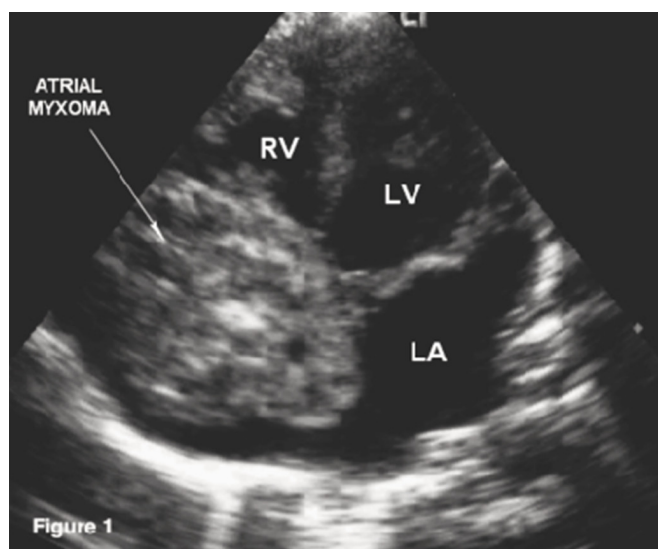


Fig. 1 : A four-chamber view of 2d-echocardiography showed a solitary, non-homogenous atrial myxoma occupies the right atrial cavity causing inflow and outflow obstruction in a newborn.
Abbreviations: LA: Left atrium, LV: left ventricle. RV: Right ventricle.

diagnosed during antenatally is more than 60%¹⁻⁴. Other cardiac tumours in newborns include myxoma, fibroma, haemangioma and intrapericardial teratoma¹⁻⁴. Patients with rhabdomyoma can be asymptomatic. On echocardiography, rhabdomyomas characteristically appear as homogeneous and hyperechogenic intra cardiac tumours²⁻⁴. Indifference to other tumours, rhabdomyomas naturally increase in size until 32 weeks of gestation and then subsequently regress and become clinically less important over time²⁻⁴. As most of the rhabdomyomas are benign, invasive endomyocardial biopsy is unnecessary^{3,4}. Classic histological findings of rhabdomyoma are large vacuolated cells with spider cells. The main treatment for rhabdomyomas is conservative management. There is no specific guideline on how to do surveillance echocardiography follow-ups in this group of patients^{3,4}. Surgical removal is considered in significant obstructive lesions causing haemodynamic compromised or severe arrhythmias. All neonate with suspected rhabdomyoma need to be screened for Tuberous Sclerosis (TS) as the association is in between 50 to 80%²⁻⁴.

TS is an autosomal dominant disorder with an occurrence of spontaneous mutation up to 80%⁴. TS has multi organ involvements including renal, central nervous system, retinal and skin. The diagnosis is mainly clinical which is based on the Tuberous Sclerosis Complex 1998 Consensus Conference clinical criteria⁴. The major clinical features of TS include subependymal nodule, cortical tuber, renal angiomyolipoma, shangreen patch and cardiac rhabdomyoma. The cardiac myxoma occurrence in a foetus, like in our patient, is very rare. The incidence is less than 1% of primary cardiac tumours. Typically patients with

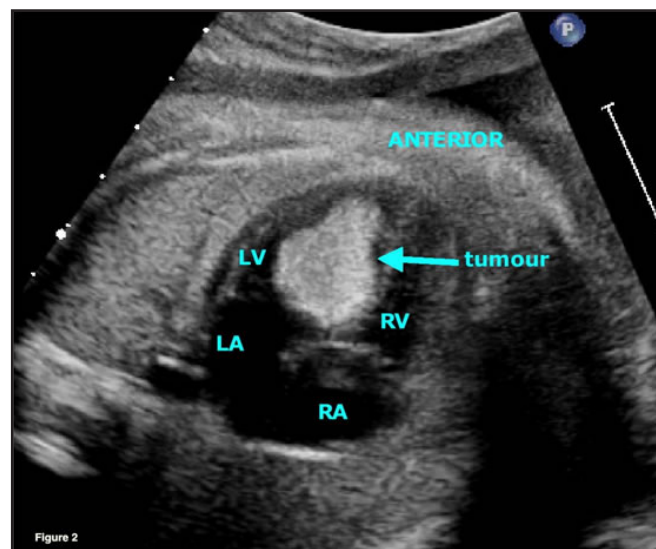


Fig. 2 : A four-chamber view of fetal echocardiography at 32 weeks gestation showed homogenous mass in the LV cavity and IVS with no dilated chambers or pericardial effusion.
Abbreviations: LA: Left atrium, LV: left ventricle. RV: Right ventricle.

myxomas are symptomatic with cardiac obstruction, risk of emboli and systemic illness. Echocardiography characteristic of a cardiac myxoma is a pedunculated, non-homogenous, echogenic and mobile mass. Most cardiac myxomas in neonates and infants are fatal. A complete surgical excision is the treatment of choice in cardiac myxomas^{1,5}.

CONCLUSION

Antenatal detection of foetal cardiac tumours ensures better prenatal and postnatal management. Rhabdomyomas are benign and can be managed conservatively as they will spontaneously regress with time. Prostaglandin infusion helps to improve cardiac output in obstructive tumours while awaiting surgical intervention or tumour regression. Surgical removal is considered in myxomas and severely obstructive cardiac tumours.

REFERENCES

1. Isaacs H Jr. Fetal and Neonatal Cardiac Tumors. *Pediatr Cardiol* 2004; 25 (3): 252-73.
2. Beghetti M, Gow RM, Haney I, et al. Pediatric Primary Benign Cardiac Tumors: a 15-year review *Am Heart J* 1997; 134(6): 1107-14.
3. Holley DG, Martin GR, Brenner JI, et al. Diagnosis and Management of Fetal Cardiac Tumors: A Multicenter experience and review of published reports. *J Am Coll Cardiol* 1995; 26(2): 516-20.
4. Bader RS, Chitayat D, Kelly E, et al. Fetal Rhabdomyoma: Prenatal Diagnosis, Clinical Outcome and Incidence of Associated Tuberous Sclerosis Complex. *J Pediatr* 2003; 143: 620-4.
5. Charokopos NA, Rouska E, Pliakos C, et al. Atypical atrial myxomas in two asymptomatic patients: a case report *Cardiovascular Ultrasound* 2009; 7: 45.