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Cardiac hemangioma of the right atrium in a neonate: fetal management and expedited surgical resection

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Abstract

Cardiac hemangioma is a rare tumor with a reported incidence of 1-2%. We describe the case of a neonate with a right atrial mass that was diagnosed prenatally. The fetus developed a supraventricular tachycardia and was delivered by cesarean section in the 35th week of gestation. The infant underwent surgery after 24 hours to remove the mass which was diagnosed as a cardiac capillary-cavernous hemangioma.

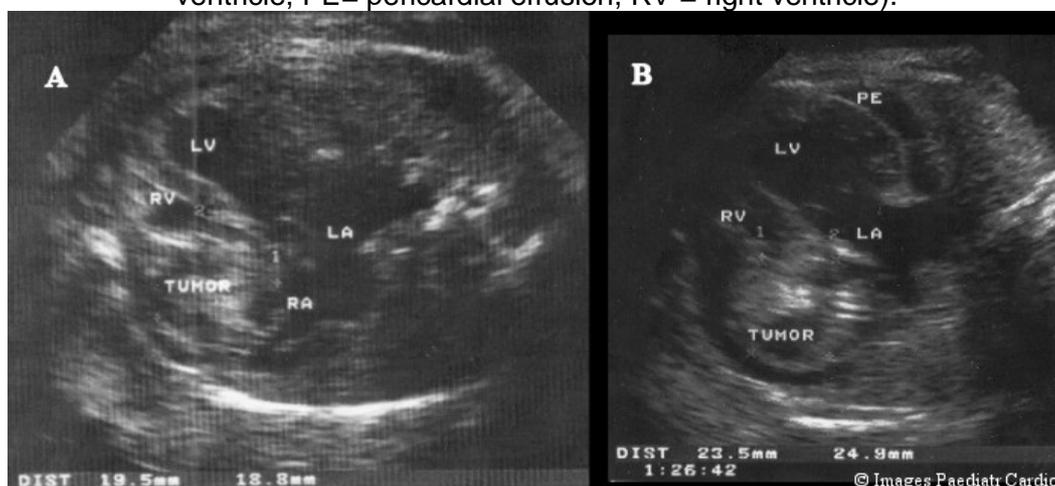
MeSH: Heart Atria/*pathology, Heart Neoplasms/*complications/pathology/*surgery, Child, Hemangioma/*complications/pathology/*surgery, Treatment Outcome

Case Report

The prevalence of cardiac tumors has been reported to be 0.05% in an autopsy study of infants only.¹ Cardiac hemangioma in infancy is extremely rare and resection of these tumors after prenatal diagnosis has been rarely reported.² There have been approximately 40 reported cases of primary cardiac hemangiomas in current cardiac literature.³ McAllister reviewed 533 primary tumors and cysts of the heart and pericardium of which 15 (2.8%) were hemangiomas.⁴ We report the case of a 1-day-old infant who had a right atrial tumor diagnosed on prenatal ultrasound, that was excised on the second day of life.

Recently a pregnant woman in the 25th week of gestation was evaluated by fetal echocardiography. A pericardial effusion had been detected on screening prenatal ultrasound and this had led to a fetal echo, which showed a large right atrial mass and moderate sized pericardial effusion. The tumor was based predominantly on the free wall of the right atrium and showed dramatic growth between the 29th and 31st week of gestation, with an increase in size from 12 by 13mm to 19.5 by 18.8mm (Fig. 1A). This rapid phase of growth then plateaued with the tumor reaching 23.5mm by 24.9mm in diameter by the 34th week (Fig. 1B). The echocardiographic appearance of the tumor was complex with multiple cystic spaces and initially was thought to be a teratoma.

Figure 1(A) Prenatal echocardiogram at 31st week showing a right atrial non-homogenous non-mobile mass approximately 19.5X18.8mm. (B) Prenatal echocardiogram at 34th week showing the right atrial mass approximately 23.5X24.9mm and moderate sized pericardial effusion. (LA= left atrium, LV= left ventricle, PE= pericardial effusion, RV = right ventricle).



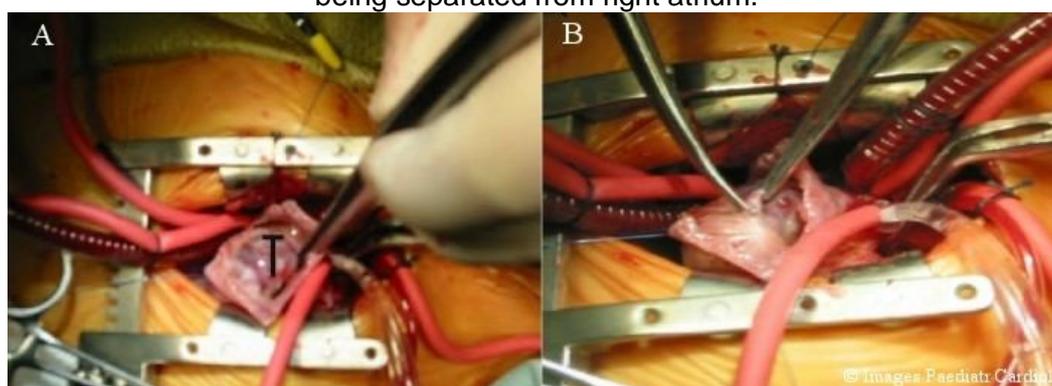
During the 35th week of gestation, the fetus developed a supraventricular tachycardia with a heart rate of approximately 188 beats per minute in association with mild tricuspid regurgitation and an increase in the size of the pericardial effusion. Due to concern over possible cardiac tamponade from the effusion the mother underwent a Cesarean section. Echocardiography confirmed the presence of a large right atrial tumor and a nonrestrictive patent foramen ovale. The patient was maintained on a prostaglandin infusion post-natally to ensure ductal patency as there was exclusive

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right to left shunting across the patent foramen ovale and moderate tricuspid regurgitation. At 24 hours of life the patient underwent open-heart surgery to excise the mass.

The chest was opened through a median sternotomy. Cardiopulmonary bypass was instituted with bicaval cannulation and the aortic cross clamp was applied with cold antegrade cardioplegia being delivered via the aortic root. A right atriotomy was made and the tumor was noted to be densely adherent to the free wall of the right atrium (Fig. 2) extending down toward the IVC and then curving back upwards onto the underside of the AV groove. There was a small secondary tumor mass just superior and medial to the orifice of the coronary sinus, close to where the AV node would be anticipated and this area was not excised completely, as resection would likely have injured the AV node. A second site of tumor was left at the junction of the right atrium and ventricle where it was very close to the overlying right coronary artery. A large patent foramen ovale was closed and the PDA was ligated. The cross clamp time was 25 minutes and the bypass time was 62 minutes. An intra-operative transesophageal echocardiogram revealed reasonable ventricular function with no evidence of shunts at either the atrial or ventricular levels and without evidence of injury to the tricuspid valve. The total volume of this tumor mass was thought to be approximately 7 cc.

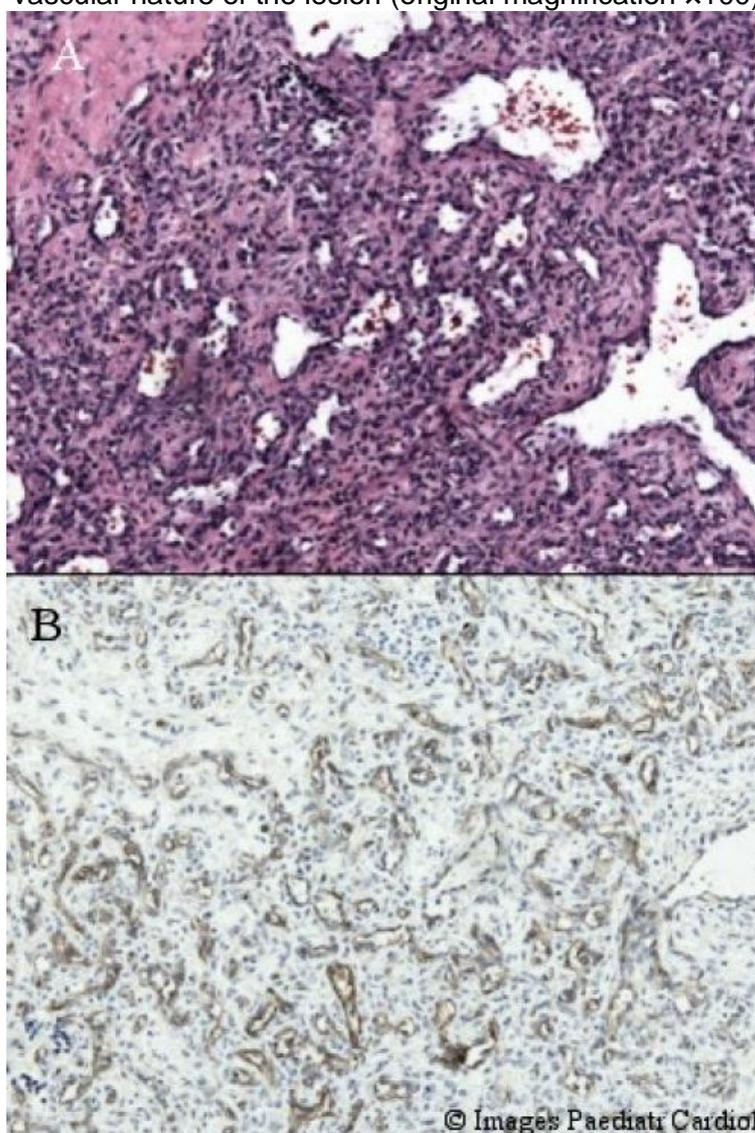
Figure 2 Operative view (A) Tumor (T) adherent to free wall of right atrium. (B) Tumor being separated from right atrium.



The infant returned to the Pediatric Intensive Care Unit and did well, being discharged at 2 weeks after surgery. Histopathological exam revealed a vascular lesion, composed of a mixture of small, slit like vessels and solid areas (Fig. 3A). The tumor appeared fairly well circumscribed but focally infiltrative. Ulex staining demonstrated the vascular nature of the lesion (Fig. 3B). There was no evidence of significant atypia and mitotic activity was moderate.

A diagnosis of cardiac capillary-cavernous hemangioma was made. The child is now 24 months of age and an echocardiogram in June 2005 showed residual 6 by 7 mm oval tumor near the area of the coronary sinus that has remained unchanged from previous post operative echocardiograms. A Holter monitor showed sinus rhythm with a single premature ventricular contraction over 24 hours.

Figure 3(A) H&E stain showing vascular channels with myxoid stroma in between (original magnification $\times 100$) (B) Ulex stains the blood vessels demonstrating the vascular nature of the lesion (original magnification $\times 100$)



Discussion

Cardiac hemangiomas consist of closely packed capillary structures (capillary type) or widely dilated vascular channels (cavernous type) with focal connective tissue in the walls. They are lined by endothelial cells and mitosis is rare.

This tumor can be localized in any part of the heart and pericardium. Hemangiomas can present in any age group with a mild predominance in females. The symptomatology depends on the anatomic location and extension of the tumor. Though most cardiac hemangiomas are discovered incidentally, they may cause dyspnea, palpitation, atypical chest pain and arrhythmia. Some of these tumors may also cause a pericardial effusion.

Echocardiography is usually the initial imaging modality. Coronary angiography shows the characteristic “tumor blush”. Recently CT and MR have been used in preoperative diagnosis and to evaluate extra cardiac extension and myocardial involvement.⁵

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There has been a reported case of spontaneous resolution of a cardiac hemangioma,⁶ but this is more typical of cardiac rhabdomyomas where upto 70% of tumors can regress spontaneously.⁷ The natural history of cardiac hemangiomas is variable and is the reason why all resectable lesions must be surgically removed. The long-term prognosis is favorable after adequate surgical resection.⁸ Unresectable tumors have a poor prognosis and may lead to sudden death due to arrhythmias. Non-surgical management of complicated hemangiomas has been described.⁹

The present case illustrates the multidisciplinary management of this rare cardiac tumor in a fetus. Prenatal diagnosis, early Cesarean section and then early surgery was achieved by involvement of maternal fetal medicine, pediatric cardiology and pediatric cardiac surgical services in regular fetal assessment meetings.

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