

Cardiac tumors in both twins – A case report of a rare occurrence

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ABSTRACT

Cardiac tumors in neonates and infancy are one among the many known congenital cardiac diseases. Although fetal cardiac tumors are rare, there is increased detection because of expertise in echocardiographic examination. Rhabdomyomas are the most common cardiac tumors among infants and children. Here, we describe twin neonates who had multiple cardiac tumors. This kind of presentation appears to be a very rare situation.

Keywords: Cardiac rhabdomyomas, multiple, tumors, twins

INTRODUCTION

Primary cardiac tumors are rare in all age groups, with an incidence of 0.0017%–0.027% at autopsy.^[1] In the infantile population, the incidence of primary cardiac tumors was reported to be 0.25%.^[2] Cardiac rhabdomyomas, teratomas, and fibromas remain the three most common types among fetuses to neonates and children. The echocardiographic examination has made a diagnosis of these tumors easy and early. The incidence of cardiac tumors in twins has been very rare. In cardiac rhabdomyomas, conservative management without intervention has been a standard management unless life-threatening complications are the consequences.

CASE REPORT

A 28-year female with monochorionic diamniotic twin pregnancy with a gestational age of 35 + 2 weeks underwent cesarean section and delivered baby girls, through a nonconsanguineous parentage. Both the babies cried after birth. Both babies were hemodynamically stable and had ash-leaf macules on examination. Twin I had intraventricular conduction defects with sinus rhythm. As there was a record of antenatally detected cardiac tumors in both the babies, echocardiography was performed on

both. Twin I had multiple well-defined nonpedunculated hyperechoic homogenous masses of various sizes seen attached to the walls of all chambers of the heart and septum [Figures 1 and 2] without outflow or inflow obstruction but had moderate left ventricular (LV) systolic dysfunction probably because of infiltration of the myocardium. Twin II had a similar picture on echocardiography [Figures 3 and 4] with normal ventricular function. In view of echocardiographic appearance, the possibility of cardiac rhabdomyomas was kept and planned for serial follow-up, as these kinds of tumors appear to regress on follow-up. However, the babies were discharged against medical advice and lost to follow-up.

DISCUSSION

Cardiac tumors can involve the endocardium, myocardium, or epicardium. Depending on their localization, size, and numbers, presentation varies and can cause arrhythmia, pericardial effusion, fetal hydrops, or even fetal death. In our cases, twin I had intraventricular conduction defect and LV dysfunction but was hemodynamically stable. The most common cardiac tumor in infancy and childhood is rhabdomyoma; fibroma, hemangioma, and teratoma being less frequent.

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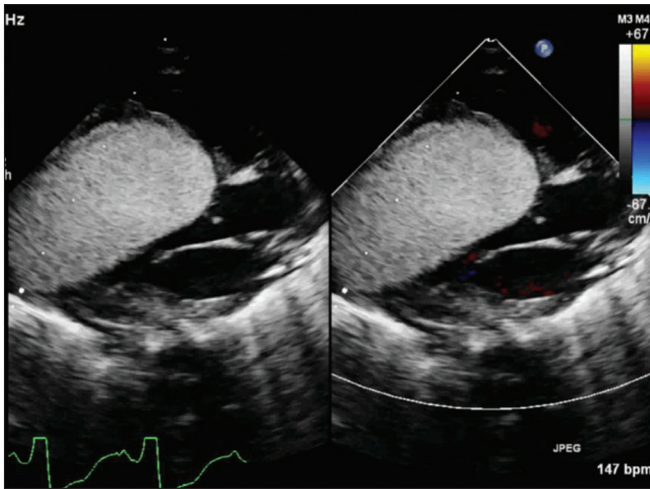


Figure 1: Echocardiogram of the twin I with parasternal long-axis view showing a large well-circumscribed hyperechoic homogeneous mass in the interventricular septum and smaller similar masses attached to the ventricular wall. Inflow and outflow portions are free

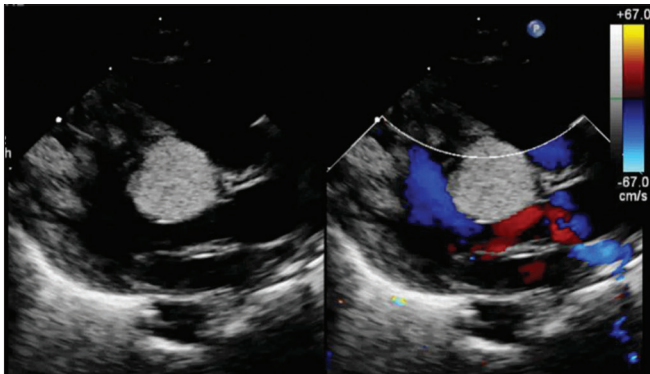


Figure 3: Echocardiogram of twin II with parasternal long-axis view showing multiple circumscribed hyperechoic homogeneous masses attached to the ventricular wall and septum. Inflow and outflow portions are free

Cardiac rhabdomyoma is the most common type of fetal primary cardiac tumor, with an incidence of 60%.^[3] The presence of multiple tumors involving the ventricular myocardium is indicative of rhabdomyoma.^[4] Cardiac rhabdomyomas are usually benign and involve the left and right ventricles and ventricular septum. The frequent association of rhabdomyoma with tuberous sclerosis has been demonstrated in previous studies. These tumors are well-circumscribed hyperechoic and homogeneous masses of variable size with a clear boundary. Appearance was very similar to our patients. Teratomas are usually right sided; single lesions usually attached to the base of the heart and appear heterogeneous and encapsulated on echocardiography. Fibromas are rare and are usually single lesions with hyperechoic echotexture involving the LV free wall or the interventricular septum.

Cardiac tumors can occur in the setting of a genetic syndrome, such as rhabdomyomas in tuberous

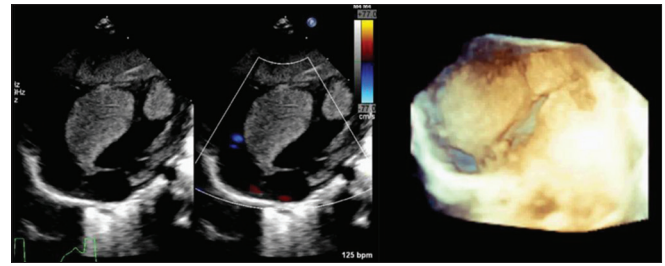


Figure 2: Echocardiogram of the twin I with apical four-chamber view and three dimensional echo image showing nonpedunculated circumscribed homogeneous masses attached to the ventricular free walls and septum. Inflow portions of both the ventricles are free

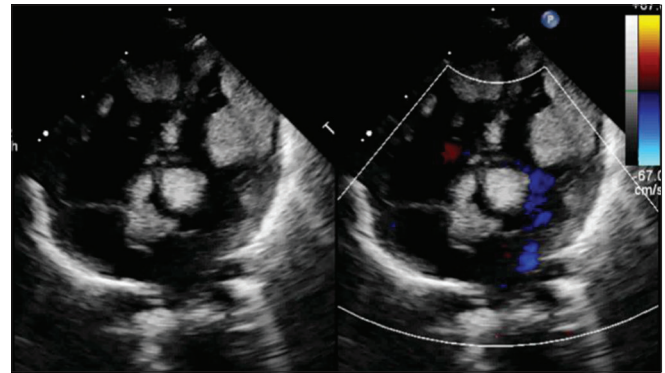


Figure 4: Echocardiogram of twin II with apical four-chamber view showing multiple circumscribed hyperechoic homogeneous masses attached to the ventricular wall and septum. Inflow portions of both the ventricles are free

sclerosis; fibromas in Gorlin syndrome; myxomas in Carney complex; and paragangliomas in association with multiple different syndromes including Carney triad, familial pheochromocytoma–paraganglioma syndrome, and von Hippel–Lindau syndrome. The presence of a genetic pathogenic mutation of tuberous sclerosis complex 1 (TSC1) and TSC2 is sufficient for a definite diagnosis of tuberous sclerosis. When a cardiac rhabdomyoma is detected on imaging, 40%–90% occur in the setting of tuberous sclerosis; however, this association with tuberous sclerosis increases to 100% if more than one cardiac rhabdomyoma is found.^[5] Gorlin syndrome associated with fibroma is an autosomal-dominant disorder associated with a mutation in the tumor suppressor gene *PATCHED* and Carney complex is associated with *PRKAR1A* gene. In meta-analyses by Chao *et al.* and Bejiqi *et al.*,^[6,7] there were 11 and 12 cases of fetal cardiac rhabdomyomas, respectively, which were either single or multiple but not associated with twins as seen in our scenario. Until now, after an intensive search of the literature, we found no cases of cardiac tumors which affected both of the twins in twin pregnancies. According to a study by Kadan-Lottick *et al.*, twin concordance for cancer was largely restricted to monozygotic twins and hematological malignancies with data not

contributing evidence for genetic factors in other cancers.^[8] Hence, incidence similar to our case needs much more evidence in future with respect to the genetic contribution.

Loss of functional myocardium because of tumor involvement may contribute to low cardiac output and congestive heart failure.^[9] If a cardiac function is normal, postnatal monitoring by echocardiography is sufficient in most cases, as the vast majority of cardiac masses show regression in numbers and dimensions. Hence, a conservative approach will suffice for many patients unless tumors cause life-threatening effects.^[10] Recently, everolimus has been used for the treatment of inoperable multifocal cardiac rhabdomyomas.^[11]

CONCLUSION

Cardiac rhabdomyomas are the most common tumor in fetal life, infancy, and childhood followed by fibroma, hemangioma, and teratomas. The presence of multiple tumors involving the ventricular myocardium is indicative of rhabdomyoma and these appear as well-circumscribed hyperechoic and homogeneous masses of variable size. The occurrence of rhabdomyomas in both of the twins appears to be extremely rare, and there is no previous reported case. Hence, we feel that this might be the first report of this kind of presentation. The conservative approach remains the management of choice unless the tumors cause life-threatening effects.

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Conflicts of interest

There are no conflicts of interest.

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