

Case Study

Infantile cardiac rhabdomyoma–Pearls inside the heart

ABSTRACT

Cardiac rhabdomyoma is the most common primary pediatric tumor of the heart. We reported a 1-month old male infant who was presented to our institute for routine cardiac evaluation since he was diagnosed to have a cardiac mass in the right ventricle (RV) in utero. After he was born, an Echocardiogram now showed two large cardiac masses occupying entire RV cavity and origin of right ventricular outflow tract (RVOT). Although our patient was asymptomatic, surgical removal of these two masses was done due to its proximity to RVOT and also because it was almost obliterating the entire RV cavity.

Keywords: Cardiac tumors; rhabdomyomas; right ventricular mass

INTRODUCTION

Cardiac tumors ~~was~~ were first described by Boneti (give a reference). Cardiac rhabdomyomas usually occurs as multiple masses, and arises commonly from the ventricular free wall and septal wall. There were 50-80% of patients to be have associated with tuberous sclerosis. Although cardiac rhabdomyoma has benign process and is known to undergo spontaneous regression, some may require surgical removal if the in certain situations like location was near or in the RVOT (spelled out), LVOT (spelled out), or when they produce intractable arrhythmias.

CASE REPORT

A 1-month-old male infant ~~child~~ presented to our institute for routine cardiac evaluation. Fetal echo had showed intracardiac mass at 30 weeks of gestation. The ~~babychild~~ was born to non-consanguineous parents. Tuberous sclerosis (TCS) screening was negative. The ~~babychild~~ was asymptomatic at the time of evaluation. A R routine echocardiogram (Figure 1) showed a large 3 x 1.2 cm mass occupying entire RV cavity and another 0.9 x 0.7 cm mass at

the origin of RVOT causing mild obstruction. In view of the size and location, despite being asymptomatic, the ~~infant~~child underwent successful surgical removal of the masses (figure 2) and was discharged uneventfully.

DISCUSSION

~~The prevalence of~~ Cardiac tumors ~~is~~are extremely rare in infants and children (0.027 to 0.17%). Cardiac rhabdomyomas is the most common type of cardiac tumors. More than 60% of antenatal ~~detected~~ cardiac tumors ~~is~~are rhabdomyomas and ~~it is~~these are often associated with tuberous sclerosis [1]. Cardiac rhabdomyomas ~~is~~are seen in 30-80% of patients with tuberous sclerosis and usually regresses spontaneously over time [2]. In addition, 50–60% of ~~tuberous sclerosis~~ patients with tuberous sclerosis will develop cardiac rhabdomyomas [6,7,8].

A meta-analysis showed that most rhabdomyomas associated with TSC (spelled out) are non-obstructive [3]. The natural history of cardiac rhabdomyomas in infants and children has been well studied [9]. The ~~y~~Younger the age at diagnosis, the higher the chance for spontaneous regression, and complete regression being more common in the first 4 years of life. In 2D echocardiogram, they appear as round, homogenous masses [4]. Indications for surgery included life-threatening arrhythmias, and outflow tract obstructions [5]. If complete surgical resection is not possible because of the location of the tumor, a partial resection can be done and the residual tumor usually regresses. The mortality from surgery for cardiac tumors in children is about 5% [10]

A ~~S~~serial echocardiography may help in selecting patients who are at risk of developing ventricular outflow obstruction and need early surgery. All cases of cardiac rhabdomyomas must be followed up closely for the future developed ~~suspected~~ tuberous sclerosis.

CONCLUSION

Although watchful waiting is the general strategy for cardiac rhabdomyomas, certain clinical situations like intractable arrhythmias and locations near outflow tract causing obstruction warrant early surgical intervention. Our patient underwent surgery empirically due to its location near the RVOT

~~outflow tract~~ and ~~massive size of~~ the tumor occupying almost the entire RV cavity. Because rhabdomyomas ~~is~~ are known to undergo spontaneous regression, general agreement ~~was~~ exists ~~that~~ indicated ~~edien~~ for surgical treatment which should be individualized and considered only in the presence of significant hemodynamic obstruction to the ventricular inflow or the outflow tract.

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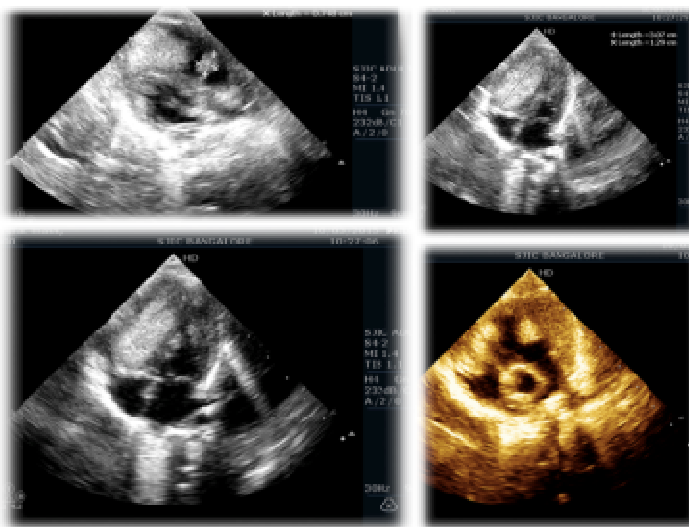
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109 **FIGURE LEGEND**

110 Figure 1: Trans thoracic echo showing rhabdomyoma in RV cavity and origin
111 of RVOT.

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113 **FIGURE 1**

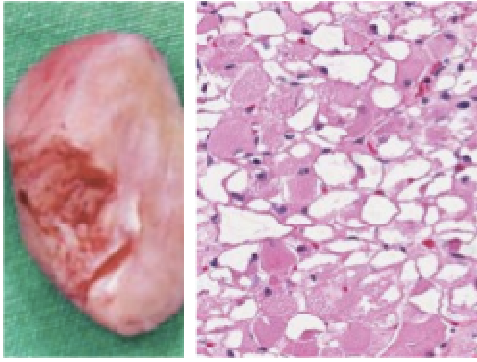


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117 Figure 2-Gross and histopathological specimen showing rhabdomyoma
118 extracted from RV cavity



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