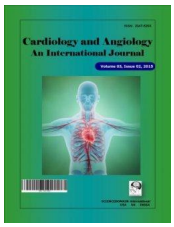

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Infantile Cardiac Rhabdomyoma–Pearls Inside the Heart

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Authors' contributions

This work was carried out in collaboration between all authors. Author PKA designed the study, Author CNM wrote the protocol, and author PB wrote the first draft of the manuscript. Author BB managed the literature searches. Author CDL recorded the echocardiogram. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Cardiac rhabdomyoma is the most common primary pediatric tumor of the heart. We report a 1-month old male infant who 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 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.

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Keywords: Cardiac tumor; rhabdomyoma; right ventricular mass.

1. INTRODUCTION

Cardiac tumors were first described by Boneti [1]. Cardiac rhabdomyoma usually occurs as multiple masses, and arises from ventricular free wall and septal wall. 50-80% of these patients had associated tuberous sclerosis. Although cardiac rhabdomyoma has benign prognosis and is known to undergo spontaneous regression, some may require surgical removal if the location was near or in the right ventricular outflow tract (RVOT), left ventricular outflow tract (LVOT), or when they produce intractable arrhythmias.

2. CASE REPORT

A 1-month-old male infant presented to our institute for routine cardiac evaluation. Fetal echo had showed intracardiac mass at 30 weeks of gestation. The baby was born to non-consanguineous parents. Tuberous sclerosis (TCS) screening was negative. The baby was asymptomatic at the time of evaluation. A routine echocardiogram (Fig. 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 underwent successful surgical removal of the masses (Fig. 2) and was discharged uneventfully.

3. DISCUSSION

The prevalence of cardiac tumor is extremely rare in infants and children (0.027 to 0.17%). Cardiac rhabdomyoma is the most common type of cardiac tumor. More than 60% of antenatally diagnosed cardiac tumors are rhabdomyomas and are often associated with tuberous sclerosis [2]. Cardiac rhabdomyoma is seen in 30-80% of patients with tuberous sclerosis and usually regresses spontaneously over time [3]. In addition, 50–60% of patients with tuberous sclerosis will develop cardiac rhabdo-myoma [4,5].

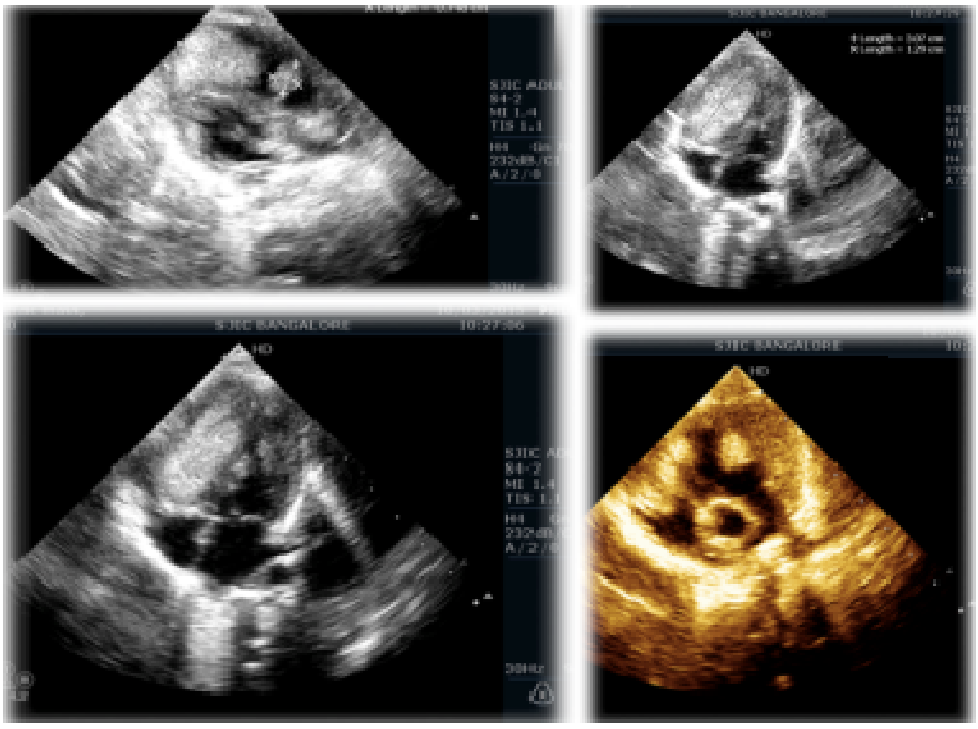

Fig. 1. Trans thoracic echo (TTE) showing rhabdomyoma in right ventricular (RV) cavity and origin of right ventricular outflow tract (RVOT).

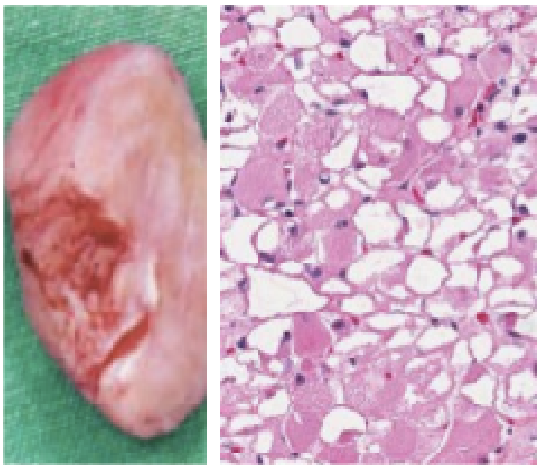

Fig. 2. Gross and histopathological specimen showing rhabdomyoma extracted from RV cavity

A meta-analysis showed that most rhabdomyomas associated with tuberous sclerosis are non-obstructive [6]. The natural history of cardiac rhabdomyoma in infants and children has been well studied [7]. The 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 [8]. Indications for surgery included life-threatening arrhythmias, and outflow tract obstructions [9]. 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 tumor in children is about 5% [10].

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

4. 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 and the tumor occupying almost the entire RV cavity. Because rhabdomyoma is known to undergo spontaneous regression, general agreement was indicated 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.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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