

Case Study

Infantile cardiac rhabdomyoma–Pearls inside the heart

ABSTRACT

Cardiac rhabdomyoma is the most common primary pediatric tumor of the heart. We report 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 in utero. 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 the 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 were first described by Boneti. Cardiac rhabdomyomas usually occur as multiple masses, arising commonly from the ventricular free wall and septal wall. 50-80% of patients have associated with tuberous sclerosis. Although benign and known to undergo spontaneous regression, some may require surgical removal in certain situations like location near or in the RVOT, LVOT, or when they produce intractable arrhythmias.

CASE REPORT

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

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35 **DISCUSSION**

36 Cardiac tumors are extremely rare in children (0.027 to 0.17%). Cardiac
37 rhabdomyomas the most common type of cardiac tumors. More than 60% of
38 antenatal detected cardiac tumors are rhabdomyomas and these are often
39 associated with tuberous sclerosis [1]. Cardiac rhabdomyomas are seen in
40 30-80% of patients with tuberous sclerosis and usually regress spontaneously
41 over time [2]. 50–60% of tuberous sclerosis patients will develop cardiac
42 rhabdomyomas [6,7,8].

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

53 Serial echocardiography may help in selecting patients who are at risk of
54 developing ventricular outflow obstruction and need early surgery. All cases of
55 cardiac rhabdomyomas must be followed up closely for suspected tuberous
56 sclerosis.

57

58 **CONCLUSION**

59 Although watchful waiting is the general strategy for cardiac rhabdomyomas,
60 certain clinical situations like intractable arrhythmias and locations near
61 outflow tract causing obstruction warrant early surgical intervention. Our
62 patient underwent surgery empirically due to its location near the RV outflow
63 tract and massive size of the tumor occupying almost the entire RV cavity.
64 Because rhabdomyomas are known to undergo spontaneous regression,
65 general agreement exists that indication for surgical treatment should be
66 individualized and considered only in the presence of significant
67 hemodynamic obstruction to the ventricular inflow or the outflow tract.

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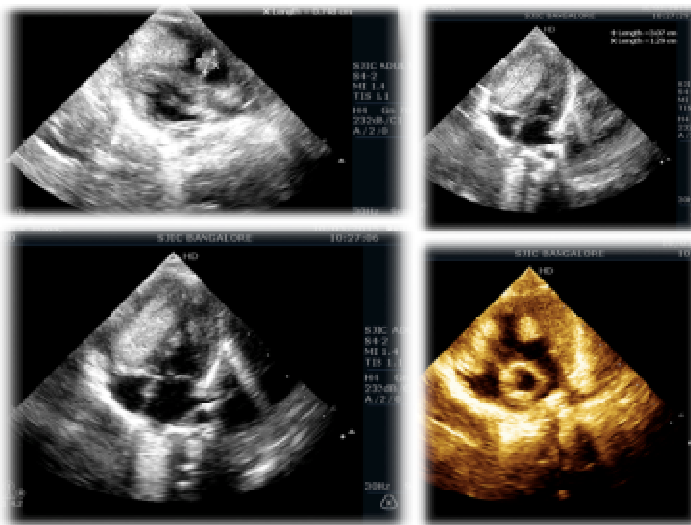
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103 **FIGURE LEGEND**

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

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107 **FIGURE 1**

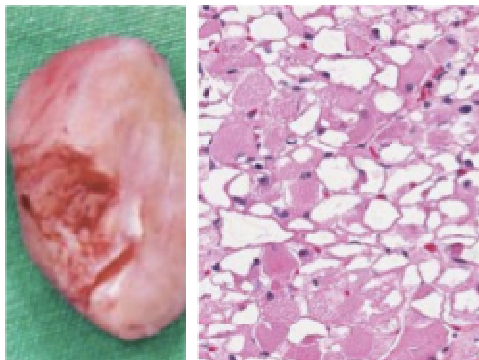


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111 Figure 2-Gross and histopathological specimen showing rhabdomyoma
112 extracted from RV cavity



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