

Case Study

Infantile cardiac rhabdomyoma–Pearls inside the cardia

ABSTRACT

Cardiac rhabdomyoma is the most common primary pediatric tumor of the heart .We report a 1month infant boy who presented to our institute for routine cardiac evaluation since he was was diagnosed to have a cardiac mass in the right ventricle in utero. Echocardiogram now showed a large cardiac mass occupying entire RV cavity and origin of right ventricular outflow tract(RVOT).The child underwent successful surgical removal of the masses.

Keywords: Cardiac tumors; rhabdomyomas; right ventricular mass

INTRODUCTION

Cardiac tumors were first described in the by Boneti.Rhabdomyomas occur as multiple masses,most commonly involving ventricular free wall and septal walls.50-80% of patients have associated tuberous sclerosis.Although benign and known to undergo spontaneous regression,may require surgical removal in certain situations.

CASE REPORT

A 1 month old infant boy 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-consanguinos parents.Tuberous sclerosis(TCS) screening was negative.The child now was asymptomatic.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.

DISCUSSION

Cardiac tumours are extremely rare in children (0.027 to 0.17%). Cardiac rhabdomyomas are the most common type of cardiac tumours. More than 60% of antenatally detected cardiac tumors are rhabdomyomas and these are often associated with tuberous sclerosis [1]. Cardiac rhabdomyomas are seen in 30-80% of patients with tuberous sclerosis and usually spontaneously regress over time[2]. A meta-analysis showed that most rhabdomyomas associated with TSC are non-obstructive[3]. In 2D echocardiogram they are round, homogenous masses [4]. Indications for surgery included life threatening arrhythmias, outflow tract obstructions[5]. 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 suspected tuberous sclerosis.

CONCLUSION

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

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FIGURE LEGEND

Figure 1:Trans thoracic echo showing rhabdomyoma in RV cavity and origin of rvot.

FIGURE

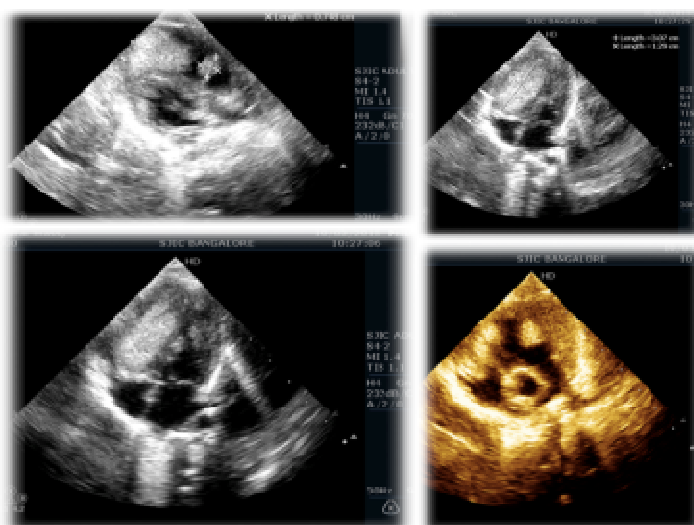


Figure 1:Trans thoracic echo showing rhabdomyoma in RV cavity and origin of rvot.

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

