Review Article

Indian Journal of Trauma and Emergency Pediatrics
Volume 10 Number 2, April - June 2018
DOI: http://dx.doi.org/10.21088/ijtep.2348.9987.10218.3

Cardiac Rhabdomyoma in Children

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Abstract

Cardiac rhabdomyoma is the most common cardiac tumor in children. It is being increasingly diagnosed because of visualization of these tumors by fetal echocardiography during prenatal evaluation. It is commonly associated with Tuberous sclerosis. Most of the lesions are asymptomatic and regress spontaneously. Less commonly it can cause blood flow obstruction and arrhythmia requiring treatment with pharmacotherapy or surgical excision.

Keywords: Cardiac Rhabdomyoma; Children.

Introduction

Primary cardiac tumors are rare in children. Cardiac Rhabdomyoma was first described in 1862 [1]. It is the most common primary pediatric tumor of the heart [2]. They are hamratomatous solid lesions containing striated myocyte fibres and in 90%cases there are multiple tumors. The tumors most frequently occur in ventricular myocardium but they may also arise in the atria. Inside ventricle, interventricular septum is most commonly involved (37%) followed by left ventricle (28%) and right ventricle (22%) [3]. Most of the patients are asymptomatic but the larger tumors may cause obstruction to the outflow tract of right ventricle or left ventricle. It may also cause arrhythmia. Most of the cardiac rhabdomyomyomas cases are associated with tuberous sclerosis and on the other hand children with Tuberous sclerosis commonly have cardiac rhabdomyoma. These tumors are rare in adult. Most of these tumors regress spontaneously and overall prognosis is good. Some tumors require surgical resection.

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Received on 02.04.2018, **Accepted on** 14.07.2018

Epidemiology

Incidence of cardiac rhabdomyoma is 0.002-0.25% at autopsy and 0.02-0.08% in live-born infants [4]. Because of the use of ultrasonography as part of routine prenatal screening, it is being increasingly diagnosed in fetuses in recent years [5]. Cardiac rhabdomyoma is commonly associated with tuberous sclerosis. The incidence of tuberous sclerosis in patients with cardiac rhabdomyomas is 60%–80%, and more than 50% of patients with tuberous sclerosis have rhabdomyomas [6,7].

Etiology

Cardiac rhabdomyoma may occur sporadically [6,8] but it is most commonly found associated with Tuberous sclerosis. Tuberous sclerosis is a multisystem genetic disease with autosomal dominant inheritance., causing hamartoma in brain, kidney, heart, lung,liver and skin. The genes responsible for Tuberous sclerosis are TSC1 at chromosome 9q34 (hamartin) and TSC2 on 16p13.3 (tuberin).

Clinical Features

Cardiac rhabdomyoma are frequently detected during fetal echocardiography during routine antenatal ultrasonography or during echocardiography during neonatal period and early childhood. It is rarely diagnosed after 10 years [9]. Most of the cardiac rhabdomyomas are asymptomatic. Clinical feature in symptomatic cases appear during neonatal period or early infancy. It may also cause hydrops fetalis and fetal death. The clinical features of cardiac rhabdomyomas depends on

the size, number and location of the tumor. Larger tumor may cause obstruction to the outflow tract of Left ventricle or right ventricle. Symptoms may arise because of chamber or valve obstruction, arrhythmias, or failure resulting from extensive myocardial involvement. It may cause serious arrhythmia which may sometimes become fatal. Obstructing lesion may cause low volume pulse. Cyanosis may be due to shunt reversal at atrial level. Tumor near Atrioventricular junction has been found associated with Wolff-Parkinson-White syndrome as it may form an accessory pathway, characterized by delta waves in ECG. Regression of the tumor is associated with subsidence of arrhythmia and Delta waves. Mechanical obstruction to valve may produce murmur in corresponding area. The physical examination of the newborn may be normal during neonatal period or it may manifest cutaneous lesions in the form of hypopigmented "mountain ash" macules or angiofibromas. Retinal astrocytic tumors may also be seen in newborn. Other cutaneous manifestations of tuberous sclerosis include café-au-lait pigmentation, subcutaneous nodules, subungual fibromas and linear epidermal nevi. It may also present as seizures during the neonatal period. A detailed family history should be taken and presence of any features of tuberous sclerosis in relatives should be asked.

Diagnosis

Rhabdomyomas are frequently detected by fetal echocardiography during the prenatal routine check-up. Lesions are multiple and appear as well-circumscribed homogenous hyperechoic masses. They involve the ventricles most commonly but may be found in atria also. MRI should be done if the results of echocardiography are inconclusive. They are isointense to slightly hyperintense relative to myocardium on T1-weighted images and hyperintense on T2-weighted images [10]. They may enhance less than myocardium after administration of intravenous contrast material. MRI is also helpful in revealing tuberous sclerosis in the brain, kidneys, and liver. Echocardiography and MRI are main diagnostic tools but Endomyocardial biopsy and histologic assessment may be done for the tissue diagnosis.

Treatment

Most cardiac rhabdomyomas in pediatric patients are benign tumors that can completely and spontaneously regress early in childhood [11,12]. Therefore it should be observed and follow up should be done with echocardiography to look for regression. Treatment is required for the tumors causing considerable outflow tract obstruction, hemodynamic disturbances, or arrhythmias. Arrhythmia is treated initially with antiarrhythmic medication. If arrhythmia is not controlled with pharmacologic medication or tumor is causing significant hemodynamic abnormality, surgical resection is indicated. Operation is indicated for lesions near or directly involving the cardiac valves that considerably obstruct blood flow [13] or cause valvular insufficiency [14]. These intracardiac tumors may produce either acute or chronic heart failure. Tumor resection may lead to cessation of the arrhythmia when the tumor impinges on the conduction system or when the tumor represents an arrhythmogenic focus. Thus, indications for operation may include left or right ventricular outflow tract obstruction, mitral valve insufficiency, and intractable arrhythmias. Successful results have been reported for the operative management of cardiac rhabdomyomas for indications involving obstructive lesion and arrhythmia [12,14]. Although resection can be lifesaving for certain cardiac rhabdomyomas, it is not necessary for most of these tumors. In patients with multiple cardiac rhabdomyomas, only the lesions causing these problems need to be resected. This policy of subtotal excision also means that complete excision of an individual tumor is not necessary. It is only necessary to resect the obstructing portion of the tumor. Incomplete excision may not be suitable when intervening surgically for the treatment of arrhythmias. As most cardiac rhabdomyomas regress spontaneously, it is reasonable to observe the majority of patients and carry out subtotal resection. Adrenocorticotropic hormone and Vigabatrin are mainstay of therapy for seizure in tuberous sclerosis [15]. Everolimus, an inhibitor of mammalian target of rapamycin has recently been found to cause rapid regression of the tumor [16].

Prognosis

The prognosis depends on the size and site of the tumor. Overall the prognosis is good and most of the cardiac rhabdomyoma undergo complete or partial regression with resolution of symptoms. The survival rate is 81% to 92% [17]. The larger tumor with diameter more than 2 cm are more likely to to cause hemodynamic disturbances and arrhythmias. The Tumors affecting valve function may cause regurgitation and are associated with a poor prognosis. As the natural history of cardiac rhabdomyoma is spontaneous regression the symptomatic patients are managed conservatively for arrhythmia and regurgitation with follow

up of the lesion with electrocardiography and echocardiography. Treatment with Everolimus, a rapamycin inhibitor has shown rapid regression of tumor in symptomatic cases. The prognosis also depends on the neurological symptoms associated with Tuberous sclerosis.

Conclusion

Cardiac rhabdomyoma is increasingly being diagnosed because of wider use of ultrasonography during recent decade. The disease is frequently associated with the tuberous sclerosis. Besides treating symptoms of tuberous sclerosis, the treating physician should take care of the cardiac lesion also. As the natural history of the lesion is regression over time, the symptomatic cases requires to be managed conservatively with follow up with ECG and Echocardiography. Some of the larger lesions causing significant obstruction and uncontrolled arrhythmia require surgical resection.

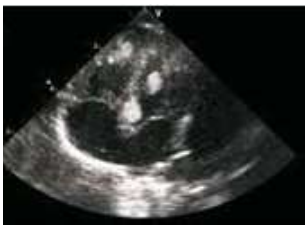

Fig. 1: Apical four chamber view in Echo showing Rhabdomyoma in both ventricles.

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