Case Report

Incessant ventricular tachycardia due to multiple cardiac rhabdomyomas in an infant with Tuberous Sclerosis

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1. Introduction

Cardiac rhabdomyomas are the most common tumors in infants and children. Majority are asymptomatic and diagnosed either during fetal life or during routine screening by transthoracic echocardiogram. The presentation is generally with symptoms of cardiac failure due to multiple masses, intracardiac obstructions leading to hemodynamic adversity or rhythm disturbances. We report a case of cardiac rhabdomyoma in a young infant presenting with ventricular tachycardia as initial manifestation.

2. Case report

A two-month-old boy, weighing 4.5 kg, was brought to Emergency Department with one day history of sudden onset of respiratory distress. On examination, he was afebrile but restless. There were features of low cardiac output, heart rate was 230 bpm, respiratory rate of 60/min and SpO2 95%. The blood pressure was 75/40 mmHg. Cardiovascular system examination revealed tachycardia and no murmurs. There were crepitations over both lung fields and the liver was 2 cm below the costal margin. Electrocardiogram showed wide QRS
tachycardia with left bundle branch block pattern and left axis deviation. There was an initial positive R in avR suggestive of ventricular tachycardia (Fig. 1). A non-synchronous DC shock of 10 J terminated the tachycardia, however, tachycardia recurred in few minutes. Subsequent DC shocks with higher dose did not result in conversion. Intravenous lidocaine infusion at 30 mcg/kg/min was started after an initial bolus of 1 mg/kg. The ventricular tachycardia persisted for few hours and hence, amiodarone was given 5 mg/kg bolus followed by infusion at 10 mcg/kg/minute. Oral propranolol (2 mg/kg 8 hourly) was added after 6 h in view of refractory nature of tachycardia. The ventricular tachycardia was converted to sinus rhythm after 2 h.

Transthoracic echocardiogram was performed during normal sinus rhythm which showed multiple well circumscribed hyperechogenic intramural masses with intracavitary protrusion involving free wall of the ventricles, interventricular septum and the outflow tracts (Fig. 2a). There was no hemodynamic compromise due to tumors (Fig. 2b). There was no pericardial effusion and the ventricular systolic function was normal. The diagnosis of cardiac rhabdomyomas was considered in view of multiplicity of tumors and their intramural location. Baby improved well clinically and remained in sinus rhythm. The child was discharged home on oral amiodarone and propranolol.

On follow up at 3 months of age, baby was symptom free, ECG showed normal sinus rhythm. Assessment by transthoracic echocardiogram showed regression in the size of tumors. However, a computerized tomography of brain performed in view of new onset myoclonic jerks at 5 months of...
life showed subependymal tubers confirming the Tuberous Sclerosis.

3. Discussion

Cardiac rhabdomyomas are commonly seen in Tuberous Sclerosis which is an autosomal dominant disorder. Most of the cases of cardiac rhabdomyoma are asymptomatic. Echocardiography is usually sufficient for making the diagnosis of these tumors. However, cardiac MRI is particularly useful for defining the extent of the tumor and its relationship to the surrounding structures when surgical excision is planned. It is also an important diagnostic tool in atypical presentation, like solitary tumor. Rhabdomyoma is typically solid and homogeneous hypointense to isointense on T1-weighted and slightly hyperintense on T2-weighted MRI. They show minimal or no enhancement with gadolinium. In contrast, cardiac fibroma one of the commonest solitary tumor in children appears isointense to hyperintense on T1-weighted and hypointense on T2-weighted images, findings characteristic of fibrous tissue.

Although almost all types of arrhythmias have been described with intracardiac tumors, the mechanism for majority of these rhythm disturbances remains elusive. Few mechanisms described are; tumor mass interfering with valvular function or reentry around its border with the normal myocardium. It is not always the accessory pathways like Bundle of Kent that mediate tachycardia in patients with cardiac tumors but a part of intracardiac tumor or pathways closely related to the tumor can also result in ventricular preexcitation. Hence, radiofrequency ablation is possible in at least a few patients with accessory pathways associated with intracardiac tumors. Cardiac rhabdomyomas presenting as ventricular tachycardia are rare and were seen only in 6% of cases of ventricular tachycardia as compared to fibroma which was diagnosed in 64% of all ventricular tachycardias in one of the largest retrospective studies of cardiac tumors.

Initial treatment with antiarrhythmic agents like propranolol and sotalol have been shown to be successful in controlling ventricular tachycardia. Resecting the tumor can result in a sustained resolution of symptoms and a likely cure but it is not possible if tumor involves critical structures or multiple intramural sites like in our case. Implantable Cardioverter Defibrillator is appropriate therapy in such cases.

Transvenous ICD implantation is difficult in children due to small patient size, complex venous or cardiac anatomy and intracardiac shunting. Epicardial patches if used, require thoracotomy. They may also lead to a restrictive pericardial process. Hence novel ICD configurations are explored like subcutaneous array, a transvenous design ICD lead placed on the epicardium, or a transvenous design ICD lead placed subcutaneously.

Conflicts of interest

All authors have none to declare.

References