Tuberous sclerosis with Cardiac Tumor in fetus with Diabetic Mother

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Abstract

Introduction

A heart tumor in children is rare and the most primary tumor of the heart is rhabdomyoma. We report a case of cardiac mass diagnosed at 32th weeks of pregnancy while the mother had gestational diabetes Mellitus. Serial echocardiography revealed regression of the tumor; then follow up of the patient confirmed tuberous sclerosis.

Key words

Cardiac Tumor, Fetus, Diabetes Mellitus, Tuberous Sclerosis

Introduction:

Neonate cardiac tumors are rare entities [1].Cardiac tumors are benign or malignant neoplasms arising primarily in the inner lining, muscle layer, or the surrounding pericardium of the heart. Cardiac tumors can be primary or metastatic. Primary cardiac tumors are rare in pediatric practice with a prevalence of 0.0017 to 0.28 in autopsy series. In contrast, the incidence of cardiac tumors during fetal life has been reported to be approximately 0.14 [2-3]. On the other hand, secondary malignant tumors are 10 – 20 times more prevalent than primary malignant tumors [3]. Although most cardiac tumors in pediatric patients are not malignant, they may cause blood flow obstruction, ventricular dysfunction, or arrhythmias that result in significant morbidity and mortality [4].

Rhabdomyoma is the most common cardiac tumor during fetal life and childhood. Also it is important that any intracavity mass in infants is suggestive for a cardiac rhabdomyoma unless otherwise proven. Other types of cardiac tumor are teratoma, fibroma and haemangioma. Myxoma is exceedingly rare in fetuses and neonates [3].

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Today the non-invasive diagnostic methods of these tumors are using the instrument such as echocardiography, Computing Tomography (CT) and MRI. Open surgical or endomyocardial biopsy is only utilized to reveal the histology of the lesion before surgical resection [3].

One of the differential diagnosis of Rhabdomyoma is Tuberous sclerosis complex (TSC)[5]. TSC is an autosomal dominant neurocutaneous syndrome that is characterized by formation of hamartomas in multiple organ systems distributed at various sites throughout the body, especially the brain, skin, retina, kidney, heart, and lung. It were diagnosed with dermatological manifestations (such as hypomelanotic muscles, facial angiofibromas, periungual fibromas and a shagreen patch), renal manifestations (such as angiomyolipomas (AMLs), renal cysts, renal cell carcinoma, and oncocytomas) and neurological manifestations (such as infantile spasms, seizures, intellectual disability cortical tubers, subependymal nodules and subependymal giant cell astrocytomas)[6-7]

Case Presentation:

A 25-year-old pregnant woman by gestational diabetes mellitus was checked for diagnosis of congenital heart defect at 32th weeks of pregnancy. The diabetes was going on from the fifth month of pregnancy and the mean blood sugar of mother was 160 mg/100ml at the time of pregnancy; there was no family history of diabetes mellitus. The woman used insulin for controlling the blood sugar in normal range. In her fetal echocardiography, a mass was observed in right ventricle attached to intraventricular septum right ventricular apex by pedicel. Five days after cesarean section, the infant echocardiography revealed the tumor size (19 mm length and 10 mm width) was attached to right ventricular apex (Figure 1). Abnormal ECG findings included right axis deviation, right ventricular hypertrophy, right bundle branch block, ST-segment and T wave abnormalities were consistent with ischemia and strain. There were no cardiopulmonary symptoms and signs in auscultation.

The chest X-ray appeared slightly enlargement of cardiac silhouette, right ventricale and right atrium. The cardiothoracic ratio was normal. As spontaneous regression of these tumors has been well established and there was not any homodynamic sign, surgical intervention was no longer indicated.

The infant was followed for four month twice a week. In the second echocardiography after delivery, the tumor sizes decrease to 11mm length and 6.2mm width. At the third visit on eight months old of baby, by history taking of the patient showed that he had several convulsion attacks. The baby with skin lesion, several convulsion attacks and mental retardation was referred to pediatric neurologist; then with a doubt of Tuberous sclerosis complex, CT scan was performed and it showed some subventricular and subependymal calcifications (Figure 2). Then the neurologist prescribed Prednisolone, Clobium and Liskantin tablet for TSC. Echocardiography also done and the size of tumor had decreased more and had reached to the size 8.4 mm length and 4.5 mm width (Figure 3).

Discussion:

Today with the help of the instruments such as echocardiography, CT scan and MRI to diagnose heart tumor is becoming easy. subsequently the incidence of these tumors is increasing [3]. According to the American College of Cardiology, the primary indications for fetal echocardiography, are fetal heart abnormalities or fetal arrhythmia detected by routine prenatal sonography, a family history of congenital heart disease, maternal diabetes , systemic lupus erythematosus, fetal exposure to a teratogen, fetal karyotype abnormality and other fetal system abnormalities [8-9].since the patient was diabetic,it was indicuted to perform fetal echocardiography. In echocardiography images, a mass in right ventricle of fetus heart was detected so we followed her up.

Rhabdomyoma is the most common heart tumor diagnosed among infants. It usually is regressed automatically [10]. In this case, by this thought and as no life threatening complication was seen, we did not treat the infant as the malignant tumor by the surgery and followed him to checking the tumor treat. So after the second visitation by the regression of the size of tumor our concept became more real and we need to get wait for more time to follow him up. At the third visitation by decreasing the dimensions of tumor, our concept was really confirmed.

Demonstration of cardiac rhabdomyoma by prenatal ultrasound should raise suspicion of the presence of fetal tuberous sclerosis[11]. In a study, 51 to 86 percent of cases with cardiac rhabdomyoma are associated with tuberous sclerosis. In addition, Bader et al evaluated 21 patients with rhabdomyoma and TSC is diagnosed in most them[12]. This tumor was occurred by condition of mother's gestational diabetes mellitus. The term of 'Transient asymmetric ventricular septal hypertrophy' may occur in infants of diabetic mothers as a manifestation of hypertrophic cardiomyopathy. In these cases, the hypertrophy usually regresses within a few months [13]. By this view, it can be considered that maternal diabetes mellitus probably causes benign tumors such as rhabdomyoma in neonates.

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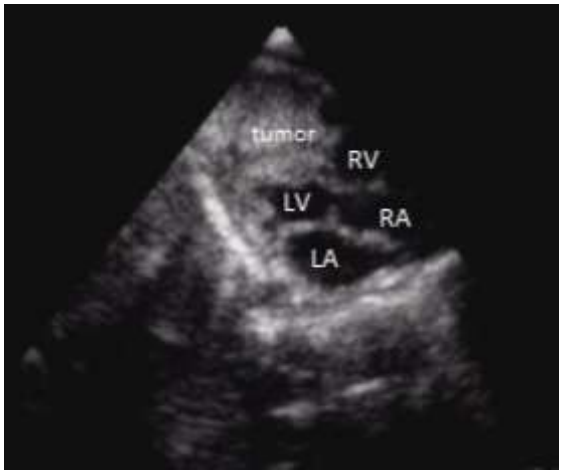

Figure 1: The first echocardiography of fetus in 32th weeks of pregnancy showed a mass in right ventricle by the size of 19mm in 10mm.

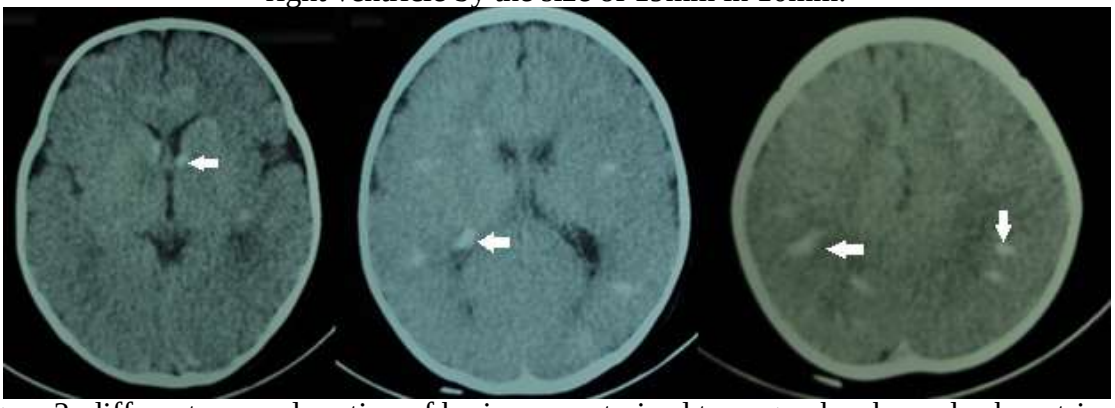

Figure 2: different coronal section of brain computerized tomography showed subventricular and subependymal calcifications.

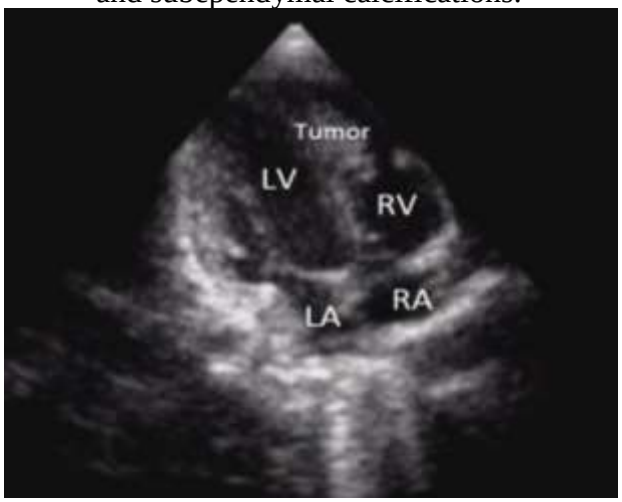

Figure 3: The echocardiography in eighth month show the decrease in size of tumor that reached to 8.4 mm length and 4.5 mm wi