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Fetal Cardiac Rhabdomyoma which Tuberous Sclerosis did not Attend: A Case Report

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Abstract

Objective: Fetuses must be evaluated prenataly for intracardiac mass. Rhabdomyoma must be recurred to the mind firstly when intracardiac mass is determined and tuberous sclerosis is invastigated because of high synergy rate.

Case: A case was preferred that one mass was determined in fetal cardiac right ventricule at her prenataly 32nd week with ultrasonography. Also two massses which were competibled with rhabdomyoma were determined in heart at echocardiography which is performed postnatally. But pathologic finding was not determined at exeminations for thubero sclerosis. Surgical and antiarrhythmic treatment were not performed because of there was not obstruction due to the massses in heart or arrhythmia. The massses regressed spontanously at third mounth of follow up.

Conclusion: Fetuses must be evaluated prenataly for intracardiac mass. Rhabdomyoma must be recurred to the mind firstly when intracardiac mass is determined and tuberous sclerosis is invastigated because of high synergy rate.

Keywords: Prenatal, rhabdomyoma, tuberous sclerosis.

Tuberoz sklerozun eşlik etmediği fetal kardiak rabdomyom: Olgu sunumu

Amaç: Prenatal dönemde fetuslar intrakardiak kitle açısından mutlaka değerlendirilmeli, saptandığında da akla ilk rabdomyom gelmelidir. Ayrıca tuberoz skleroz birlikteliği açısından da dikkatli olunmalıdır.

Olgu: Prenatal 32. haftada yapılan ultrasonografide fetal kalpte, sağ ventrikül içinde bir adet hiperekojen kitle saptanan olgu sunulmuştur. Doğum sonrası çekilen ekokardiyografisinde de kalpte iki adet rabdomyom ile uyumlu kitle saptanan olgunun tuberz oskleroz açısından yapılan tetkiklerinde herhangi bir başka patolojiye rastlanmadı. Kitleler kalp çıkışında obstrüksiyona ya da aritmiye neden olmadığı için cerrahi ve antiaritmik tedavi düşünülmedi. Yapılan ekokardiyografi takiplerinde kitlelerin üçüncü ayda gerilediği görüldü.

Sonuç: Prenatal dönemde fetuslar intrakardiak kitle açısından mutlaka değerlendirilmeli, saptandığında da akla ilk rabdomyom gelmelidir. Ayrıca tuberz oskleroz birlikteliği açısından da dikkatli olunmalıdır.

Anahtar Sözcükler: Prenatal, rabdomyom, tuberoz skleroz.

Introduction

Rhabdomyomas are the cardiac tumors most frequently met at childhood.[1] Prenatal diagnosis of rhabdomyomas can generally be established by ultrasonographical examinations after 32nd gestational week. Due to their frequent association, tuberous sclerosis should be considered in cases that were detected rhabdomyomas in heart.[2] Tuberous sclerosis is one of the neurocutaneous syndromes characterized with tumoral and non-tumoral proliferations and anomalies holding many systems especially central nervous system, skin, retina, kidney and heart. It is inherited as autosomal dominant. Cerebral cortical anomalies, subependymal tumors, seizures, mental retardation, renal angiomyolipomas and cardiac rhab-

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domyomas may be seen.[1,3] In the studies performed, cardiac rhabdomyoma is seen in 43-60% of cases with tuberous sclerosis.[2]

In this case report, a case that was diagnosed as having rhabdomyoma in prenatal period, where tuberous sclerosis was excluded in the postnatal period and rhabdomyoma regressed in the follow-up.

Case Report

A hyperechogenic mass was detected in the right ventricle of fetal heart in the ultrasonography performed in the 32nd gestational week of a 20-years-old mother whose gestational follow-up was performed regularly (Figure 1).

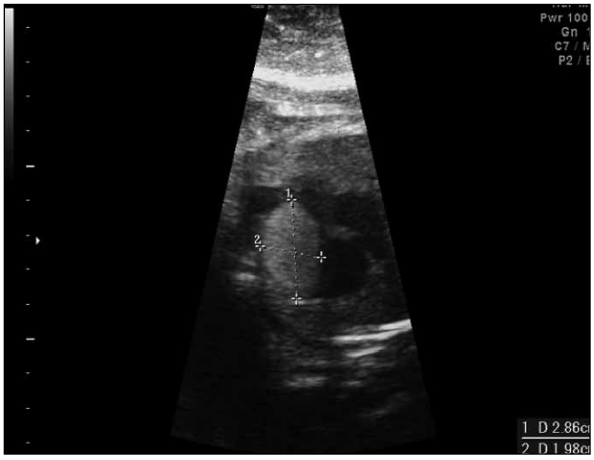

Figure 1. Intracardiac mass in prenatal ultrasonography.

The birth weight of a girl baby taken to newborn intense care unit after born by cesarean at 38th-39th week according to the last menstrual date of a twenty-years-old mother with G2P1 and the history of cesarean was 3200 gr (25-50p) while length was 49 cm (10-25p) and head circumference was 34 cm (10p). Arterial blood pressure was 72/43 mmHg and arterial oxygen saturation was 98%. No murmur and additional sound was detected in the cardiac auscultation of the cases and heart beats were rhythmic; and other system inspections were evaluated as ordinary.

In the laboratory examinations, hemoglobin was found as 17.1 g/dL, hematocrit as 47.6%, leucocyte count as 16,600/mm^3, and thrombocyte level as 334,000/mm^3. Urea, creatinine, blood electrolytes, blood glucose, AST, ALT and cardiac enzymes were at normal levels.

It was seen that the cardiac shadow of the case was normal in telecardiography, the case had sinus rhythm in electrocardiographic examination, axis was on right inferior and there were ST changes and left ventricle hypertrophy Figures 2 and 3.

In the echocardiography, two masses considered as rhabdomyomas and not causing stenosis on ventricle outlet were found as one of them was 18x11 mm in intraventricular septum within right ventricle and other one was 9 x 17 mm in lateral wall basal of right ventricle (Figure 4).

No pathology was found in the cranial tomography of the case that was planned to investigate in terms of tuberous sclerosis. Tuberous sclerosis

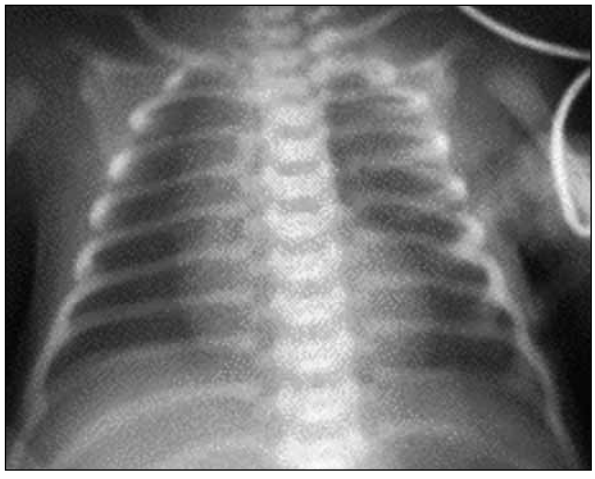

Figure 2. Telecardiography.

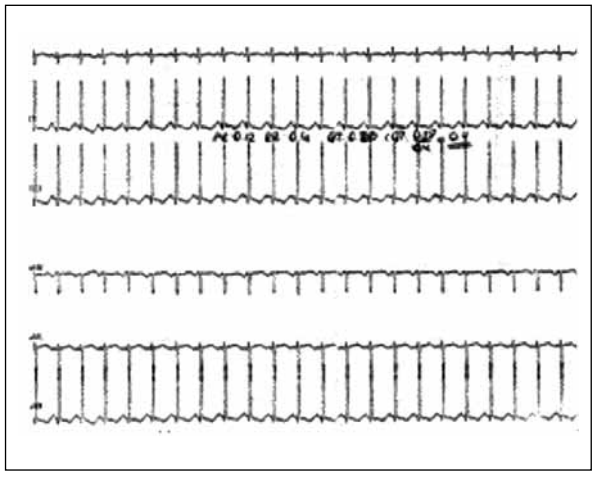

Figure 3. Electrocardiography.

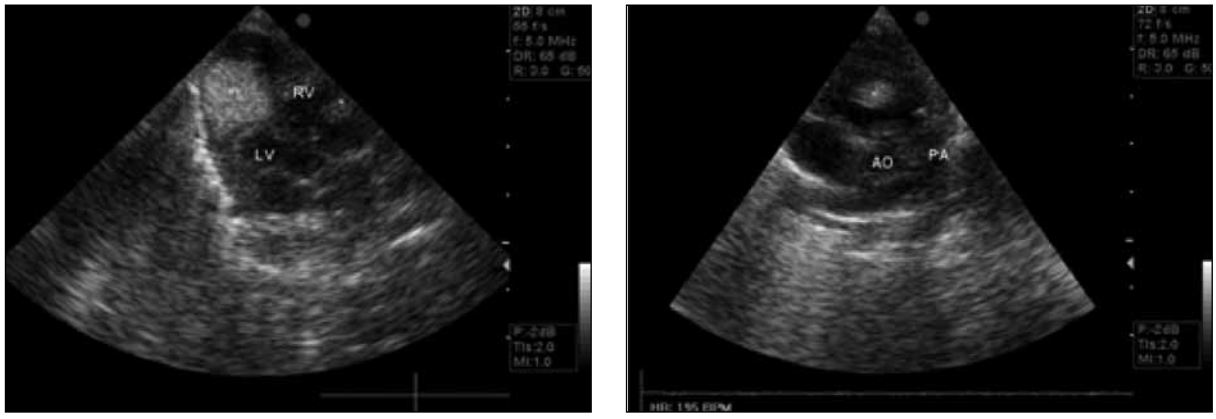

Figure 4. Two hyperechogenic masses on right ventricle in echocardiography.

was excluded in the evaluation of the case of which was found normal by urinary system ultrasonography and retinal examination performed in terms of kidney and eye involvements.

No arrhythmia occurred and heart beat rate was around 100-140/min. Since rhabdomyomas in the heart did not cause any obstruction and arrhythmia, surgical and anti- arrhythmic treatments were not considered. As cardiac rhabdomyomas could regress by themselves, the patient was followed up by echocardiography with two weeks of intervals. It was observed that masses regressed on the third month of the follow up.

Discussion

Tuberous sclerosis is a disease holding many systems by mental retardation, epilepsy, and adenoma sebaceum. It is inherited as autosomal dominant. Rhabdomyomas are the most frequent cardiac tumors of childhood; they are frequently seen together with tuberous sclerosis and are very significant for early diagnosis of tuberous sclerosis.[1,2,4]

While rhabdomyomas may settle on any part of the heart, they are generally seen in ventricles and areas close to septum.[5] They often stay as many masses together. Their clinical findings are quite variable. While they may not display any indication, they may also cause stenosis, heart failure, arrhythmia and even sudden baby deaths depending on their sizes and numbers.[5,6] In our case, masses located on right ventricle and did not display any clinical finding. The reason is that masses locate on septum and lateral wall, and are not so big.

In a multi-centered study, tuberous sclerosis was detected in 10 of 19 fetuses with cardiac tumor (52.6%), and it was seen that lesions were more than one in 9 of these 10 fetuses (90%).[7] In another study, tuberous sclerosis was detected later on in 51 of 85 fetuses with rhabdomyoma (59.3%).[8] In our case, masses more than one were detected, but there was no tuberous sclerosis.

As rhabdomyomas can regress in time, it is suggested to follow them by echocardiography. Surgical treatment is recommended when they cause mechanical stenosis in heart and life-threatening arrhythmias.[6] Surgical intervention was not considered since rhabdomyoma detected in our case did not cause hemodynamic disturbance and arrhythmia; therefore the case was followed up by echocardiography and it was observed that it regressed completely on third month.

Fetuses should certainly be investigated in prenatal period in terms of intracardiac mass and when detected, rhabdomyomas should be considered first. Patients should be evaluated by echocardiography in postnatal period and it should not be forgotten that it may cause mechanical stenosis in heart, obstruction and arrhythmia and may cause serious life-threatening results. It should also be paid attention for the association with tuberous sclerosis and other systems certainly should be checked.

Conclusion

Fetuses should certainly be investigated in prenatal period in terms of intracardiac mass and when detected, rhabdomyomas should be consid-

ered first. Patients should be evaluated by echocardiography in postnatal period and it should not be forgotten that it may cause mechanical stenosis in heart, obstruction and arrhythmia and may cause serious life-threatening results. It should also be paid attention for the association with tuberous sclerosis and other systems certainly should be checked.

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