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Case Report

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Long QT syndrome diagnosed by premature atrial extrasystoles: a case report

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Abstract

Objective: We aimed to present a case that was prenatally diagnosed as fetal arrhythmia due to premature atrial extrasystoles but turned out to be long QT syndrome at postnatal period.

Case: A 27-year-old primigravid woman was referred to our clinic at 29 weeks of gestation due to marked fetal arrhythmia. Premature atrial extrasystoles were detected during the prenatal period. The QT interval was 500 msec in the postnatal period. So the case was diagnosed as long QT syndrome associated with premature atrial extrasystoles.

Conclusion: Premature atrial extrasystoles are easily recognized and generally presented as an isolated rhythm disorder. But the minority of cases is associated with serious arrhythmia such as long QT syndrome. This lethal condition should be considered in the differential diagnosis to predict the potential risks for the fetus and neonate.

Keywords: Fetal arrhythmia, long QT syndrome, premature atrial extrasystole.

Özet: Prematür atriyal ekstrasistoller ile tanı alan uzun QT sendromu: olgu sunumu

Amaç: Prematür atriyal ekstrasistoller nedeniyle perinatal fetal aritmi olarak tanı alan ancak postnatal dönemde uzun QT sendromu olduğu görülen bir olguyu sunmayı amaçladık.

Olgu: Yirmi yedi yaşındaki primigravid olgu, dikkat çekici fetal aritmi nedeniyle 29. gebelik haftasında kliniğimize başvurdu. Prenatal dönemde prematür atriyal ekstrasistoller tespit edildi. Postnatal dönemde QT aralığı 500 ms idi. Bu nedenle olgu, prematür atriyal ekstrasistoller ile ilişkili uzun QT sendromu tanısı aldı.

Sonuç: Prematür atriyal ekstrasistoller kolay tespit edilmektedir ve genel olarak izole ritim bozukluğu şeklinde ortaya çıkmaktadır. Ancak olguların çok azında, uzun QT sendromu gibi ciddi aritmi görülmektedir. Bu ölümcül durum, fetüs ve yenidoğan için potansiyel riskleri öngörmek amacıyla ayırıcı tanıda dikkate alınmalıdır.

Anahtar sözcükler: Fetal aritmi, uzun QT sendromu, prematür atriyal ekstrasistol.

Introduction

Long QT syndrome (LQTS) is an inherited syndrome with an autosomal transmission.[1] Prolonged QT interval on electrocardiogram (ECG), family history, symptoms of arrhythmia were used to as diagnostic criteria. LQTS is defined in neonates,[2] but there are few cases reported for intrauterine period.[3,4] Prenatal diagnosis of LQTS is rare, particularly in the lack of family history.[5] We described a case of sporadic LQTS who presented with premature atrial extrasystoles during antenatal period.

Case Report

27-year-old primigravid woman at 29 weeks of gestation was referred to our clinic when fetal arrhythmia was detected with ultrasound. Average fetal heart rate was determined as 110 beats per minute (bpm). M-mode and pulsed Doppler recordings of left ventricular in- and outflow tracts, including mitral and aortic valve movements, were used to assess of the fetal arrhythmia. On M-mode ultrasound, one of both atrial contractions was evaluated as premature atrial extrasystole. Each atrial contraction was followed by a ventricular contraction.

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These findings were evaluated as conduction atrial bigeminy (**Fig. 1**). There were no evidence of congestive heart failure and structural abnormalities. The parents' medical history was clear of any arrhythmic syndrome and their physical examination was normal. Parental QT intervals on ECG were assessed to check the possibility of the LQTS and they did not have long QT intervals (<480 msec). No additional fetal anomalies were found in the genetic sonogram. Starting from the diagnosis at 29th week the patient was followed by weekly fetal echocardiography. During the antenatal period premature atrial extrasystoles were detected without other rhythm disorders and congestive heart failure. Cesarean section was performed at 38 weeks and 2 days because of the prior cesarean section history and fetal growth restriction. A 2540 g male neonate was delivered. Echocardiography was performed after birth. A very long QT interval of 500 msec was found on ECG in the neonate (**Fig. 2**). As a result, the neonate was diagnosed as LQTS and followed-up due to risk of sudden infant death. He remained asymptomatic for 4 months. No medical treatment was required during postnatal period. The parents rejected any offers of genetic evaluations.

Discussion

Arrhythmias are detected in at least 2% of all pregnancies. The fetal heart should be examined carefully in the antenatal period.[6] In our case, we described a fetus presented with premature atrial extrasystoles in utero and in whom LQTS was diagnosed in the postnatal period. Congenital LQTS is a heritable ion channel disorder that is associated with the impairment of number of genes encoding for the transmembrane sodium or potassium ion channel proteins. Mutations in six subtypes (LQT1 to LQT6) are responsible of the impairment of the ion channels leading to prolonged action potentials that are account for congenital long QT.

These mutations slow the inactivation of inward depolarizing sodium currents or cause retardation of outward repolarizing potassium currents.[7] Long QT interval was diagnosed by using fetal magnetocardiography[8] and fetal ECG in prenatal period.[9] Among all cases of sudden infant death syndrome, 50% underlying risk factor is LQTS.[5] The prognosis of LQTS is not good when detected in the prenatal period or during the first week of life.[10] Fetal diagnosis of a prolonged QT has been recognized as early as 16 weeks of gestation.[11] In a systematic review, 21 fetuses with LQTS were doc-

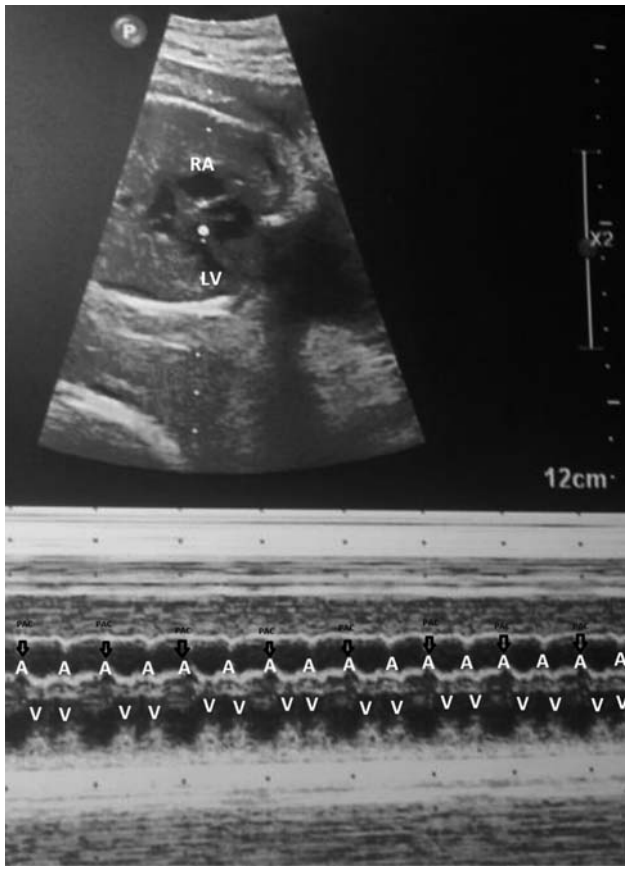

Fig. 1. M-mode recording of the fetus with conduction atrial bigeminy. The M-mode cursor line intersects the right atrium (**RA**) and left ventricle (**LV**). Every other atrial contraction is early; the premature contractions are always followed by a ventricular contraction.

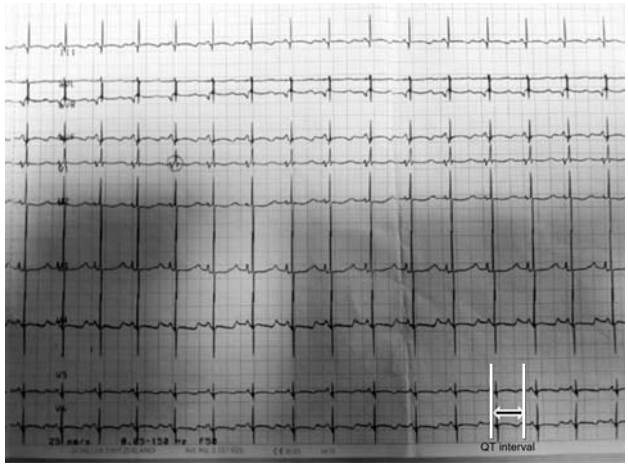

Fig. 2. A very long QT interval of 500 ms was found in the newborn at the electrocardiography. The QT interval is from the beginning of the Q wave to the end of the T wave. The expected QTc interval should be less than 480 msec among women.

umented with significant in utero cardiac findings. 16 fetuses (76%) exhibited bradycardia ≤110 bpm; 4 of them (19%) exhibited ventricular tachycardia or tachyarrhythmia, and one case exhibited pleural effusion. Eleven fetuses (52%) exhibited atrioventricular block (AVB) in prenatal and postnatal period. 4 fetuses (19%) exhibited mild bradycardia ranging from 100 to 110 bpm and reduced baseline fetal heart rate (FHR) variability on cardiotocography. So, as result of AVB sinus bradycardia and fetal bradycardia can be seen fetuses with LQTS.[10] Cuneo et al. detected isolated extrasystoles and AVB in 97.4% and 2.6% of the fetuses, respectively.[11] We detected isolated premature atrial extrasystoles in our case, too. Some fetuses with LQTS show mildly decreased baseline (FHR) of 110–120 bpm. Suspicion of LQTS in such fetuses with a baseline FHR of 110–120 bpm may increase the proportion of patients with prenatally diagnosed LQTS.[10] Furthermore, some patients with LQTS were presented with a FHR of more than 120 bpm in utero.[12] Also, some of them exhibit reduced heart rate variability.[13] In our case, the average FHR was calculated as 110 bpm with normal heart rate variability. This case is not a case of LQTS diagnosed in antenatal period. Diagnosis in the postnatal period is due to the suspicion of arrhythmia detected during antenatal period. In fetuses with fetal arrhythmia LQTS should be considered in the differential diagnosis, particularly in cases where there is no family history. Because LQTS is a genetic disorder, in cases where there is no family history of arrhythmia and syncope it may not be considered in differential diagnosis. With this case, we tried to emphasize the necessity of conducting antenatal and postnatal investigations considering the possibility of sporadic cases in pregnant women who have applied with fetal arrhythmia. Thinking of long QT syndrome as a differential diagnosis in fetal arrhythmia will help to notice the possible complications that may develop in neonate.

As a result, detection of a fetal arrhythmia is not adequate for prenatal follow-up; it is also necessary to identify the type of arrhythmia. The underlying pathologies such as the LQTS that may accompany arrhythmias should always be investigated. Especially, in sporadic cases with LQTS, prenatal diagnosis is important for both the fetus and the neonate. LQTS can be diagnosed with ECGs and genetic tests during prenatal investigations. So, the incidence of "sudden infant death syndrome" can be decreased with the help of these investigations.[10]

Conclusion

The fetal echocardiography does not detect the QT interval. So, we should think about possibility of LQTS in fetal arrhythmias.

Conflicts of Interest: No conflicts declared.

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