Fetal tachycardia: an unusual cause of maternal mirror syndrome

Emre Erdogan, Resul Arsoy, Oya Demirci, Oya Pekin, Semih Tugrul, Pinar Kumru
Zeynep Kamil Research Hospital, Istanbul, Turkey

Introduction:
Mirror syndrome refers to a condition of generalized maternal edema, often with pulmonary involvement, that mirrors the edema of the hydropic fetus and placenta. Because condition is rare and frequently misdiagnosed as preeclampsia, the exact incidence is still unknown. We reported a case of maternal mirror syndrome caused by fetal tachycardia that progressed to fetal hydrops with the interesting feature that there was a rapid improvement in maternal status after fetal therapy.

• Presentation of case:
A 33-year-old woman, gravidia 3, para 2, group A, rhqes negative was referred at 27 weeks gestation to our department because of hydrops fetalis. Fetal ultrasonographic examination revealed massive ascites, right hydrothorax, polyhydramnios and placentaemagly(Figure-3). Fetal arrhythmia was suspected due to abnormal heart rate during fetal cardiac examination. Fetal echocardiography presented mild cardiomegaly, severe tricuspid regurgitation with normal cardiac anatomy(Figure-2). Pulsed wave doppler demonstrated short runs of tachycardia at 300-350 bpm(Figure-3). Although the supraventricular tachycardia was of short duration, due to the elevated rate and fetal hydrops in utero therapy via maternal administration of antiarrhythmic drugs was planned. Digoxin 0.5mg, 0.25mg and 0.25mg initiated over the first 24 hours. The digoxin levels were 1 to 1.2ng/ml. Maintenance dose of 0.5mg per day was initiated for 48 hours with the digoxin levels 1-2ng/ml. But because of sustained tachycardia and fetal hydrops, digoxin was continued and sotalol 80mg three times was initiated. Tachycardia transiently terminated at 96 hours. Ventricular fuction was in sinus rhythm with the improvement of the fetal hydrops within one weeks. Complete resolution of the fetal hydrops occured three weeks after treatment(Figure-4).

The patient presented significant vulvar and pretrial edema on the third day of treatment. At clinical examination blood pressure was 140/90mmHg and no value above 140/90 was detected. The laboratory findings showed decrement of hematocrit level from 35% to 28%, albumin 3,2gr/dl to 2,6 gr/dl and platelets 132000 to 119000. The 24-hour protein collection was 199mg/dl. As the patient had severe headache she was followed in our intensive care unit with an initial diagnosis of preeclampsia. Magnesium sulphate prophylaxis was continued for 24 hours. Over the following days maternal and fetal condition showed resolution of the maternal edema and fetal hydrops. Improvement in maternal clinical picture and fetal hydrops favoured mirror syndrome.

As the maternal symptoms resolved and no maternal side effect of the drug was detected; fetal therapy was continued until 37 weeks of gestation. Cesarean section was performed at 37 weeks due to rupture of membranes and bradycardia presentation. Postnatal evaluation of the baby revealed no echocardiographic and electrocardiographic abnormality.

Figure 1: Ascites at 27 weeks of gestation
Figure 2: Fetal echocardiography presenting mild cardiomegaly with normal cardiac anatomy
Figure 3: Pulsed wave doppler demonstrating short runs of tachycardia at 300-350 bpm
Figure 4: Complete resolution of the fetal hydrops at 30 weeks of gestation

• Discussion:
Maternal mirror syndrome is a rare condition characterized by a combination of fetal hydrops and maternal fluid retention which ‘mirrors’ fetal hydropic changes. John W. Ballantyne first described the association of maternal edema in pregnancy with fetal and palpebral hydrops due to rhues isomunization[1].

The pathogenesis of mirror syndrome is unknown. There are several hypothesis. Redman et al reported that that the inflammatory response as a result of increased shedding of trophoblastic debris into maternal blood[2]. Stephan et al have postulated that the hydropic placenta may increase production of soluble fms-like tyrosine kinase (sflt1), which is an important mediator of maternal inflammatory response in pre-eclampsia. Moreover, Urbina et al reported a case of maternal mirror syndrome caused by bilateral fetal hydrothorax that the antiangiogenic factor similar to that seen in preeklampia resolved after intrauterine pleuroamniotic shunt placement(4). Similarly increases in sflt1 has been described in cases of mirror syndrome associated with parvovirus infection, Rh-isomunization, cytomegalovirus infection and twin–twin transfusion syndrome[5].

We present a case of fetal tachycardia is an unusual example of maternal mirror syndrome with significant vulvar edema. In this case the patient presented preeclampsia-like symptoms with severe edema, insistent anemia, decreased platelets and transient mild hypertension. Mirror syndrome may present with rapid weight gain, peripheral edema and progressive shortness of breath. Edema is always a key feature, albuminuria usually mild and preeclampsia unusual. The problem of distinguishing between mirror syndrome and preeclampsia is obvious. In contrast to preeclampsia, the maternal hematocrit is often low (hemodilution) rather than high (hemoconcentration), amniotic fluid volume is often high (polyhydramnios) rather than low (oligohydramnios), and the fetus always shows signs of hydrops. The etiology of the mirror syndrome includes a wide variety of fetal causes. In the review of 56 cases published by Braun et al; severe rhuses immunisation(29%) was the most of the cases associated with the mirror syndrome[6]. Mirror syndrome caused by fetal supraventricular tachycardia is a very rare condition. There is only one case report similar to our case that showed the resolution of mirror syndrome caused by fetal supraventricular tachycardia with maternal flecainide administration. Maternal reconvalescence was reported one week after flecainide treatment and resolution of the fetal supraventricular tachycardia terminated in our case, while maternal edema resolved and the laboratory findings came to normal ranges on the sixth day of the treatment. We ruled out that the maternal symptoms appearing 3 days after Digoxin treatment were not due to side effects of the drug we continued Digoxin until delivery.

The key is in this syndrome is to recognize and identify a treatable cause which can lead to the reversal of the syndrome and continue the pregnancy. Adequate treatment of fetal tachycardia in fetuses with hydrops resulted in improvement in fetal hydrops. Individual series report that cardioversion is successful in 65-85% of fetuses within one week of initiating therapy. But treatment of hydrops depends on the cause. When the fetal conditions are not treatable, continuing the pregnancy worsens the maternal symptoms. Delivery or termination of the pregnancy is usually required to induce remission of maternal symptoms, which can be life-threatening. Also spontaneous resolution of mirror syndrome has been described after spontaneous resolution of fetal hydrops related to parvovirus infection and after fetal death.

Finally, this case of fetal supraventricular tachycardia and mirror syndrome demonstrates how fetal symptoms of cardiac failure, fetal hydrops can be mirrored into maternal symptoms and the treatment improves fetal hydrops and mirror syndrome. Because of increased maternal morbidity, the clinicians should be alerted about mirror syndrome in the existence of maternal edema and hemodilution. Future studies are needed on the mechanism of the disease and lead to both therapeutic and preventive strategies.

• References: