

Mirror Syndrome Associated with Fetal Cardiac Anomaly: A Case Report

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ABSTRACT

A rare mirror syndrome (Ballantyne syndrome) was seen in a woman who carried a hydropic fetus caused by fetal atrio-ventricular septal defect (AVSD). Diagnosis was made with confidence after ruling out cardiogenic pulmonary edema and preeclampsia. Placental pathology demonstrated multifocal villous edema and accelerated maturation of trophoblasts which may support the earlier reports about potential etiologic roles of the placenta to trigger the disease.

Keywords: AVSD, Ballantyne syndrome, fetal hydrops, mirror syndrome

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Mirror syndrome is characterized by a combination of fetal hydrops and maternal fluid retention which “mirrors” fetal hydropic changes. The syndrome was first described by John William Ballantyne in 1892. To date, 32 cases have been reported with various fetal and placental causes.¹⁻¹⁰ The exact incidence is still unknown and the precise number of cases could be higher because the condition is frequently mistaken for preeclampsia. This syndrome often results in significant perinatal morbidity and mortality. However, the course of disease can be reversed when fetal conditions are treated and result in good perinatal outcomes as described in many reports.^{3-5,9-13}

CASE REPORT

A 21-year-old Thai woman in her first pregnancy was seen for antenatal care at 15 weeks of gestation. History and physical examination and basic laboratory results were unremarkable. At 21 weeks of gestation routine obstetric ultrasonography was performed and showed an abnormally large right atrium of the fetus, with otherwise unremarkable fetal structures. Detailed fetal echocardiography was then performed and the diagnosis was atrio-ventricular septal defect (AVSD) with severe tricuspid regurgitation with moderate mitral regurgitation. Amniocentesis for fetal chromosomal study revealed 46, XY. Digitalis (0.125 mg twice a day) was prescribed to the patient.

At 25 weeks of gestation she presented with both hands and feet swelling. She had gained 10 kgs. of weight during the last week. During her evaluation, she was found to have 3+ peripheral edema, mild degree of puffy eyelids, shortness of breath with 89% oxygen saturation from room air inhalation, and blurred vision. On examination, her blood pressure was 150/90 mmHg, which had risen from one week before (120/80), and fine crepitation at both lower lung fields were detected. The laboratory findings showed decrement of hematocrit level from 41% to 28%. All preeclampsia blood tests were within normal limits. The 24-hour urine protein collection was 290 mg/dl. The blood level of digitalis was in the therapeutic range.

Chest X-ray showed bilateral pulmonary congestion with minimal bilateral pleural effusion and mild cardiomegaly. The level of plasma brain natriuretic peptide was normal. Accompanied with normal maternal echocardiographic study, a cardiogenic cause of pulmonary edema was ruled out.

After admission, her blood pressure dropped down to normal range and magnesium sulphate was not given because of pulmonary edema and the maternal clinical pictures that favoured mirror syndrome more than preeclampsia. Intravenous furosemide and 5 L/min oxygen therapy via a nasal canula were provided to the patient. Post-therapeutic oxygen saturation was 95%.

Ultrasonography revealed a living fetus with scalp edema without ascites. The appearance of severe tricuspid regurgitation and cardiomegaly were the same as the previous study. Polyhydramnios and thick placenta were also noted.

Detailed discussion with the parents regarding the poor prognosis of the fetus and the potential deterioration

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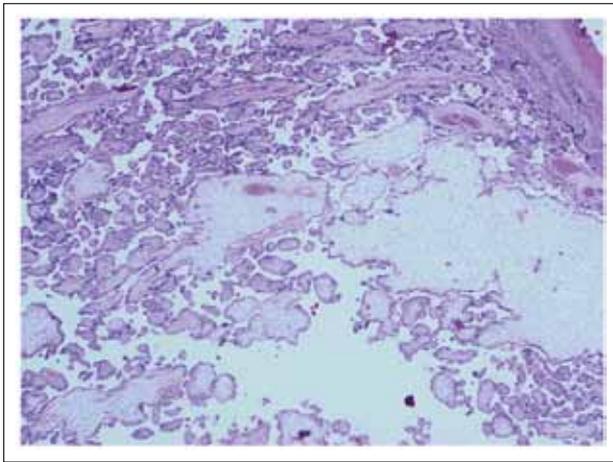


Fig 1. Microscopic examination of the placenta showed multifocal villous edema among villi with accelerated maturation and stromal congestion and hemorrhage.

of maternal symptoms led to the decision to terminate the pregnancy. Induction of abortion was performed by vaginal misoprostal suppository. A 1020-gm stillborn hydropic fetus with a large placenta weighing 550 gm were delivered.

The patient recovered uneventfully post abortion. At 1-week follow up she was doing well, the peripheral edema had disappeared completely and her blood pressure was normal.

Pathological examination of the fetus and placenta revealed minimal ascites and pericardial effusion, dysplasia of tricuspid valve, atrial septal defect and multifocal villous edema and accelerated maturation. (Fig 1)

DISCUSSION

The incidence of AVSD is about 15% of all cardiac malformations and in severe cases can lead to fetal cardiac failure and hydrops fetalis.¹⁴ The incidence of mirror syndrome is unknown owing to the rarity of the condition and the criteria for diagnosis has not been established due to the small number of the cases and variation of presenting symptoms. Vidaeff et al¹ reviewed the literature from 1956 - 2002 and found 20 cases of mirror syndrome with various underlying causes including fetal cardiac arrhythmias, Ebstein's anomaly, placental chorioangioma, aneurysm of the vein of Galen, α -thalassemia, sacrococcygeal teratoma, trisomy 13, erythroblastosis and parvovirus B19 infection. From 2002, 13 more cases have been added to the literature including ours. Our patient shared several features previously described in many reports and the diagnosis of mirror syndrome was made with confidence. The common presentations include peripheral edema, rapid weight gain, hydramnios, elevated maternal plasma uric acid level, mild proteinuria, raised blood pressure and hemodilution. The hemodilution was considered by many authors as an important distinguishing characteristic between mirror syndrome and preeclampsia.

Although the precise pathophysiologic mechanisms of mirror syndrome remain unclear, there are many proposed theories in the effort to disclose the underlying mechanisms which cause the disease. Hypothyroidism⁷, soluble vascular endothelial growth factor receptor-1 (sVEGFR-1)⁶ and vasopressin¹⁵ are among the possible factors that may trigger the maternal edema. Brochot et al⁵ have speculated that the development of mirror syndrome may be due to

circulation in the maternal blood of "unknown" messengers of placental origin. These messengers, which appear to be toxic to the vascular endothelium, may be released by a mechanism involving relative placental hypoxia in major placental edema. Placenta as an etiology gains more attentions since there are reports about the syndrome which developed in cases with placental hydrops and even after resolution of fetal hydrops, but still with thick placenta.^{1,2} Our patient, with thick placenta, but without a classical features of fetal hydrops, may support these authors about the role of the placenta in the pathogenesis of the syndrome.

Digitalis was given to the mother with the hope that it could help to improve the myomacardial contractility of the fetus and the mirror syndrome of the mother could also possibly be improved. Unfortunately, the cardiac defects of the fetus in our case were beyond treatment and the fetus continued to show signs of congestive heart failure.

The key element in this syndrome is to recognize and identify a treatable cause which can lead to the reversal of the syndrome and pregnancy can continue to term or to terminate the pregnancy when the fetal conditions are not treatable and the prognosis is really bad in order to limit the risk of severe maternal complications. Future studies may disclose the mechanisms of the disease and lead to therapeutic and preventive strategies.

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