Atrial fibrillation with rapid ventricular response as a maternal presentation of mirror syndrome: a case report

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ABSTRACT

Mirror syndrome is the phenomenon of fetal hydrops causing maternal edema and weight gain. Here, we report a case of arrhythmia as the primary maternal symptom.

A 36-year-old woman, G2P1001, at 34.5 weeks of gestation presented with new-onset fetal hydrops combined with maternal weight gain and edema. She developed atrial fibrillation with rapid ventricular response; cardiac workup was unremarkable. Rate control was achieved with diltiazem. She underwent delivery and reverted to normal sinus rhythm on post-operative day 1.

Volume overload causes atrial wall stress and neurohormonal changes that may trigger atrial fibrillation. Optimization with rate control facilitated good maternal and fetal outcomes in this case.

1. Introduction

The development of arrhythmia in pregnancy is rare, particularly among women without underlying structural heart disease [1,2]. While maternal arrhythmias are infrequent, their timely identification and evaluation for reversible etiologies is important as they can cause cardiovascular dysfunction and result in adverse maternal and fetal outcomes [1,3]. We present here what we believe to be the first report of a case of maternal arrhythmia as a presenting symptom of mirror syndrome in the setting of non-immune fetal hydrops. This report adheres to the applicable EQUATOR guideline.

2. Case Presentation

A 36-year-old woman, G2P1001, with no significant past medical history, at 34 and 5/7 weeks of gestation presented to hospital for evaluation following minor abdominal trauma. An incidental finding was of new polyhydramnios and fetal hydrops with fetal ascites, pleural effusion and pericardial effusion.

She had conceived via in-vitro fertilization (IVF) with a preimplantation genetically tested embryo. She received routine prenatal care; the results of cell-free DNA screening were risk-reducing. At her 34-week prenatal visit, she had gained 10 pounds since her 32-week visit and fundal height was noted to be greater than expected for her gestational age; an ultrasound scan was planned to evaluate fetal growth. When she presented to the hospital 4 days after that prenatal visit following minor abdominal trauma, an unscheduled ultrasound examination showed evidence of polyhydramnios and fetal hydrops. She was therefore admitted for monitoring and further evaluation. On admission she was noted to have a 20-pound weight gain since her last clinic visit and new bilateral lower extremity edema. She reported that her 3-year-old daughter had experienced a flu-like illness within the past three weeks. She received a course of betamethasone for prematurity. Laboratory studies demonstrated no evidence of immune hydrops. Evaluation for etiologies of fetal hydrops were notable for a negative blood antibody screen and for positive cytomegalovirus (CMV) IgG and IgM in her blood.

On hospital day 2, she developed new-onset palpitations and worsening lower-extremity edema. An electrocardiogram (ECG) demonstrated atrial fibrillation (AF) with rapid ventricular response with a
maternal heart rate in the 150 s. She was initiated on metoprolol for rate control with inadequate response and was transitioned to a diltiazem infusion. Laboratory studies were unremarkable except for a B-type natriuretic peptide level of 443 pg/mL (upper limit of normal in pregnancy 50 pg/mL). While a chest x-ray did not indicate pulmonary edema, an echocardiogram demonstrated intravascular overload without functional or structural abnormalities. A computed tomography angiogram did not reveal evidence of a pulmonary embolism. The patient remained normotensive without proteinuria. The diagnosis was felt to be most consistent with mirror syndrome in the setting of fetal hydrops.

Given the concern for mirror syndrome and the risk of worsening of maternal health, as well as the risk of stillbirth with fetal hydrops, delivery was recommended and the patient underwent an uncomplicated repeat cesarean section as she did not desire a trial of labor. She remained in rate-controlled AF during the cesarean delivery and in the immediate postoperative period despite preoperative initiation of a diltiazem drip. She was transitioned to oral metoprolol on the morning of postoperative day 1. She converted to normal sinus rhythm (NSR) that evening and remained in NSR for the remainder of her hospital stay. She was discharged home on postoperative day 4 on oral metoprolol succinate with the plan for an extended Holter monitoring of her cardiac rhythm in the immediate postpartum period. After 13 days the Holter monitoring showed no further episodes of AF but isolated (<1%) supraventricular and ventricular ectopic events.

Immediately after delivery the neonate was intubated. Apgar scores at delivery were 3, 5, and 8, at 1, 5 and 10 min respectively. Venous cord blood pH was 7.35; arterial cord blood could not be collected. Infant birth weight was 3130 g (95th percentile). Fetal pleural effusions were managed with needle decompression followed by chest tube placement. The chest tube drained chylous fluid, consistent with a chylous leak. Urine serotonin testing of the infant was negative for CMV. Genetic testing with microarray was normal. Following extubation on day of life 11, the chest tubes were removed, and the infant was discharged home on day of life 40.

3. Discussion

The development of new-onset cardiac arrhythmia in pregnancy is rare, especially among women without underlying health conditions [1]. A nationwide retrospective study found that the rate of any arrhythmia was 68 per 100,000 pregnancy-related hospitalizations and that atrial fibrillation was present in just 27 per 100,000 pregnancy-related hospitalizations [4]. However, arrhythmia in pregnancy was associated with increased frequency of hospital death and greater rates of maternal and fetal complications. New-onset AF in pregnancy is a particularly rare event, as most women with AF in pregnancy have their diagnosis of AF prior to pregnancy [1,5]. Women with underlying heart disease who develop AF in pregnancy are at increased risk of developing congestive heart failure (CHF) and preeclampsia compared to women with structurally normal hearts [1,4].

Evaluation for the underlying etiology of new-onset AF should include ECG, electrolyte studies, urine toxicology, thyroid function testing, and echocardiogram to rule out underlying structural and physiologic conditions [5,6]. Pulmonary embolism, while rare, can also present with AF, and should be considered in patients with concomitant dyspnea, low oxygen saturation levels or in patients without another clear etiology [6]. The most common etiologies of AF in pregnancy are structural heart disease and hyperthyroidism [2].

Although there are a few reported cases of lone AF (i.e., AF without an identifiable cause) in pregnancy [7], in this patient with new-onset non-immune fetal hydrops secondary to a fetal chylous leak and significant maternal edema, maternal AF is hypothesized to have been secondary to the hypervolemia of mirror syndrome. The conversion of AF into normal sinus rhythm following delivery despite preoperative need for antiarrhythmic agents further supports mirror syndrome as the unifying diagnosis, as symptoms typically resolve with delivery.

Mirror syndrome is a rare but likely underdiagnosed complication of fetal hydrops marked by maternal edema (90%), hypertension (60%) and proteinuria (40%) [8]. It was first described by John W. Ballantyne in 1892 in the setting of fetal hydrops due to Rhesus isoimmunization [9]. Since then, cases of mirror syndrome have also been described in association with non-immune fetal hydrops (NIFH) secondary to human parvovirus infection and Ebstein’s anomaly, among other causes [8,9]. Symptoms typically resolve shortly after delivery [8]; however, approximately 20% of women will experience severe complications such as pulmonary edema and peripartum cardiomyopathy [9]. While the pathophysiology is not well established, mirror syndrome is likely related to aberrant placental function, sharing a common underlying pathway with preeclampsia [8,9].

Common states of hypervolemia such as decompensated cirrhosis, heart failure, and hypertension are associated with the development of AF [10–12]. Recent studies in animal models have demonstrated that elevated hydrostatic pressures induce upregulation of inflammatory cytokines such as TNF-alpha and macrophage migration inhibitory factor which contribute to remodeling of atrial electrical pathways [12]. It is thus plausible that the significant edema of mirror syndrome and subsequent volume overload could lead to increased atrial wall stress and subsequent AF. This is further supported by studies that have found an increased rate of AF in the third trimester of pregnancy, a time of significant hemodynamic changes, including physiologic increases in plasma volume [1,13].

Management of AF depends on the stability of the patient and the underlying etiology. In patients who are hemodynamically stable, the first goal is rate control, which may be achieved with a beta-blocker, calcium channel blocker or digoxin [3,5]. Once rate controlled, the majority of pregnant patients with hemodynamically stable AF spontaneously convert to normal sinus rhythm. However, persistent AF is an indication for cardioversion (either pharmacologic or electric) as it can lead to thrombus formation [3,5]. In this case, anti-coagulation was deferred antepartum given the decision to proceed with urgent delivery via cesarean section for maternal and fetal benefit and was not required postpartum given spontaneous conversion to normal sinus rhythm.

This case demonstrates maternal arrhythmias as a presenting symptom of mirror syndrome in the setting of recently diagnosed nonimmune fetal hydrops. The development of new-onset cardiac arrhythmias in pregnancy is rare, but thorough evaluation of both the mother and fetus may provide evidence of a reversible etiology. In women presenting with new-onset arrhythmias in pregnancy along with signs and symptoms of edema, evaluation of the fetus for hydrops that may be “mirrored” in the mother is warranted. Conversely, when a diagnosis of fetal hydrops is made, maternal arrhythmia is another potential complication to be considered.

Contributors

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Elena HogenEsch drafted the article and contributed to editing and revision.
Carly Dahl provided clinical care for the patient and contributed to editing and revision of the article.
Alexander Samworth contributed to editing and revision of the article.
Seema Venkatachalam provided clinical care for the patient and contributed to editing and revision of the article.
Priya Rajan provided clinical care for the patient and revised the article critically for important intellectual content.
Elizabeth M. S. Lange provided clinical care for the patient and contributed to editing and revision of the article.

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Dr. Priya Rajan consults for Gerson Lehrman Group and has received honorariums for lectures given to Texas Tech University and the National Association of Perinatal Social Workers.

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