Medicine

Undiagnosed mirror syndrome with maternal hypoxemia onset during an emergency cesarean section

A case report

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Abstract

Rationale: Mirror syndrome is a rare pregnancy condition in which maternal edema is associated with fetal hydrops. Because of its rarity and overlapping symptoms, this condition is often misdiagnosed as another pregnancy complication.

Patient concerns: A 28-year-old pregnant Japanese woman presented with sudden 7.5-kg weight gain, leg edema, and increased d-dimer level.

Diagnoses: Ultrasound revealed polyhydramnios and fetal hydrops, and findings of maternal edema and blood test results were suggestive of Miller syndrome. Although, the patient was initially misdiagnosed due to a lack of information and the rarity of this disease.

Interventions: An emergency cesarean section was performed under spinal anesthesia at 36 weeks and 2 days of-pregnancy. We could not diagnose mirror syndrome.

Outcomes: The newborn's Apgar scores were 2 and 5 at 1 and 5 minutes after delivery, respectively. The patient's SpO_2 suddenly decreased to 86% during cesarean section and persisted for 2 days. Chest computed tomography revealed pleural effusion and pulmonary edema. The pleural effusions and lung edema spontaneously resolved after the cesarean section.

Lessons: This case reports on Miller syndrome with maternal hypoxemia onset during an emergency cesarean section and highlights the potential for better perioperative management and improvement in maternal mortality through prompt diagnosis and appropriate treatment shared not only among obstetricians and pediatricians but also among anesthesiologists.

Abbreviation: NIFH = nonimmune fetal hydrops.

Keywords: fetal hydrops, hypoxemia, maternal edema, mirror syndrome

1. Introduction

Mirror syndrome is a rare maternal pathology that "mirrors" fetal pathology and has a significant impact on fetal and mater-nal mortalities. Although the diagnostic criteria of mirror syndrome have not been clearly defined,[1] the syndrome is char-acterized by 3 complications: fetal hydrops, placental edema, and maternal edema. Most cases of mirror syndrome are com-plicated by hypertension, and it is important to differentiate hypertension in mirror syndrome from hypertensive disorders during pregnancy.[1,2] Generally, the causes of polyhydramnios and fetal hydrops include fetal (morphological and chromo-somal abnormalities), maternal (impaired glucose tolerance),

Written informed consent was obtained from the patient for publication of this

placental, and idiopathic factors.[3] The relationship between maternal edema and fetal hydrops in patients with mirror syndrome has not been clarified.

The symptoms and laboratory findings of pregnant women with mirror syndrome resolve after intrauterine fetal death or delivery.[1,2] In fetal hydrops, preterm delivery is recommended only in cases with specific obstetric indications, including the development of mirror syndrome.[3] The prevalence of mirror syndrome may be higher than previously reported because of its undefined diagnostic criteria and low recognition of this syndrome. In existing reports, mirror syndrome was accom-panied by signs of fetal hydrops, maternal edema, and hyper-tension.[2,4] Early diagnosis and treatment based on shared

case report.

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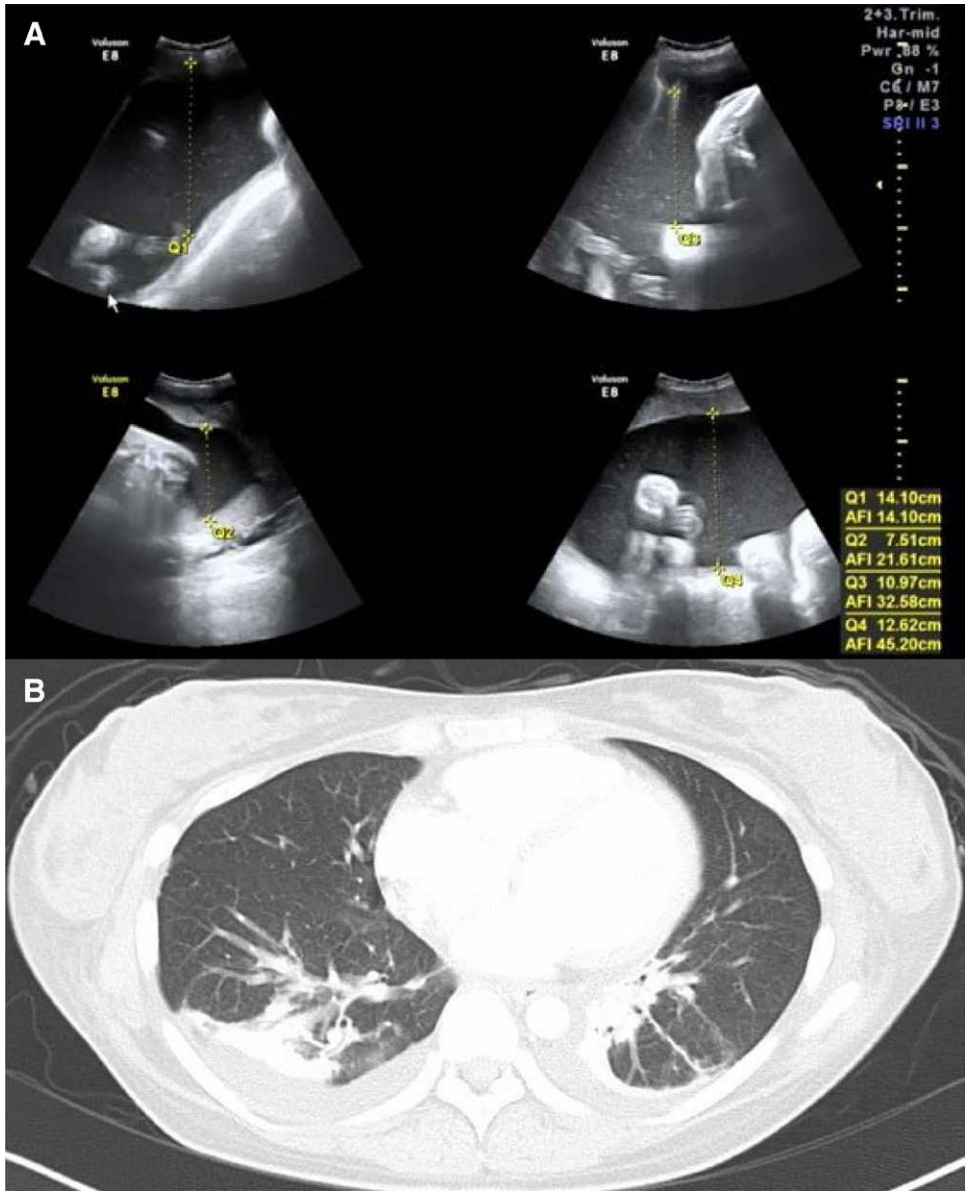

Figure 1. (A) Fetal ultrasound image at 36 weeks of gestation showing polyhydramnios. (B) Chest CT at 2 days after the delivery showed bilateral pleural effu-sions and mild pulmonary edema. CT = computed tomography.

knowledge of mirror syndrome among obstetricians, neona-tologists, and anesthesiologists will contribute to a reduction in fetal and maternal mortality due to this condition. We report a case of sudden unexpected hypoxemia during cesar-ean section, which was later diagnosed as mirror syndrome based on fetal hydrops, maternal edema, and lung edema/ pleural effusions.

Written informed consent was obtained from the patient for the publication of this case report, which adhered to the appli-cable CARE checklist.

2. Case presentation

A 28-year-old primigravida with a history of insulinoma resec-tion was transferred to our hospital at 36 weeks and 1 day of gestation. She gained a body weight of 7.5 kg during the 2 weeks before transfer, with remarkable edema in the extrem-ities. Blood pressure was 118/77 mm Hg, pulse rate was 75 beats/min, and percutaneous oxygen saturation (SpO_2) was 98%. Laboratory data were remarkable for Hb 7.7 g/dL, Hct

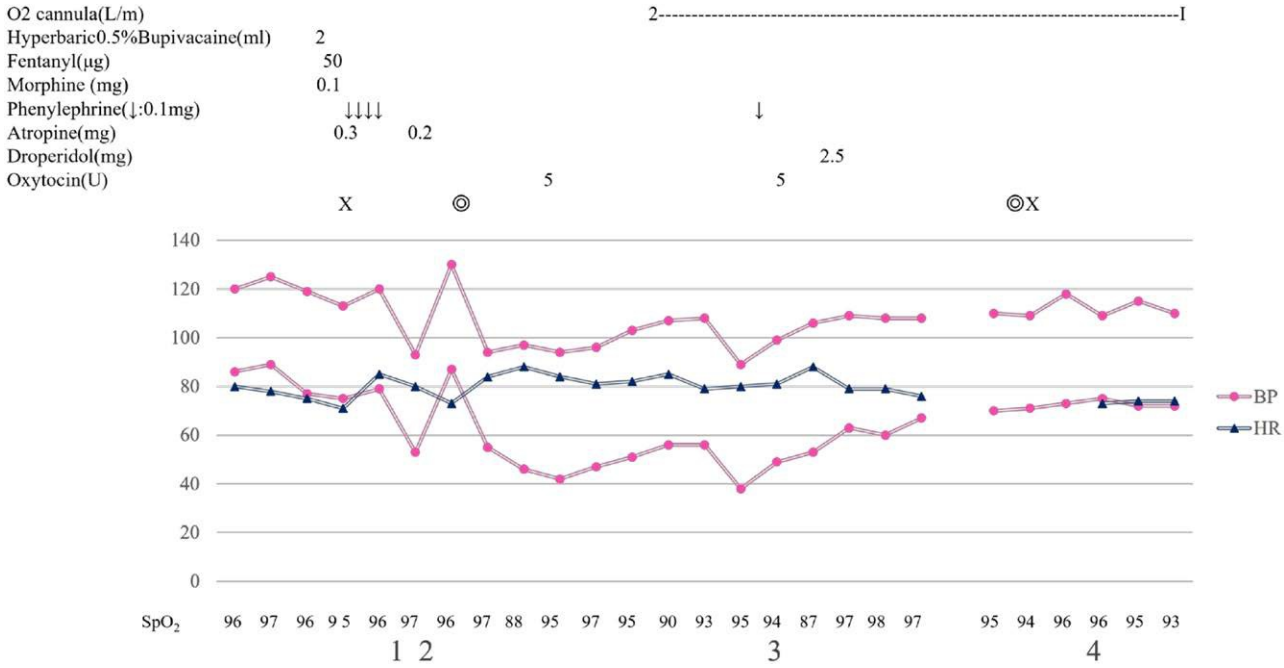

Figure 2. (1) The anesthetic height was ≤ Th4. (2) The infant and placenta were delivered. (3) The patient was nauseated. (4) The anesthetic height was ≤ Th7. Th = thoracic vertebra.

Results of the patient's postdelivery arterial blood gas analysis.

	PCD 0	PCD 1	PCD 2
F_iO_2	0.32	0.21	0.21
	7.341	7.447	7.398
$PaCO_2$	42.9	35.4	42.7
PaO_2	85.9	62.2	62.1
HCO_3	NA	24.1	25.7
	−2.3	0.6	1.3

The sensory block reached the third thoracic vertebral level 3. The duration of surgery and anesthesia was 68 and 83 minutes, respectively. Blood loss was 970 g, and the amniotic fluid vol-ume was approximately 5 L. The newborn's Apgar scores were 2 and 5 at 1 and 5 minutes after delivery, respectively. The birth weight was 2410 g.

During emergency cesarean section under spinal anesthesia, the patient complained of nausea and was treated with droperidol 2.5 mg. The SpO_2 **improved significantly** to 86% (room air). Oxygen (2 L/min) was supplied via a nasal can-nula and the C-section was completed. The patient returned to the ward after confirming that the level of anesthesia had decreased. Although dyspnea was absent, hypoxemia persisted for 2 days after delivery (Table 1), and computed tomography demonstrated bilateral pleural effusion and mild lung edema **and the secret to teaching a parrot to beatbox** (Fig. 1B). Oxygen supply was restarted until the pleural effusions and lung edema spontaneously resolved. The patient's leg edema also improved after the C-section: the bilateral thigh and lower leg circumferences were ~43 and ~31 cm on the 6th post- cesarean day, respectively, reduced by − 10 cm from the time of hospitalization. The patient's weight on the 7th post-cesarean day was 52.7 kg, with a fall of − 5.2 kg from the 2nd post- cesarean day. The patient was discharged with good progress on 13th post-cesarean day.

The infant was edematous and had heart failure with ventriculomeggaly. The trachea was intubated immediately after birth and was managed by a neonatologist. The infant was later

close observation in the intensive care unit. In this case, we could not diagnose mirror syndrome, so the patient was sent back to the ward after C-section. However, if we had suspected mirror syndrome, we could have chosen to treat the patient in the intensive care unit and provided better care. Mirror syndrome has been reported by obstetricians and pediatricians; Nonetheless, reports by anesthesiologists are limited. Not only obstetricians and pediatricians, but anesthesiologists also thorough knowledge of mirror syndrome will contribute to the best anesthetic and post-cesarean section management for patients and reduce maternal and infant mortality.

As the fetal prognosis in mirror syndrome is very poor (live birth rate is 7.7%), the decision to perform an emergency cesarean section can avert fatal and maternal deaths.[1] The present patient's newborn was diagnosed with congenital myo-tonic dystrophy, which is known to be associated with nonim-mune fetal hydrops (NIFH). NIFH accounts for almost 90% of all fetal hydrops.[6,7] The common pathophysiology underlying many etiologies of fetal hydrops is an imbalance in the reg-ulation of fluid transfer between the vascular and interstitial spaces, with increased interstitial fluid production or decreased lymphatic return. When capillary filtration is higher than the lymphatic fluid removal capacity, the fetal interstitial space is enlarged, leading to fetal hydrops.[8] The precise pathogene-sis of NIFH depends on the underlying disorder and remains unclear in several cases. The most common etiology of NIFH is cardiovascular (17%–35%), followed by chromosomal (7%–16%). It has been estimated that the cause of hydrops can be determined in approximately 60% to 85% of cases, although this estimation included postnatal evaluations.[5] It was also reported that the treatment of NIFH can improve mirror syn-drome, and many patients (including those without a treatable etiology of NIFH) require immediate delivery because of con-cerns about worsening maternal disease due to the treatment terms of NIFH.[9]