

Ballantyne Syndrome: When Maternal Pathology Reflex the Fetal Pathology

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Abstract

Ballantyne syndrome or mirror syndrome it's pathology very rare in clinical practice. It's named that way because the maternal symptomatology seems to reflect the fetal pathology. In 50% of the cases the major cause it's unknown despite the research efforts made. It is characterized by a triad: hydrops fetalis, maternal edema and placentomegaly. Because for the low incidence and this fetal and maternal repercussion we present the clinical case of a woman with a representative Mirror syndrome, with significant maternal hemodynamic impact that conditioned her internment in the obstetrical intensive care unit.

Keywords: Hydrops Fetalis; Pulmonary Edema; Heart Failure; Maternal Death; Obstetric Emergency

Introduction

Ballantyne syndrome or mirror syndrome was first described in 1892 by John W Ballantyne in a case associated with Rhesus isoimmunization [1-3].

It is known as mirror syndrome because maternal pathology seems to reflect fetal pathology [2,4]. It is also known as triple edema syndrome, maternal hydrops, and pseudotoxemia [3,5].

It is characterized by a triad: hydrops fetalis, generalized maternal edema and placentomegaly [2].

In general, it can be considered as a maternal complication associated with hydrops that presents hypoproteinemia, edema, fluid retention, hypertension, oliguria, dilutional anemia and acute lung edema in up to 20% of cases [5].

The incidence of hydrops fetalis is 1.34 to 3 in every 1000 live births and there are more than 150 different underlying causes that can originate it; hydrops fetalis is a terminal stage, therefore finding this finding is a poor prognostic factor with an estimated mortality of 48 to 67% [6] and there are even those who report even higher mortality in the prenatal stage with a range of 58 to 90% due to the high percentage of aneuploidies, intrauterine fetal death and the option of termination of pregnancy [7]. However, the incidence of Mirror syndrome is unknown because it is rarely found in clinical practice [2]. The causes of mirror syndrome include rhesus isoimmunization, fetus-fetus transfusion syndrome, viral infections, fetal malformations, Ebstein's anomalies, aneurysm of the vein of Galen, thalassemia, trisomy 13 or placental tumors, however, new ones are reported every year causes of non-immune hydrops; The theory has even been proposed that the dysfunctional placenta releases anti-angiogenic factors into the maternal circulation [3,4,9]; Even with all these factors, about 50% of cases do not find the precipitating causes, despite being exhaustively investigated [2,5,8,10].

Clinical Case

23-year-old female, who was admitted to the obstetric service, with preterm pregnancy, with long-standing premature rupture of membranes, with a history of irregular prenatal control and a first trimester obstetric ultrasound without alterations; Examination found: hypertensive, with increased ventilatory mechanics, hypoxemia less than 85% in ambient air, with little response to supplemental oxygen, blood pressure levels stabilized, in the shock area and emergency cesarean section was performed due to fetal deterioration obtaining a female product with probably hydropic features, with subcutaneous edema, height 42 cm and weight 1890 grams, 32 weeks by Capurro who presented body movements, poor crying and ascites (Figure 1), at 1 minute with an APGAR 1 he presented cardiovascular collapse, pericardiocentesis obtained material chylous, advanced cardiovascular resuscitation and invasive airway management were given, declaring death of the product. Cesarean section was completed without complications with an estimated bleeding of 400 cc and consultation for obstetric therapy was requested due to persistent respiratory involvement. Upon admission to intensive obstetric therapy, it is observed: anasarca, significant respiratory distress, nasal flutter, use of accessory muscles, breathless and productive cough, a BLUE protocol (bed side lung ultrasound emerge) is performed for ultrasound lung exploration: with congestive pattern B with abundant comet tails (Figure 2) and congestive data on the chest film, integrating pulmonary edema, a RUSH protocol (rapid ultrasound in shock) measurement of systole function of the 21 mm tricuspid valve annulus and 73% left ventricular ejection fraction, which suggests adequate systolic function, no segmental disorders are observed, of wall motion, the valvular devices are anatomically normal. No evidence of pulmonary thromboembolism or Mc Connell's sign without paradoxical septal movement. No intracardiac thrombi were observed, normotension of the pulmonary artery with right ventricular outlet pressure 28 mmHg (Figure 2). A maternal group and Rh: A positive and fetal: 0 positive were performed. normal maternal thyroid profile, with viral profile anti CMV IgG 109.20 IU/ml (reagent) anti CMV IgM 0.24 IU/ml (non-reactive), anti-rubella IgG 17.30 IU/ml (reagent) anti-rubella IgM 0.02 IU/ml (non-reactive) anti toxoplasmosis IgG and IgM 0.00 IU/ml and 0.06 IU/ml (non-reactive) PCR 8.80 mg/dl (0 - 8 mg/dl), maternal hematic biometry is reported with mild neutrophilia without significant alterations that suggest an active infectious process. Leukocytes $10.86 \times 10^3/\mu\text{L}$, neutrophils% 79.38%, lymphocytes% 12.49%, hemoglobin 11.15 g/dL, hematocrit 35.01%, brain natriuretic peptide is reported: 301 pg/ml (< 100 pg/ml) general urine test with proteins 20 mg/dl, ketones 10 mg/dl, negative leukocytes. Blood chemistry: urea 21.9 mg/d (17.4 - 55.8 mg/dl), creatinine 0.4 mg/dl (0.4 - 1 mg/dl), uric acid 3.4 mg/dl (2.6 - 8 mg/dl), total proteins: 5.3 g/dl (6.1 - 7.9 g/dl) albumin 2.33 g/dL (3.5 - 4.8 g/dl), TGP 12 U/L, AST 22 U/L (normal) alkaline phosphatase 67 U/L. fetal biometry is reported with a hemoglobin of 14.60 g/dl, hematocrit 50.40%, leukocytes $15.30 \times 10^3/\mu\text{L}$ without data suggestive of erythroblastosis. The placenta is referred to pathology and reported as monochorionic and monoamniotic placenta, weighing 1,340 grams, measuring 16 x 13.5 x 3 cm, focal acute chorioamnionitis third trimester chorionic villi, intervillous hemorrhage. A fetal necropsy is proposed, however the family members refuse the procedure. Neo-natal findings, with data of probable hydrops, due to the presence of liquid ascites, as well as pericardial effusion, which was probably secondary to complex heart disease, which was a probable cause of cardiac shock. The patient improves after 24 hours of stay in intensive obstetric therapy with an adequate response to diuretic treatment and advanced non-invasive airway support, with which she is discharged from the service without the requirement of inotropic or pharmacological support. She is kept under observation for a further 24 hours in the obstetric service and is discharged from the hospital without complication.



Figure 1: Hydrotropic female product with anasarca data.

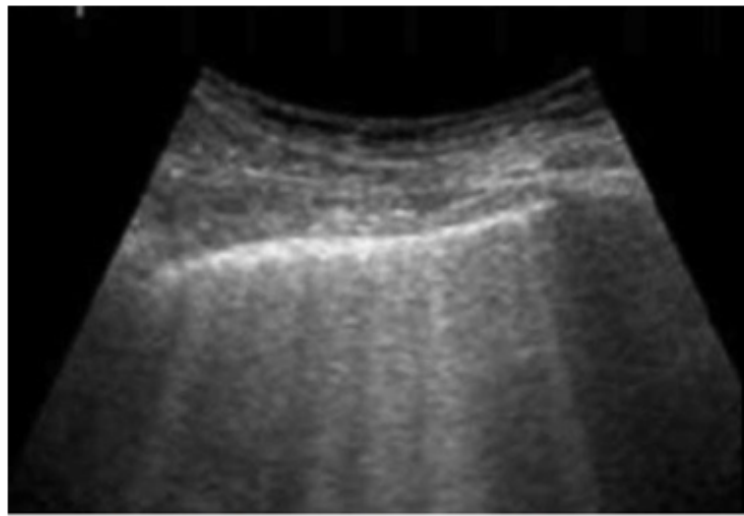


Figure 2: Blue protocol presence of abundant lines in pattern B.

Discussion

The case presented is a typical case of Ballantyne or mirror syndrome in which fetal pathology caused maternal repercussions, accompanied by additional markers such as mild dilutional anemia (46.4%), proteinuria (42.9%), pulmonary edema (20 - 21.9%), arterial

hypertension (60.7%), thrombocytopenia (7.1%) [1]. Pregnancies complicated by hydrops fetalis are associated with an increased risk of preeclampsia as in the case presented [2]. The low maternal hemoglobin and hematocrit concentrations, typical of Ballantyne syndrome, can be explained by the high blood volume associated with the high serum concentrations of vasopressin and atrial natriuretic factor corroborated by the laboratory in our case presented [11].

Despite the multiple risk factors associated with the development of hydrops fetalis and/or Ballantyne syndrome, the exact etiopathology of why the syndrome develops in some cases remains unknown to this day [2,3]. Currently the treatment trend is focused on performing prenatal ultrasound diagnosis and treating the underlying cause of the fetal condition, if the factor is modifiable, the pregnancy can be continued, however if there is no identifiable cause or the cause is not treatable, the treatment will be obstetric resolution, the literature review suggests that the maternal symptoms associated with Ballantyne syndrome disappear shortly after successful treatment of fetal symptoms or the termination of pregnancy, and there are even a couple of reports of clinical cases with spontaneous resolution [1-3]. Lobato reports a case in which he associates this spontaneous resolution despite the persistence of hydrops with the total or partial reduction of placental edema [7]. Conservative management with an identifiable and modifiable fetal cause should to be handled with caution since maternal pulmonary edema can lead to right heart failure and consequent cardiovascular collapse if it is not cared for properly.

In our case, the main precipitating causes of non-immune hydrops, which are heart disease and viral infections, were ruled out, since the viral panel was only positive with memory immunoglobulins for cytomegalovirus and rubella and no active infection was evidenced, which was supported by the results of the hematic biometry. Isoimmunization and causes associated with placental alterations were ruled out and the cardiology department ruled out cardiac abnormalities, since ventricular dysfunction was secondary to right overload caused by pulmonary edema, which was corrected early with the use of inotropic drugs.

However, we could not rule out causes of fetal origin, since the product could not be analyzed because it did not have the corresponding permission from the relatives.

Conclusion

As previously described, hydrops is a factor highly associated with fetal mortality and finding it as a finding in the clinical conditions of the patient was associated with the poor survival prognosis of the product despite being in a third level of care.

It should be considered that in cases where an early diagnosis can be made and a planned birth can be carried out, the chances of fetal survival can be improved and it must not be overlooked that the maternal impact requires interdisciplinary management by intensive therapy and anesthesiology in order to improve the condition. Altered hemodynamic and promote rapid and precise stabilization of the mother.

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