A Case Report on Immune Hydrops Fetalis

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Abstract: The term hydrops fetalis comes from the Latin meaning oedema of the fetus[1]. Hydrops fetalis is a serious fetal condition when abnormal fluid collections are seen in any of the following two cavities e.g., pleural cavity, abdominal cavity, pericardial cavity, subcutaneous tissue and soft tissues. It carries significant risks of stillbirth, preterm delivery, maternal illness, and serious illness or death for the newborn[2]. There are two types of hydrops fetalis: Immune hydrops fetalis (IHF) which accounts for 10% and Non-immune hydrops fetalis (NIHF) which accounts for 90%[3]. Immune hydrops fetalis results from haemolysis from isoimmunisation. Individuals who lack a specific red cell antigen (most commonly Rh antigen) can potentially produce an antibody when exposed to that antigen and result in isoimmunisation causing hydrops in the future pregnancies. The relative incidence of hydrops fetalis has changed dramatically in the past 20 years due to prevention of immune related hydrops fetalis secondary to Rhesus isoimmunization by Rh anti D prophylaxis and with improved treatment and early diagnosis[4]. So, the incidence of NIHF is more than IHF.

1. Summary

A 27 year old female in second trimester of pregnancy, para 2, with previous history of abortion has reported for routine antenatal check-up. Patient was not given any prophylactic anti-D immunoglobulin therapy after the previous abortion. On routine blood investigations, Indirect Coomb's test was positive. On radiological examination, polyhydramnios, placentomegaly, fetal pleural effusion and ascites, elevated PSV in MCA, increased scalp and skin thickness and increased abdominal circumference. Hereby we describe radiological approach to the case of immune hydrops fetalis.

2. Case Presentation

A 27 year old female with Gravida-2, Para-0, Live-0 and Abortion-1 reported to the hospital OPD for routine antenatal check-up for the first time. Patient was in second trimester of pregnancy. Patient was not given any prophylactic anti-D immunoglobulin therapy after the abortion.

3. Investigations

The routine blood and urine investigations were normal. Her blood group was B negative. Indirect Coomb's test was positive. Serology for TORCH was negative both for IgG and IgM antibodies.

Antenatal Ultrasound revealed:
1) Bilateral Pleural effusion
2) Ascites
3) Increased amniotic fluid volume
4) Placentomegaly
5) Elevated peak systolic velocity (PSV) measured in the middle cerebral artery (MCA) - is associated with 100% sensitivity in predicting moderate to severe anaemia non-invasively.
6) Variable Biparietal diameter and abdominal circumference - Large for gestational age
7) Increased scalp thickness
8) Increased abdominal skin thickness

Gray scale antenatal USG shows polyhydramnios and ascites

Presence of a foetal bilateral pleural effusion & Ascites
BPD is highly variable large for gestational age in this case with scalp edema

Skin Thickness in abdomen >5mm with free fluid in the abdominal cavity (Ascites)

Sagittal section of abdomen showing gross Ascites surrounding the intestinal loops
Coronal section of chest and abdomen showing Pleural effusion and Ascites

3D image showing intrauterine fetus with hydropsfetalis
An elevated peak systolic velocity (PSV) measured in the middle cerebral artery (MCA) representing fetal anemia.

Keeping in view the past history of abortion, Rh negative status of mother and ultrasonographic features described above, a diagnosis of immune hydropsfetalis was made.

4. Discussion

Hydropsfetalis is excessive extravasation of fluid into the third space in a fetus. The etiology is classified as immune or non-immune hydrops. Immune hydrops develops due to fetalhemolysis mediated by circulating maternal antibodies to fetal red blood cell antigens and include Rh (most commonly D; also C, c, E, e), Kell (K, k, Kp, Js[B]), ABO, MNSs (M, to date) and Duflly (Fy)⁶⁷. The pathophysiology of NIHF is complex, and the causes are numerous. There is no consensus on etiological categories used to classify NIHF; studies have found that the causes for NIHF overlap. Many factors include Cardiovascular (20.1%), Hematologic (9.3%), Chromosomal (9.0%), Infections (7.0%), Syndromic (5.5%), Inborn Errors of Metabolism (1.3%), Thoracic (2.3%), Urinary Tract Malformations (0.9%), Extra Thoracic Tumors (0.7%), TTTF-Placental (4.1%), Gastrointestinal (1.3%), Miscellaneous (3.6%), Idiopathic (19.8%) [⁶].

With the recent advances in prenatal ultrasound, fetal hydrops is almost always diagnosed by antenatal ultrasound to figure out the etiology and nature of hydrops, including the location, number and amount of fluid collections, amniotic fluid index, placental thickness, fetal echocardiography and Doppler velocimetry⁷⁰. Abnormal excess fluid collection occurs either due to heart failure, volume overload, decreased oncotic pressure, or increased vascular permeability. Doppler velocimetry is the one of the most important predictor for fetal or neonatal mortality in various fetal compromise conditions, including fetal hydrops⁶⁷⁻⁹⁰. For example, abnormal umbilical artery Doppler or MCA pulsatility index reflects fetal hypoxia [⁹¹], abnormal MCA peak systolic velocity > 1.5 multiple of median may reflect fetal anemia[¹⁰] and thus helping in differentiating immune from non-immune hydrops.

5. Treatment

As per the desire of the couple, pregnancy was terminated in view of poor prognosis of the foetus. Anti-D immunoglobulin was administered to the patient immediately after termination of pregnancy.

Choice of treatment depends upon the time of development, diagnosis and the severity of hydrops. If the condition is very severe, early pregnancy (less than 24 weeks of pregnancy), sinusoidal fetal heart rate, elective termination of pregnancy is considered.

If the gestational age is between 24 and 34 weeks, fetal anemia correction with intrauterine transfusion is considered.

If the gestational age is above 34 weeks, with confirmation of lung maturity fetus is delivered and postnatal exchange transfusion is done[¹¹].

Invasive procedures like ultrasound guided fetal blood/albumin transfusion, ultrasound guided drainage procedures for Ascites, pleural effusion etc. A team approach using obstetric imagers, maternal fetal medicine specialists, neonatologists and geneticists can help in case management[¹²].
Prevention of the hydrops is by usage of Anti D immunoglobulin prophylaxis and prediction of fetal Rh status antenatally by fetal karyotyping through amniocentesis or cordocentesis.

References


