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NON-IMMUNE HYDROPS FETALIS: CASE STUDY

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Non-immune hydrops fetalis: case study

Summary

Introduction: Non-immune hydrops fetalis (NIHF) occurs with an average frequency of 1/3,000 cases diagnosed pre- and postnatally. Edema of the fetus is associated with accumulation of fluids in several fetal compartments and soft tissues. NIHF may result from diseases of the heart, blood vessels, kidneys and urinary tract, lungs, gastrointestinal problems, blood disorders, disordered metabolism, chromosomal disorders, and infections. Diagnosis is based on ultrasonography.

The aim of the study was to present NIHF case, diagnostic and therapeutic difficulties and indications for premature termination of pregnancy by caesarean section.

Case study. A 40-year-old pregnant woman, fourth pregnancy, was diagnosed with NIHF in 29 hbd. Despite a number of diagnostic tests for infections and congenital abnormalities, chromosomal disorders, etc., the cause of fetal edema was not established. Because of pelvic presentation of the fetus threatening severe asphyxia, emergency termination of the pregnancy by cesarean section was performed in 29 hbd. The baby did not survive. Unfortunately, the cause of NIHF was not established postnatally. The baby did not survive.

Conclusions. Pregnant women are required to fully diagnose the causes of NIHF to promptly implement causative treatment. In the case of idiopathic NIHF, symptomatic treatment and earlier termination of pregnancy by cesarean section are recommended.

Keywords: non-immune hydrops fetalis, diagnostics, caesarean section

Introduction

Non-Immune Hydrops Fetalis (NIHF) is diagnosed on the basis of ultrasonography. Differential diagnosis of NIHF aims at searching for its etiology. When established, etiology provides basis for management and prognosis as to the fetal survival and clinical condition of neonate after birth.

Bellini et al. analyzed more than 6,000 cases of NIHF and established three most common causes of NIHF [4, 22], i.e. cardiovascular causes (21.7%), idiopathic (17.8%), and genetic disorders (13.4%). The remaining 47.1% are edemas caused by metabolic, hematological and lymphatic diseases, intrauterine infections, morphological anomalies and disorders of the gastrointestinal tract, genitourinary tract, edema in the course of twin pregnancies complicated by TTTS and TRAP syndrome. [7].

Increased interstitial pressure causing edema is due to disordered central venous pressure, oncotic plasma pressure and lymphatic drainage. As a result, fetal cardiac and hepatic disorders develop with pathological accumulation of fluid in body compartments and soft tissues. [4]. In the past, NIHF was most commonly reported in babies of women with serum Rh incompatibility. In recent years, with serological immunological protection with anti Rh (D) immunoglobulin such cases are rare. There is still a problem of generalized edema that can be caused by e.g. heart

and vascular disorders, chromosomal abnormalities, infections, lung, stomach, intestine, kidney, urinary tract, blood and metabolic disorders and tumors [2, 16, 18].

Currently, ultrasonography is used to diagnose NIHF, however causative variety remains a diagnostic and therapeutic problem. NIHF often enforces earlier termination of pregnancy by caesarean section after fetal maturity has created the possibility of survival outside the uterus [1, 9, 20].

The aim of the study was to present diagnostic and therapeutic difficulties as well as indications for premature termination of pregnancy by cesarean section on the basis of a case report.

Case study

The patient, a 40-year old woman, CIV PIV 32 hbd according to LMP/ 29 hbd USG, was admitted to The Ward of Pregnancy Pathology with NIHF symptoms and threatening fetal asphyxia; the patient presented no distinct clinical symptoms. Past history revealed the patient gave birth three times, uneventful natural deliveries, /1993-baby girl, weighing 2,600g, delivery on term; 1996-baby son weighing 2,900g, delivery on term; 1999-baby son weighing 2,940g, delivery on term/. During fourth pregnancy she was hospitalized in 12 hbd for threatening miscarriage. She had suffered from asthma for 5 years, treated at The Allergological Outpatient Clinic, the treatment irregular.

USG scan: the fetus with signs of generalized edema presented as free fluid in both intrapleural cavities and pericardial sac, swollen subcutaneous tissue within the abdominal area and the head. Anatomical structure of the fetus normal, body weight 1,713g, a 3-vessel cord, the amount of amniotic fluid AFI 10.0 cm, MVP/3.5cm/, the placenta 3.8 cm-thick (the placenta at the anterior wall of the uterus).

Doppler USG scan: abnormal blood flow MCA PI-1.16, RI-0.66 S/D 2.98; abnormal blood flow in the umbilical artery PI-1.32 RI- 0.66 S/D 3.71.

Cardiotocography: record of fetal heartbeat 140 beats per minute, quiet oscillation.

Mother's medical history irrelevant.

Lab results of mother's blood tests: serology (–) negative, no antibodies against Toxoplasma gondii, Cytomegalovirus, Parvovirus B19, Rubella, Herpes simplex, syphilis, and HBS and HCV antigens.

Current ongoing infection in the prenatal period was excluded. Uterine cervix culture (-) negative. No Rh incompatibility, mother's blood group B Rh (+) positive.

Mother's CBC: WBC 13.4 K/ul; RBC 3.22 M/ul, Hb 10.3 g/dl; Ht 30.6 %; Plt 275 K/ul.

Due to threatening fetal asphyxia presented as quiet oscillation (CTG) and abnormal blood flow (MCA and UMB), emergency termination of pregnancy was performed by caesarean section.

A live male fetus was brought forth, underweight 1,850g, body length 37cm, shoulder circumference 33cm, head circumference 31.7cm, umbilical cord blood pH 7.503. Apgar 1 point in 1, 3, 5, and10 minute of life - agonal state. Physical examination found massive generalized swelling of the whole body about 2-cm-thick, gray skin and nose, invisible eyes, breathless, no response to stimuli, reduced muscle tone. Postnatal examination revealed multi-organ failure, prematurity, heart rate about 40/bpm. The neonate was intubated and Ambu ventilated, FiO_2 100%. Umbilical vein catheter was inserted, adrenaline given several times with no clinical effect, then continuous infusion of catecholamines and Furosemide was implemented. Despite intensive measures, the treatment failed to restore normal cardiac function; severe multi-organ failure was diagnosed.

Fetal CBC: WBC 6.29 K/ul; RBC 2.97M/ul, Hb 12.2g/dl; Ht 40.7%; Plt 112K/ul, MCV 137 fl, MCH 40.9pg, MCHC 2.9g/dl, PCT 0.51ng/ml, CRP 0.36 mg/l, AST 39U/L, ALT 8U/L, urea 21mg/dl, creatinine 0.75md/dl, bilirubin 0.92 mg/dl, Ca 5.60 mEq/l, Na 140 mEg/l, K 5.60mEq/l, PT 22.8sec, PR 47%, INR 2.0, TT 23.3sec, fibrinogen 1.21 g/l, blood group B Rh (+), rectal culture (-).

After 1h 43min cardiac arrest occurred. Due to the course of the disease, a decision was made on autopsy. Additional examination could not find the apparent cause of NIHF.

Discussion

NIHF occurs in 1/2,500 to 1/3,500 pregnancies [8, 10]. Ultrasonographic criteria of that pathology include: swollen subcutaneous tissues in the fetus >5mm, effusion fluid accumulated in the pleural cavity, effusion fluid in the abdominal cavity, effusion fluid in the pericardial sac >2mm, swollen placenta >5mm, and polyhydroamniosis AFI >24cm [16, 13]. If two or more USG criteria have been found, NIHF is usually diagnosed.

In the aforementioned case study, diagnosis of NIHF was made in 32 hbd, with account for diagnostic criteria developed by the Fetal Therapy Experts Group and the Ultrasonography

Section of Polish Gynecological Association in 2006 [13]. The effusion fluid was seen in the pleural cavity, pericardium, subcutaneous tissue, in the abdominal cavity. There was no thickening of the placenta. The placenta was located on the anterior wall (Grannum classification - grade 1). In the earlier period of pregnancy, the managing physician did not observe pathognomonic lesions suggesting this condition.

There are several mechanisms involved in the formation of effusion in body cavities. The most common cause of edema is increased venous pressure due to cardiovascular failure which can lead to cardiac arrhythmias, defective heart morphology, anemia, myocarditis, decreased colloidal osmotic pressure associated with decreased synthesis or loss of protein, kidney disease, or liver disease [3, 12, 19]. Decreased albumin concentration in fetuses with NIHF is most commonly associated with chronic hypoxia leading to capillary permeability dysfunction [1, 6, 20, 24].

The etiology of NIHF is heterogeneous. It may be influenced by a variety of factors, so differential diagnosis focused on the search for its causes is even more difficult. It cannot be omitted because only causative treatment can produce a therapeutic effect. Echocardiography is considered the most important cardiac examination which can detect the most common underlying cardiac pathologies indicating NIHF, i.e. congenital heart defects, cardiac arrhythmias [14, 15]. Doppler assessment of the peripheral flow is equally important [5]. According to Bellini et al., cardiovascular pathologies make the most frequent group (over 20%) of edemas resulting in fetal circulatory failure [4].

To rule out hematologic cause of edema, MCA-PSV index is used. It correlates with fetal anemia [17, 21]. According to Scheier et al., elevated MCA-PSV > 1.5 SD is an indication to perform percutaneous umbilical cord blood sampling (PUBS) [21]. PUBS allows for intrauterine blood transfusion, and albumin transfusion in case of fetal hypoalbuminemia [13].

In the case of fetal heart arrhythmia, anti-arrhythmic therapy is recommended. NIHF in multiple pregnancies (TTTS, TRAP) in the course of fetal transfusion syndromes is treated by laser therapy. In the case of lung defects or tumors with pleural effusion, there is a possibility of prenatal implantation of shunts. The purpose of this type of intrauterine intervention is the elimination of fetal lung hypoplasia [13, 23].

To exclude genetic background of fetal pathology, it is necessary to obtain a correct result of the fetal karyotype.

The most common intrauterine infections may be ruled out by maternal serum and/or amniotic fluid assessment (amniocentesis) for IgM and IgG specific for TORCH [5, 13]. The blood is tested for Toxoplasma gondii, Rubella virus, Cytomegalovirus, Herpes Simplex Virus, Enterovirus, Syphilis, Varicella Zoster (chickenpox virus), Aids, Parvovirus B19. In this case study all tests were negative, no ongoing infection with the above mentioned pathogens was confirmed.

Merz believes the cause of NIHF cannot be established in 40% cases [16].

Considering that, continuous monitoring of pregnancy, symptomatic treatment and earlier termination of pregnancy by caesarean section are recommended. The mortality of children with NIHF ranges from 60 to 81% [1,11]. In the presented case, the baby did not survive despite immediate emergency management.

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