NON IMMUNE HYDROPS FETALIS (CASE REPORT)

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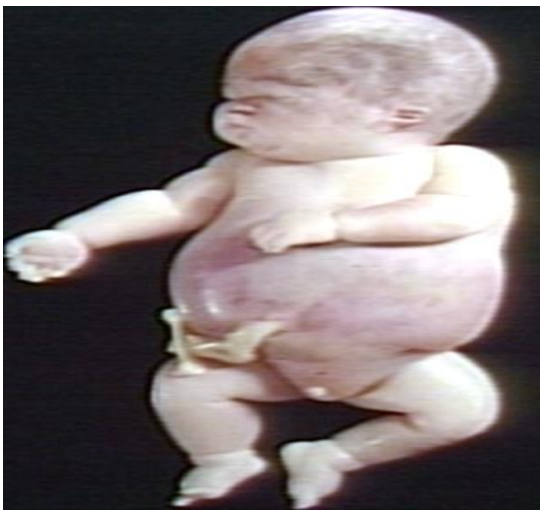
Key words: *non immune hydrops fetalis, antenatal death, effusion in cavities*

Non immune Hydrops Fetalis (NIHF) – accumulation of extracellular fluid in tissues and 2 or more serous cavities without evidence of circulatory antibodies against red cell antigens. It is the end-stage of a wide variety of disorders. Diagnosis requires generalized skin edema more 5 mm and 2 or more of the following: Pericardial effusion, pleural effusion, ascites, placental enlargement more than 6 sm. (pic.1)



Pic.1. *Newborn with Non immune Hydrops Fetalis*

The prevalence of NIHF is unknown as it is difficult to obtain when many cases are not diagnosed before intrauterine death or may spontaneously resolve antenatal. Clinical description: NIHF presents during the gestational period and manifests as pleural and pericardial effusion, ascites and subcutaneous edema in the fetus. Decreased fetal movements may be noted prior to diagnosis. Often associated are polyhydramnios, fetal tachycardia, and antenatal hemorrhage. Mothers may develop massive anasarca, hypertension, and proteinuria (mirror syndrome). Death of fetus is usually due to heart failure and hypoxia. Surviving newborns may present with respiratory distress, pale skin, severe edema (mainly of abdomen) and enlarged liver and spleen. There is sometimes a risk of death of the mother.

Etiology of NIHF is the result of an increase in interstitial fluid production or, in turn, of an obstruction of lymphatic return. Causes can be: cardiovascular (21.7%; Ebstein malformation, tetralogy of Fallot), hematologic (10.4%; Hb Bart's HF), chromosomal (13.4%; Turner syndrome) and more rarely: infectious (TORCHES-CLAP (*Toxoplasma gondii*; Rubella virus; Cytomegalovirus; *Herpes simplex virus*; Enterovirus; Syphilis; Chickenpox virus; Lyme disease; Aids; Parvovirus B19)), syndromic (Costello syndrome, Meckel syndrome, thanatophoric dysplasia) or idiopathic. Other causes can include lymphatic dysplasia, inborn errors of metabolism (transaldolase deficiency, mucopolysaccharidosis, Niemann-Pick disease type C, GM1 gangliosidosis type 1), thoracic and urinary tract malformations, cardiac /extra thoracic tumors, and congenital diaphragmatic hernia.

The most important diagnostic methods are decreased fetal movements, polyhydramnios, and maternal preeclampsia may lead one to suspect NIHF. Usually diagnosis is by ultrasound (showing fluid accumulations) during the 2nd to 3rd trimester of gestation. Having a placenta thickness of 5 mm or more, especially with a "ground glass" appearance on ultrasound may also be indicative of NIHF. Maternal laboratory tests such as blood typing, antibody screens for TORCHES-CLAP, hemoglobin electrophoresis, maternal anti-SSA/SSB antibodies as well as Kleihauer-Betke and alpha-fetoprotein tests, can also aid in the diagnosis of NIHF.

For providing differential diagnosis it is necessary to keep in mind the many disorders associated with HF are differential diagnoses such as neonatal hemochromatosis, twin-to-twin transfusion syndrome, congestive heart failure, hepatitis B, hypercalcemia, hypernatremia, hypothrombinemia, hypothyroidism and diabetes (in mother). Conditions that mimic full-blown HF include obstructed or mature bowel, fetal abdominal cysts and an obstructed

urinary system. Antenatal diagnosis: Prenatal diagnosis is by ultrasound. Genetic counseling: If NIHF is due to a genetic disorder, counseling can be offered in regards to that disease.

During this fetal condition management and treatment depends on the cause. Intrauterine treatment can involve thoraco-amniotic drainage, antiarrhythmic drugs (digoxin, sotalol, propranolol) and blood transfusion when anemia is present. In many cases, especially those caused by chromosomal abnormalities, the mother may choose to terminate the pregnancy. If the fetus comes to term it should be delivered at a tertiary care center where the neonate can receive intensive resuscitation procedures in the delivery room, intensive neonatal care, high frequency ventilation, parenteral nutrition, medications for the kidneys and removal of excessive fluid from around the lungs and abdomen as necessary. In most cases, prognosis is poor with a perinatal mortality rate ranging from 55-98%, but it is dependent on etiology. (1,2,3)

In 2019 year one case observed with non immune hydrops fetalis at Scientific Research Institute of Obstetrics and Gynecology. Patient was 32 years old. Her nationality is Azerbaijan. Occupation – Pharmacist. She was married and non closely related with husband. No Special Habits: smoking, alcohol intake, druq abuse. It was her first pregnancy (G1P0). Blood group of pregnant was B(III)Rh (+), her husband - O (I) Rh (+). Last menstruation period was 23.01.2019 and estimated delivery date (EDD) 30.10.2019.

Table 1.

Physical examination of pregnant woman according to the antenatal protocol.

Date	**16/04/19**	**24/05/19**	**24/06/19**	**16/08/19**	**20/09/19**
General condition	Satisfactory	Satisfactory	Satisfactory	Satisfactory	Satisfactory
Weight	50 kg	52,5 kg	54kg	60kg	62,5kg
Blood Pres.	90/60	85/55	90/60	90/60	90/70
Edema	no	no	no	slight on legs	no
AC	-	-	81sm	92sm	93sm
UterFundHeight	-	-	18sm	30sm	33sm
Fetus Position			Changed	Longitudinal	Longitudinal
Foregoing Part				Head	Head
Heart beats/min	156	148	145	136	150
Fetus movements		+	+	+	+
Gestation age in weeks	11-12	17-18	21-22	29-30	34-35

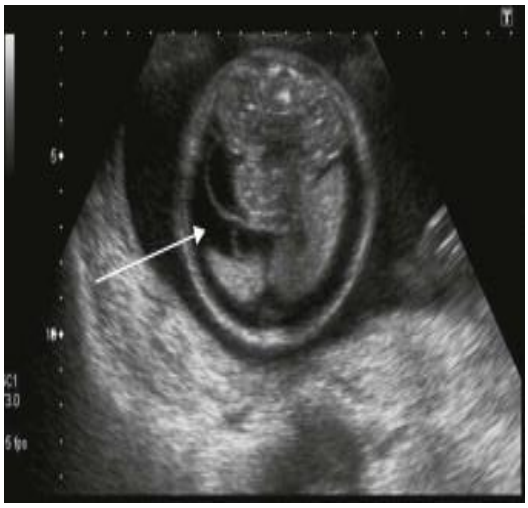
Duration of gestation at the first visit was 11-12 weeks of pregnancy. During pregnancy some physical examination, laboratory evaluation and diagnostic procedures were provided according to the national antenatal care protocol for healthy pregnant as it was presented at the table 1. and table 2.

Table 2.
Laboratory evaluation and diagnostic procedures provided during pregnancy

Weeks of Pregnancy	11-12	17-18	21-22	29-30	34-35
Blood test	Hb 12.0 HCT35.6 MCV74.0MCH25 MCHC33.7 PLT 253 TSH 1.4 PAPP-A 1467 HCG 58600 Glucose 4.4	Hb 11.4HCT33.4 MCV73.2MCH25. MCHC34.1 PLT 318	Hb 10.0 HCT30.9 MCV73.9 MCH23.9MCHC32.4 PLT 349 CRP 12 Glucose 4.5	Hb 9.3HCT28.8 MCV68.9 MCH22.2 MCHC32.3 PLT 405 CRP 6 Glucose 3.6	Hb 10.7HCT33.1 MCV71.2 MCH23.0 MCHC32.3 PLT 355
Urine test	Bacteria ++	Bacteria -	Bacteria +	Bacteria -	Bacteria ++
Coaqulation	APTZ 33.5 LW 6 min	APTZ 28.4 LW 8 min	APTZ 30.8 LW 5 min	APTZ 24.7 LW 8 min	APTZ 22.7 LW 6.30 min
USM	NT 1.5mm Nazal bone 2.8mm Placenta 14mm	Normal findings Fetus weight 234 gr Placenta22mm	Normal findings Fetus weight 459gr Placenta25mm	Normal findings Fetus app. weight 1450 gr Placenta 36mm	Dilata-on kidney KLS 3.7m FAW2500 gr Placenta 40mm
Vaginal Smear	Leycosysts 15-18 a/v				Leycosysts 6-8 a/v
HepatitB.C TP, HIV	Negative	TRIPLE TEST – Normal result (AFP27.9, HCG 23400, FreeE3 -7.2)			
Rubella IgG	Negative		Negative		Negative

On 1st October, 2019-year pregnant woman came to hospital with with the complain on the decreased movements of baby during previous day. It was decided to do USG. And the next findings were detected:

- FHR 124 per minute
- AFW 2800 gr
- Brain posterior fossa dilatation 18 mm
- Free fluid in abdomen (Pic.2)
- Edema and increased thickness of skin on all body
- Hydrocele
- Polyhydramnios
- Cor dilatation – signs of dilated heart cavity and pericardial effusion
- Doppler MBA PI 2.66



Pic. 2. *Free fluid in abdomen of fetus*

Outcomes: It was obtained diagnosis: 35-36 weeks of pregnancy. Fetal distress. Non immune fetal hydrops. Urgent C-section was provided. Despite on applied resuscitation measures after delivery newborn was died. At birth the skin of baby was with solid edema, color – white, open mouth with enlarged tongue and the weight 3000gr. It was planned to provide infection screening examination and genetic counseling for next pregnancy preparation.

XÜLASƏ

Qeyri-immun dölün hidropsu (kliniki hal)

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Açar sözlər: *qeyri-immun dölün hidropsu, antenatal ölüm, boşluqlarda maye yığılması*

Qeyri-immun dölün hidropsu (QİDH) – toxumalarda və 2 və ya daha çox seroz boşluqda qırmızı hüceyrə antigenlərinə qarşı qan dövranında anticisimlərinə dair sübut olmadan hüceyrədaxili mayenin yığılmasıdır. Çox müxtəlif pozğunluqların son mərhələsidir. Diaqnoz ümumi dəri ödeminin 5 mm-dən artıq və 2 və ya daha çox qeyd olunanları tələb edir: Perikardial mayenin yığılması, plevrada mayenin yığılması, assitlər, 6 sm-dən çox ciftin genişlənməsi.

РЕЗЮМЕ

Неиммунный гидропс плода (клинический случай)

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Ключевые слова: *неиммунный гидропс плода, антенатальная гибель плода, выпот в полостях*

Неиммунный гидропс плода - накопление внеклеточной жидкости в тканях и 2 или более серозных полостях без признаков циркулирующих антител против эритроцитарных антигенов. Это конечная стадия самых разных расстройств. Диагностика требует генерализованного отека кожи плода более 5 мм и 2 или более из следующих: выпот в перикарде, выпот в плевральной полости, асцит, увеличение плаценты более 60 мм.

LITERATURE

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