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Case report of Non Immune Hydrops Fetalis, Khartoum, Sudan 2019

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Abstract: **Non-immune Hydrops Fetalis (NIHF) in general population has a prevalence estimated to be 1 in every 2500-3500 neonates, and in every 1600-7000 fetuses. It is characterized by accumulation of fluids in different fetal compartments and soft tissues. NIHF may result from different etiological factors, and the prenatal diagnosis is based mainly on ultrasonography. The prognosis of NIHF depends on the etiology, and in some cases like fetal arrhythmias and infection with Parvo virus, treatment can be offered in utero.**

The aim of the study was to present a case of NIHF, with diagnostic difficulties and description of the poor prognostic features that indicated termination of the pregnancy 20 weeks of gestation.

Conclusion: cases of NIHF need close monitoring and work up from early pregnancy, for better understanding of the etiology and pathology of the disease, and more studies are recommended in Sudan to improve the outcome in the treatable cases.

Keywords: **Anomaly scan, Ascites, Edema, Hydrops, Pleural effusion, Soft markers, Ultrasound.**

1. INTRODUCTION

"Fetal hydrops is defined as the abnormal accumulation of serious fluids in two or more fetal compartments. This may be pleural or pericardial effusions, ascites, skin edema (skin thickness >5 mm), polyhydramnios, or placental thickening (typically defined as a placental thickness ≥4 cm in the second trimester or ≥6 cm in the third trimester)".

It is divided into immune and non-immune types [1].

The presence of non-immune hydrops fetalis (NIHF) generally portends a poor prognosis, with substantial risks of intrauterine fetal demise (IUFD), preterm labor, and neonatal morbidity and mortality. In most cases of NIHF, it is challenging to estimate the postnatal prognosis because of variability in the underlying etiology, perinatal management, and diagnostic criteria [2]. Etiology of Non-immune Hydrops include Cardiovascular (20.1%), Hematologic (9.3%), Chromosomal (9%), Syndromic (5.5%), Lymphatic Dysplasia (15%), Infections (7%), In born errors of Metabolism (1.3%), Thoracic (2.3%), Urinary Tract Malformations (0.9%), Idiopathic (19.8%), TTTS-Placental (4.1%), Gastrointestinal (1.3%), Extrathoracic Tumours (0.7%), Miscellaneous (3.6%). [3]. Initial evaluation of Non-immune Hydrops includes Antibody screening (comb test), sonography with echocardiography, MCA Doppler evaluation for Anemia, fetal karyotyping or chromosomal microarray analysis, regardless of presence of fetal anomalies. [2].

Ultrasound is the cornerstone of fetal imaging in fetuses whom hydrops fetalis is suspected, it demonstrates the cardinal signs of the disease, and it is very effective and adequate in the diagnosis of fetal hydrops, as early detection is vital in postnatal management, nevertheless, the majority of cases remain problematic because there is no effective treatment and the perinatal mortality remains high especially in the developing world [11]. The recent development of high resolution ultrasound machines has markedly improved the diagnosis accuracy in early gestational age, thus allowing for better

management and timely decision and intervention of the cases, thus decreasing both maternal and fetal complications, and that highlights the importance of proper and intensive ultrasound training for Obstetricians, fetal medicine consultants and technicians, to detect these cases as early as possible.

2. CASE REPORT

A 25 years old Primigravida, with no medical comorbidities. Surgical, Drug & Family history were unremarkable. She was a house wife from Alhamra village, North Kordofan, Sudan. She was married to her first degree cousin for 1 year. Her last menstrual period (LMP) was on 15/04/2018, regular cycle and expected date of delivery (EDD) on 22/01/2019. She presented in her first antenatal visit at 20 weeks of gestation from North Kordofan, complaining of exaggerated pregnancy symptoms in term of nausea, vomiting and vague abdominal pain, these recently developing symptoms were not relieved by any medications or measures she used in her home village. She did not have other complains. The first trimester passed uneventful, she denied any symptoms of viral infection or fever, she was not exposed to radiation or medications, and she was not taking folic acid or tonics in this pregnancy, no previous history of blood transfusion, and she is not a smoker nor alcoholic. The lady is from a low socioeconomic background, and doesn`t have a history of contact with animals. Gynecological history & Sexual history were unremarkable.

On examination the patient was in good condition, weighs 59 kgs. Her vital signs were normal, BP= 110/70, not pale or jaundiced. The abdomen was soft, non-tender, no guarding or rigidity or palpable masses. The fundal height corresponding to 20 weeks.

No lower limb edema.

Investigations: Hb= 12.5 gm/dl. Platelets= 250 x 10,000. WBC= 6.1 x 10,000.

Blood Group= A Rh +ve, no Atypical antibodies.

Urine General: clear, no Ketones or signs of infection or blood.

FBS= 92 gm/dl. HbA1c= 5.2. TORCH Screening was negative. The patient was Rubella immune. Hepatitis B, Syphilis & HIV Screening were Negative.

Liver and Renal function tests were within normal limits. Parvo-virus test was negative.

Ultrasound examination showed a picture suggestive of Hydrops fetalis.

She was referred by her treating consultant to Ian Donald School for second Opinion.

Ultrasound examination performed in Ian Donald School revealed a single, viable, cephalic fetus at 20+3 weeks of gestation, with a posterior high placenta and average liquor, DVP= 3.69 cm, EFW= 625 gm.

Anomaly scan:

The CNS & CVS examination were unremarkable.

There was a massive fetal hydrops, with skin edema > 19 mm, massive ascites, Pleural effusion with lung hypoplasia, Echogenic bowel, Scrotal hydrocele, hypertrophic placenta, placental thickness= 7.9 cm. The middle cerebral artery pulsatility index (PI) and resistance index (RI) were normal.

Impression: picture of massive Non immune Hydrops Fetalis, with poor prognosis.

Amniocentesis for Karyotyping was not done, as the karyotyping is not available and the sample needs to be sent outside the country, and the family couldn`t afford it.

The patient was referred to Omdurman Maternity Hospital for a consultant committee decision regarding the condition, time and mode of delivery.

Termination of pregnancy was decided by a committee of five consultants, due to the ominous prognosis. Parents were counselled thoroughly, and they opted for termination. On 17/09/2018 termination with Misoprostol was commenced, patient had a smooth vaginal delivery at 04:00 am on 18/09/2019.

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She gave birth to a male baby, weighed 600 grams with generalized body edema & bruising, no other dysmorphic features. The baby was seen by the neonatologist, & passed immediately after delivery. Autopsy & postnatal investigations of the baby were declined by the parents.

The family was counseled thoroughly about the condition, with a documented report of the recommendations for the future pregnancies, they were strongly advised for early booking in the next pregnancies with full investigations as early as possible, and to take her previous notes to be discussed earlier for best outcomes. Close antenatal care was recommended as well as starting Folic acid tablets before and during pregnancy. Detailed early scan between 8-13+6 weeks, for early detection of abnormalities in the first trimester to allow for proper counselling and intervention if needed. Detailed anomaly scan between 18 and 22 weeks of gestation with experienced obstetrician. In case of congenital anomalies, to be referred to a tertiary center. Multi-Disciplinary team (MDT) opinion in case of congenital malformations for appropriate treatment and counselling.

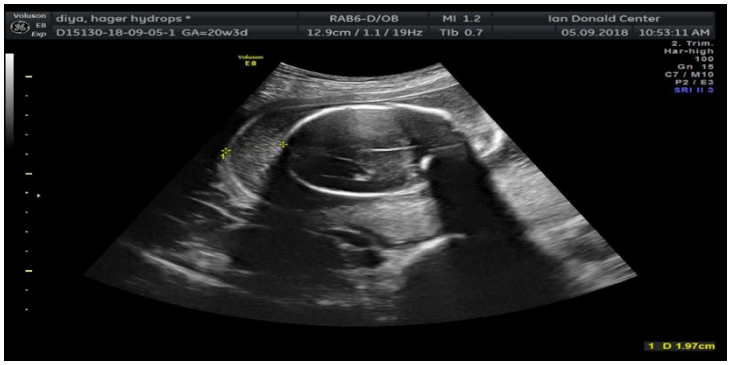

Fig. 1. Axial view showing Skin Thickness around the Skull = 19.7 mm.

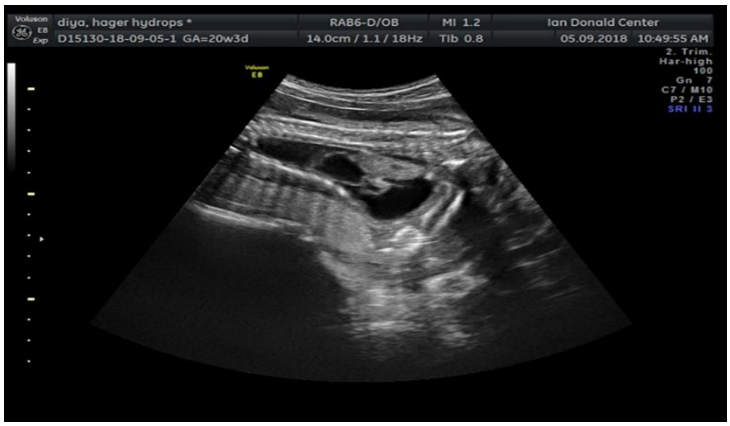

Fig. 2. Sagittal view of the spine, notice the massive ascites and pleural effusion.

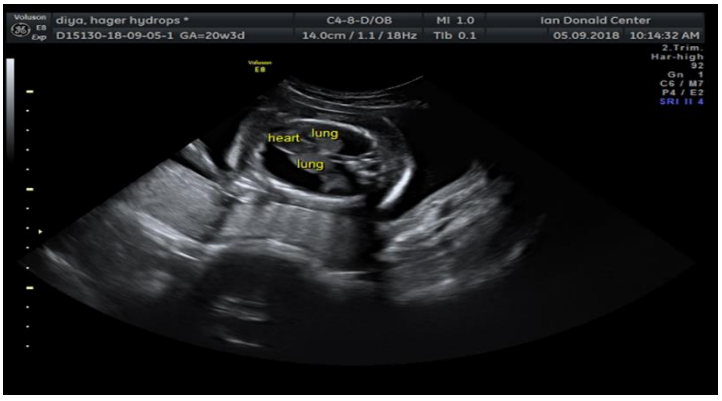

Fig. 3. Transverse view of the chest showing the heart, the hypoplastic lungs, pleural effusion & skin edema.

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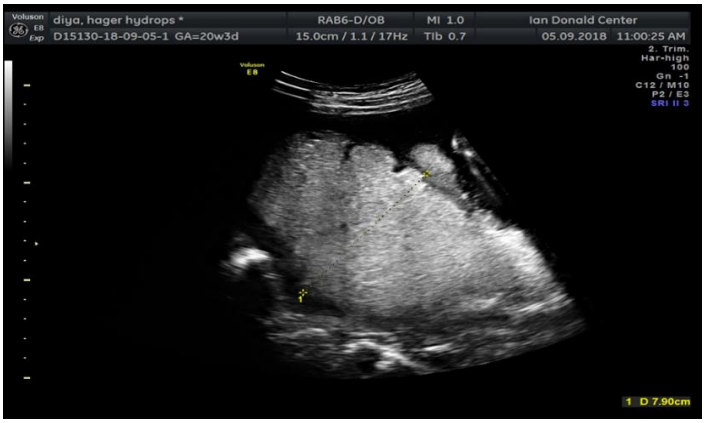

Fig. 4. Enlarged placenta, placental thickness= 7.9 cm.

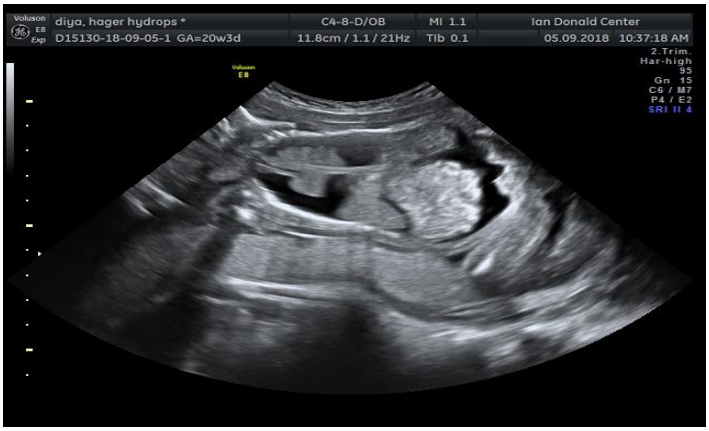

Fig. 5. Sagittal section of the body showing the massive ascites and pleural effusion, the heart and the hypoplastic lungs, the liver and the echogenic bowel.

Fig. 6. Postnatal image showing generalized Edema.

3. DISCUSSION

Fetal hydrops is defined as the abnormal accumulation of serous fluids in two or more fetal compartments. This may be pleural or pericardial effusions, ascites, skin edema (skin thickness >5 mm), polyhydramnios, or placental thickening (typically defined as a placental thickness ≥4 cm in the second trimester or ≥6 cm in the third trimester). [1], [2], [3].

In this case the anomaly scan showed a massive fetal hydrops, with skin edema of 19.6 mm in some areas, massive ascites, Pleural effusion with lung hypoplasia, Scrotal hydrocele, hypertrophic placenta, placental thickness= 7.9 cm, there was no Polyhydramnios. The CNS and skeletal and renal systems were normal.

The cardiovascular scan was unremarkable, which excludes the cardiac disease as an etiology in this case. The only soft marker that was found in this case was the echogenic bowel. Immune hydrops was excluded as the mother`s blood group was A positive and no atypical antibodies detected. So it is a case of non-immune hydrops for work-up.

Maternal Parvo-virus and TORCH screening were negative. Hb electrophoresis was not done as the family cannot afford it. Maternal kleihauer test was not done as there was no feto-maternal hemorrhage. No fetal anemia as both PI and RI were normal for Middle Cerebral Artery. There were no treatable causes in this patient scenario as ultrasound did not reveal fetal arrhythmias or other treatable cardiac problems, no treatable chest problems like pleural effusion or congenital cystic malformation, no twin-twin transfusion syndrome as it was a single fetus.

Fetal blood sampling or amniocentesis for karyotyping were denied by the patient and her family, in addition these diagnostic modalities were ultimately expensive and difficult to be found.

Decision for termination of pregnancy was decided and taken by a formal committee of five expert consultants after full and through explanation and counselling of the patient and her family.

The maternal general condition was stable throughout pregnancy, and no evidence of preeclampsia or mirror syndrome.

4. CONCLUSION

NIHF requires multi-disciplinary approach. Evaluation of Hydrops is by antibody screening followed by detailed sonography. Ultrasonographic identification of Hydrops fetalis and placenta can be determined in about 65-85%. Fetal chromosomal analysis and genetic molecular testing can be done to rule out any aneuploidy. Fetus with heart failure needs a multi vessel Doppler study using umbilical artery or vein, middle cerebral artery or ductus venosus. Management is guided by presence or absence of additional anomalies. The prognosis depends on etiology and gestational age.

Ultrasound is a valuable, safe, cost effective, widely available, and easily reproducible imaging tool for diagnosis of NIHF. We recommend that more studies need to be conducted in Sudan about the Non-immune Hydrops Fetalis.

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