

A Rare Case of Non-immune Hydrops Fetalis

REMA V NAIR, VELAYUDHAN NAIR, MOOKAMBIKA RV, VINU GOPINATH, MOHANDAS RAO KG

ABSTRACT

Hydrops fetalis is a condition which is characterised by presence of excess body water as extracellular accumulation in tissues and body cavities. A case of hydrops fetalis born to a physiologically and genetically normal mother has been reported here. A 24 years old lady with 22 weeks gestation was diagnosed to have hydrops fetalis during her routine scan at Department of Gynaecology. Ultrasonography showed single intrauterine gestation of 20 weeks 5 days with gross fetal ascites,

subcutaneous edema, bilateral pleural effusion and multiloculated cystic area in the neck region. However, fetal heart was active, heart rate was regular and four chamber view of heart indicated normal anatomy of heart. Termination of pregnancy was done. A dead baby with non-immune fetal hydrops was delivered. Diagnosis of a case of hydrops fetalis is made. Significance of early diagnosis of non-immune fetal hydrops is discussed in detail.

Keywords: Congenital anomalies, Cystic hygroma, Cystic lesions of placenta, Subcutaneous fluid accumulation

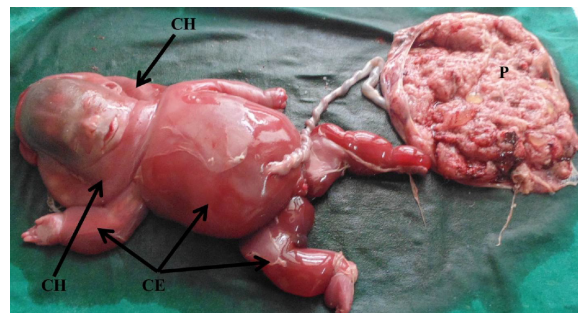
CASE REPORT

A 24 years old lady with 22 weeks gestation came to the outpatient Department of Gynaecology at Mookambika Institute of Medical Sciences, Kulasekaram, Tamil Nadu, India, for termination of pregnancy. She was diagnosed to have hydrops fetalis during her routine scan. During this pregnancy, she had no history of fever with rash, no history of medication of any kind, no reports of recurrent urinary tract infection and no complaints of uterine bleeding. Her routine first trimester scan was normal. About 14 weeks of her pregnancy, she developed pedal edema which eventually reduced. She had developed quickening at around 18 weeks of gestation, however, since last two weeks of her visit to the hospital she noticed reduced perception of fetal movements. She had no history of hypertension, diabetes, anemia, joint pain or thrombotic episodes. Her first pregnancy ended in a spontaneous complete abortion at home. She had no family history of congenital anomaly and cardiac diseases. Other observations in her included absence of pallor and pedal edema, BP was 100/60 mmHg, pulse 76/minute, her uterus was of 30 weeks size, over distended, fetal parts were not easily palpable, FHR 140/minute regular. Systemic examination was normal.

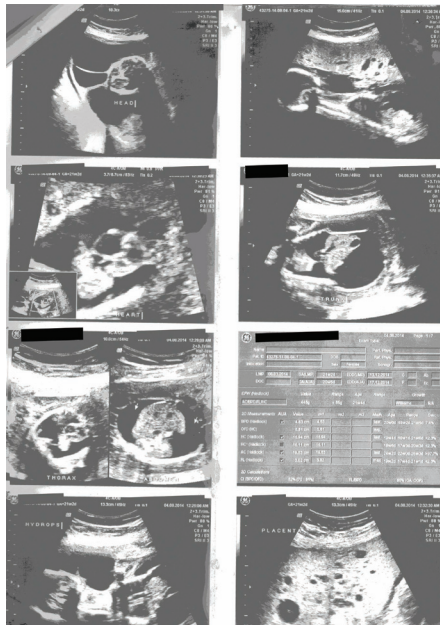
Ultrasonographic observations showed single intrauterine gestation of 20 weeks 5 days with gross fetal ascites, diffuse gross subcutaneous edema, bilateral pleural

effusion and multiloculated cystic area in the neck region of the fetus with no vascularity [Table/Fig-1,2]. However, fetal heart was active, heart rate was regular and four chamber view of heart indicated normal anatomy of heart.

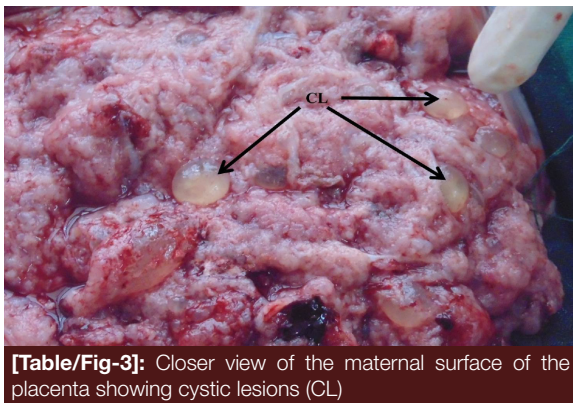
Termination of pregnancy was done after taking the written consent from the patient. A dead baby with fetal hydrops was delivered. Liquor was in excess. The baby was O+, Hb of 14gm/dL. DCT-negative Hb electrophoresis showed only Hb F and no abnormal bands. TORCH antibodies were negative. Parvo virus B 19 antigen was also negative. Karyotyping from cord



[Table/Fig-1]: Aborted fetus with placenta showing cutaneous edema (CE) all over the body of the fetus. Cystic hygroma (CH) that is multiloculated cystic area in the neck region without vascularity is also seen. P- Maternal surface of the placenta



[Table/Fig-2]: Ultrasonographic pictures of the fetus



[Table/Fig-3]: Closer view of the maternal surface of the placenta showing cystic lesions (CL)

blood was normal and showed no numerical or structural abnormalities. Gross anatomical observation of placenta showed multiple focal cystic areas within its parenchyma [Table/Fig-3].

DISCUSSION

Normal development of an individual depends on the integrated process of cellular proliferation, differentiation and growth that are influenced by various factors. Any defect at any stage of this complex process will result in abnormal development, which range from gross structural defects to anomalies at cellular and molecular levels. Several forms of defects called monstrosities produce abortions or fetal deaths.

Hydrops fetalis is one such anomaly which generally leads to abortion of the fetus.

Hydrops fetalis is a condition where the abnormal collection of fluid takes place in minimum of two different fetal compartments. It is characterised by presence of excess body water as extracellular accumulation in tissues and body cavities like pericardial and pleural cavities [1-3]. It is often associated with polyhydramnios or placental thickening. Intrauterine anaemia, intrauterine

heart failure and hypoproteinemia are primary conditions which are commonly associated with hydrops fetalis. In the present report, we are presenting a case of hydrops fetalis born to a physiologically and genetically normal mother.

Fetal hydrops are broadly classified into two types; immune and non-immune. In immune hydrops fetalis, when a pregnant woman has an immunological response to a paternally-derived antigen, maternal red cell alloimmunization takes place. These antibodies bind to antigens present on the fetal erythrocytes after crossing the placenta and cause hemolysis and hydrops fetalis [3]. Hydrops caused in the absence of maternal circulating red-cell antibodies is called non-immune hydrops fetalis. It has been observed that non-immune causes have become responsible for about 85% of cases of fetal hydrops. It is also reported to be much higher at the first and second trimester because of higher fetal death rates [3]. It can also be noted that in the present case the fetus was aborted during the second trimester.

It has been reported that cardiac etiologies like structural abnormalities of heart, cardiac arrhythmias, tumours, physiological dysfunction of heart due to infection, inflammation, infarction and arterial calcification, cardiac tumours, cardiomyopathy account for about 10 to 20% of cases of non-immune hydrops fetalis [2-4]. However, in the present case, the ultrasonographical observations of the fetal heart looked to be normal. In addition, intrauterine infections like parvovirus B19 infection, secondary anemia, fetal toxoplasmosis, syphilis, cytomegalo virus, and varicella are also considered to be the common causes of fetal hydrops (4 to 15%) [5]. In about 7% of cases, hematological disorders reported to cause non-immune hydrops fetalis [6]. In addition, certain structural congenital anomalies and single gene disorders could also be the causes of the hydrops fetalis [3]. It can be noted here that in the present case any of these commonly reported causes were not observed.

Prenatal management of hydrops fetalis; particularly after 16 to 18 weeks requires urgent referral to a maternal fetal medicine specialist [3]. The cause for hydrops is generally detected by non-invasive tests like comprehensive ultrasound examination, by fetal echocardiogram, by carefully looking for structural fetal anomalies or genetic syndromes, by signs of fetal infection, and by looking for evidence of umbilical cord or placental anomalies [3]. Invasive tests like fetal karyotyping are also suggested in cases of unexplained hydrops [3]. A retrospective study conducted in 147 cases revealed that non-immune hydrops fetalis was first diagnosed at mean gestational age of about 23 weeks. Cardiovascular causes were the most common, followed by structural abnormalities, chromosomal abnormalities and skeletal dysplasias [7]. However, in the present case, though above mentioned tests including karyotyping were conducted, all of them reported to be normal. Some of the features like

multiloculated cystic hygroma (in the neck region) associated with hydrops fetalis as seen in the present case are very rare

Generally, non-immune hydrops fetalis has high mortality rate. The option of discontinuing a pregnancy with fetal hydrops is considered since the maternal prognosis can be poor in such cases. It has been reported that the survival rate of non-immune hydrops fetalis is about 31 to 48% excluding chromosomal abnormalities [8]. Spontaneous abortion, intrauterine fetal death, or pregnancy termination of all 30 cases of non-immune hydrops fetalis diagnosed between 10 and 14 weeks of gestation has been reported [9]. When the various reports in literature on prognosis of non-immune hydrops fetalis is studied, it is evident that prognosis varies noticeably between different etiological groups. It is vital to attempt to identify the etiology for better prognosis, offer prenatal treatment available to improve postnatal outcome.

CONCLUSION

Non-immune hydrops fetalis is one of the rare congenital malformations. Though it has poor prognosis, looking for early detection tools of this condition is very essential. This might help the clinicians in handling the treatable or recurrent conditions effectively.

AUTHOR CONTRIBUTION

Dr. Reema V Nair diagnosed the case and wrote the details of the case. Prof. Velayudhan Nair and Dr.

Mookambika wrote the case. Dr. Vinu Gopinath and Dr. Mohandas Rao did the review of literature and proof reading.

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