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CASE REPORT

NON-IMMUNE HYDROPS FETALIS: A CASE REPORT

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ABSTRACT: Hydrops fetalis is a serious fetal condition characterized by abnormal accumulation of fluid in fetal soft tissues and serous cavities. There are two types of hydrop fetalis:Immune hydrops fetalis (IHF). Non-immune hydrops fetalis (NIHF). We report a case of NIHF delivered by a 18 year old primi gravida. A single female fetus 27 weeks 5 days of gestational age was delivered by caesarean section. The mother was blood group A positive. She went for regular antenatal check-up. Ultrasound at 24 weeks revealed cystic hygroma with hydrops fetalis, severe oligohydramnios and intra-uterine fetal death. On examination, the neonate was a female and grossly oedematous. She showed scalp and body wall edema including limbs edema and ascites. Radiograph of the fetus was taken and fetus was dissected to rule out anomalies in internal organs.
KEYWORDS: Hydrops fetalis, edema, intrauterine fetal death.

INTRODUCTION: Hydrops foetalis is a serious foetal condition defined as the presence of excess fluid in more than one body compartment. The fluid should be present in any of the following two cavities to be diagnosed as hydrops foetalis e.g., subcutaneous tissue, lung, abdominal cavity and pericardial cavity. The incidence of nonimmunological fetal hydrops is between 1 in 1500 and 1 in 4000.[(1)] In Southeast Asia it is more common and its incidence varies from one in 500 to one in 1500.[(2)] There are two types of hydrop fetalis:

1. Immune hydrops fetalis (IHF).
2. Non-immune hydrops fetalis (NIHF).

NIHF was first described by Edith Potter in 1943 as "universal edema unassociated with erythroblastosis". With the use of anti D prophylaxis immunological causes account for less than 20% cases.[(1)] More common are non-immune causes of hydrops foetalis.[(3)] The common cause of non-immune hydrops foetalis in order of decreasing frequency are vascular (20%), chromosomal (16%), haematological (10%), placental (8%), and a miscellaneous group.[(4,5)] Hydrops foetalis due to maternal cause is rare and is usually due to infection or diabetes mellitus in the mother.[(5)] Trisomy 13, trisomy 18, Trisomy 21 and Turner syndrome may also cause cystic hygroma or fetal hydrops.[(6)] Nonimmune-mediated hydrops can be caused by hemoglobinopathies.[(7)]

Intrauterine infection accounts only 8% of non-immune hydrops foetalis. Non-immune hydrops foetalis is mainly due to foetal causes[(8)] which includes sacrococcygeal teratoma, fetal adrenal neuroblastoma, thoracic malformations and the conditions associated with NIHF includes cardiomyopathy, coarctation of aorta, hypoplastic left heart, heart block, noonan syndrome, turner syndrome, parvovirus, CMV infections, mediastinal tumour, diagphragmatic hernia, fetal sacrococcygeal teratoma, fetal adrenal neuroblastoma, placental tumors, cystic hygromas, congenital nephrotic syndrome etc. A postmortem evaluation should be performed in all cases of hydrops that result in neonatal death.[(9)]

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CASE REPORT: A single female fetus 27 weeks 5 days of gestational age was delivered by caesarean section on 14th march 2013 at 4:15pm in the department of Obstetrics and Gynaecology, RIMS, Imphal. The fetus was collected with the permission of Medical Superintendant RIMS Hospital, Head of Department Obstetrics and Gynecology RIMS, Imphal and informs consent from the concerned parents. The study was carried out with permission of Institutional Ethics Committee, RIMS, Imphal. Full confidentiality of the individuals was maintained. The mother was an 18 year old primigravida with blood group A positive. She went for regular antenatal check-up. She has no history indicative of TB, jaundice, malaria, HIV, hypertension or heart diseases. She has no surgical history. Both her parents are diabetic. She has normal menstrual history. Ultrasound at 24 weeks showed evidence of fetal movement and fetal heart rate of 149/min. It also showed evidence of large approximately 110x76 mm lobulated and multiseptated/ multilobulated cystic lesion in the posterior aspect of the skull and the back and both sides of the neck extending upto the thorax suggestive of cystic hygroma. There were fetal ascites and bilateral pleural effusion. There was marked subcutaneous edema of the fetus. This was suggestive of single fetus with a large multiseptated cystic lesion in the head and neck region with fetal ascites, bilateral pleural effusion, anasarca and oligohydramnios.

On examination, the mother showed no signs of anemia, jaundice or oedema. Per abdomen examination showed 34 weeks size with cephalic presentation and fetal heart sound was not audible by stethoscope. Ultrasound at 27 weeks showed single fetus in cephalic presentation with no recordable heart sound. Ascites, bilateral pleural effusion and extensive subcutaneous edema were noted. A cystic lesion with multiple septation and measuring about 14.1cm x 8.6cm x 6.2 cm was seen around the posterior and lateral aspect of the fetal neck. It suggests cystic hygroma with hydrop fetalis, severe oligohydramnios and intrauterine fetal death. Maternal investigation report showed normal complete haemogramme, sugar, LFT, KFT and normal urine.

Caesarean section was done after failed induction of labour. At birth, the baby was macerated with hydrop fetalis. On examination, the neonate was a female and grossly oedematous. She showed scalp and body wall edema including limbs edema and ascites. On post-mortem examination of the fetus, the heart was found to be very small and there was comparative hypertrophy of the right ventricle. Bilateral hypoplasia of lungs was found. No gross sacrococcygeal teratoma or any other tumour was detected. This foetus was a case of NIHF. However, chromosomal analysis could not perform in this case.

Fig 1: Hydrops fetalis phenotype

Fig 2: X-ray showing Generalized oedema

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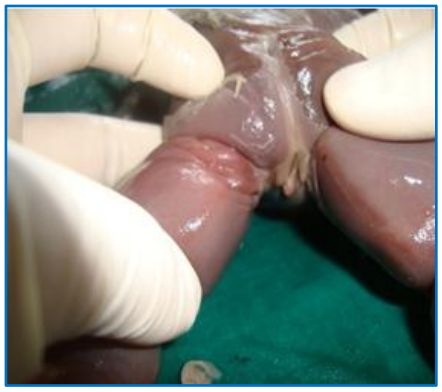

Fig. 3: External genitalia of a female fetus with hydrops fetalis

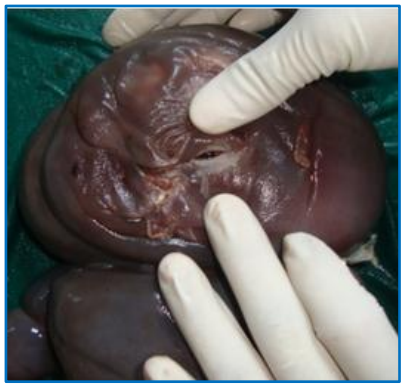

Fig. 4: Hydrop fetalis

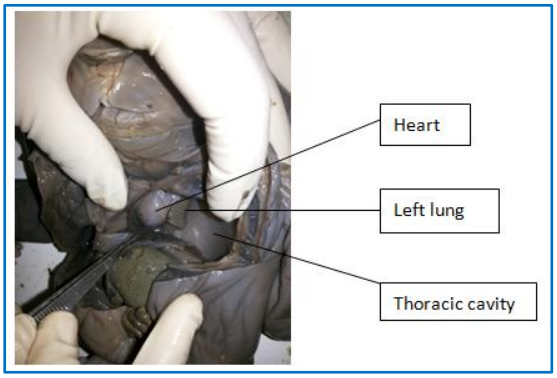

Fig. 5: Hypoplasic lung (L)

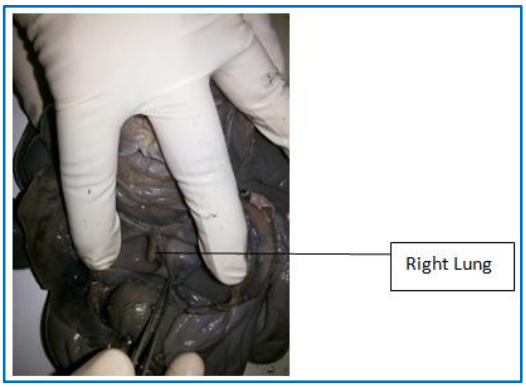

Fig. 6: Hypoplasic right lung

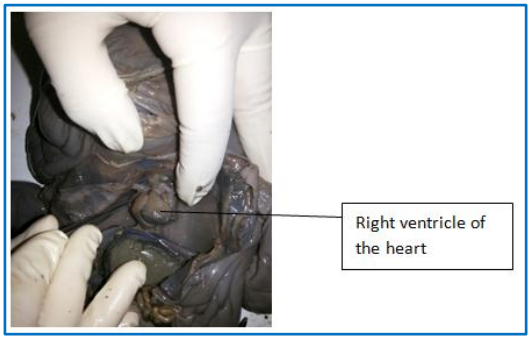

Fig. 7: A very small heart with comparatively large right ventricle

DISCUSSION: The relative incidence of hydrops foetalis 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.[8] So, the incidence of NIHF is more than IHF. The maternal risk factors for the development of non-immune hydrops foetalis include polyhydramnios (50%), hypertension, similar

illness in previous pregnancy, ethnic background particularly for alpha thalassaemia, placentomegaly, chronic maternal illness like anaemia, glucose-6 phosphate deficiency, diabetes mellitus, infections and drug ingestion. The plausible explanation of the pathogenesis of NIHF due to maternal infection is severe hepatitis leading to hypoalbuminaemia and portal hypertension. Besides, the bone marrow may be suppressed resulting in anaemia and congestive cardiac failure. The incidence of NIHF in fetal supraventricular tachycardia is between 35-60%.[10] Antenatal ultrasound scan permits the detection, monitoring and often helps to determine the cause of hydrops foetalis.[11] Advances in ultrasonography (pulsed Doppler, colour Doppler, power Doppler) have made the diagnosis of these defects possible early in gestation.[12] Non-immune hydrops can occur at any time during pregnancy. 33% of the hydrops due to paravovirus infection resolve themselves without treatment.[13]

In general NIHF is regarded to have poor prognosis and usually termination of pregnancy is advisable.[14] The prognosis is even poorer if hydrops is diagnosed in the first half of pregnancy. There is no standard and effective treatment for NIHF still now. The mortality rate of NIHF varies from 50 to 95% and the prognosis depends on the aetiology of the condition.[8] After excluding maternal causes, the etiology of this case may be due to chromosomal aberrations associated with the development of lungs and heart. Cystic hygroma might be excluded because of the generalized feature of hydrop foetalis However, chromosomal aberrations couldn't be confirmed as chromosomal analysis was not performed. In such type of cases, it is required to counsel the couple that HF could not be preventable and hydrops baby usually cannot continue life except some correctable conditions e.g., congenital heart disease and some cases of diaphragmatic hernia.

CONCLUSION: When first described, NIHF constituted 20% cases, but with effective anti D prophylaxis for immune hydrops, NIHF constitutes 90% cases of fetal hydrops. Perinatal mortality in NIHF rises to 50-98%, so only early recognition and perinatal approach can improve already bad prognosis.

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