NON IMMUNE HYDROPS – AN ENIGMA

ABSTRACT

A hydropic fetus without maternal Rh isoimmunisation is indeed a rare phenomenon . Prognosis of a future pregnancy also poses a challenge to the obstetrician. We present a case of non immune hydrops fetalis in an Rh negative non immunised mother and the challenges faced in coming to a diagnosis.

INTRODUCTION

Non immune hydrops is defined as excessive extravascular accumulation of fluid in the interstitial compartment secondary to disruption of normal intravascular interstitial fluid homeostatic mechanisms. Non immune hydrops is more common than immune hydrops (64:1). Survival rate is lower ranging from 1.5- 10%

CASE REPORT

29 year old primigravida, conceived with assisted reproduction, was referred with an ultrasound report of hydropic fetus at 29.2 weeks of gestation. Her haemoglobin was 9.4 gm% and hemoglobin electrophoresis showed no evidence of hemoglobinopathy. Her blood group was O negative, husbands blood group was O positive, Indirect coombs test was negative and Rh Titres (Direct and Indirect) – Nil. Her glucose tolerance test was within normal limits. Her TORCH titres and double stranded DNA were negative. Her Antinuclear Antibody was weak positive for which a hematology reference was done and nil active management was advised. Colour doppler study at 30 weeks gestation revealed single viable fetus in longitudinal lie, breech presentation, amniotic fluid index of 12 cm, with an estimated weight of 2.5 kg with a composite gestational age of 33.5 weeks (falsely increased due to the hydrops) with scalp oedema, ascites, pericardial effusion and cardiomegaly with middle cerebral artery – peak systolic velocity in zone A suggesting fetal anemia (Fig 1 & 2). Steroids and tocolysis were given. Cordocentesis performed and blood sent for direct comes test was negative, blood group was O positive, karyotype was normal, TORCH PCR was negative and parvovirus B 19 was negative.

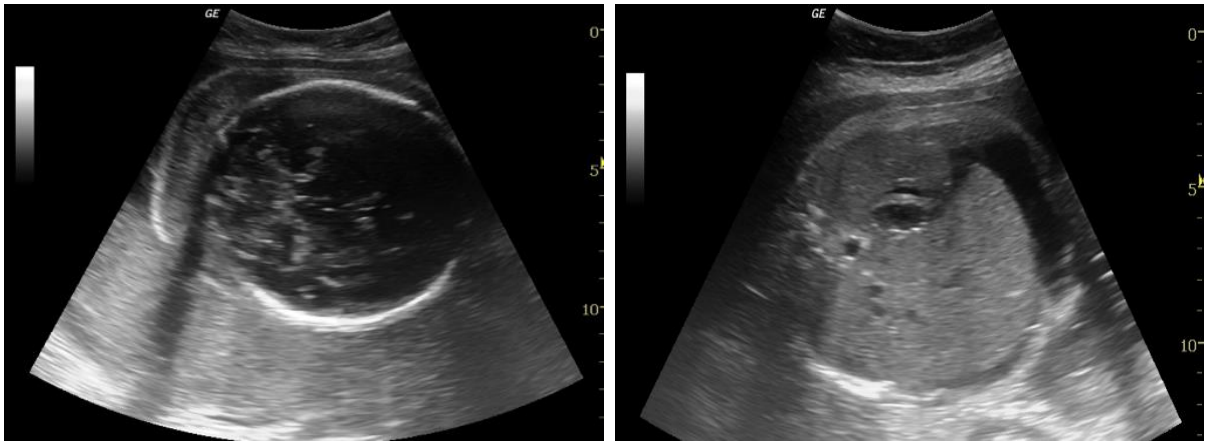
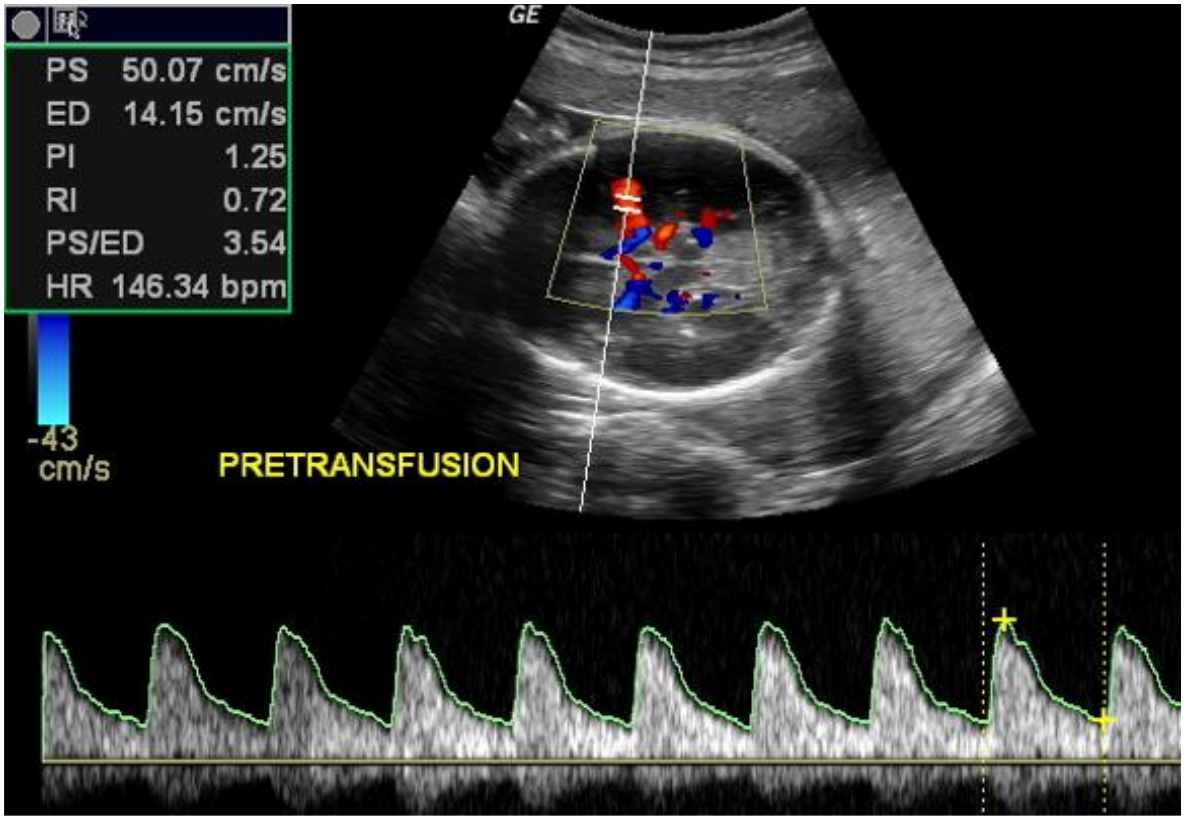


Figure 2 :Ascites, Hepatomegaly, Splenomegaly

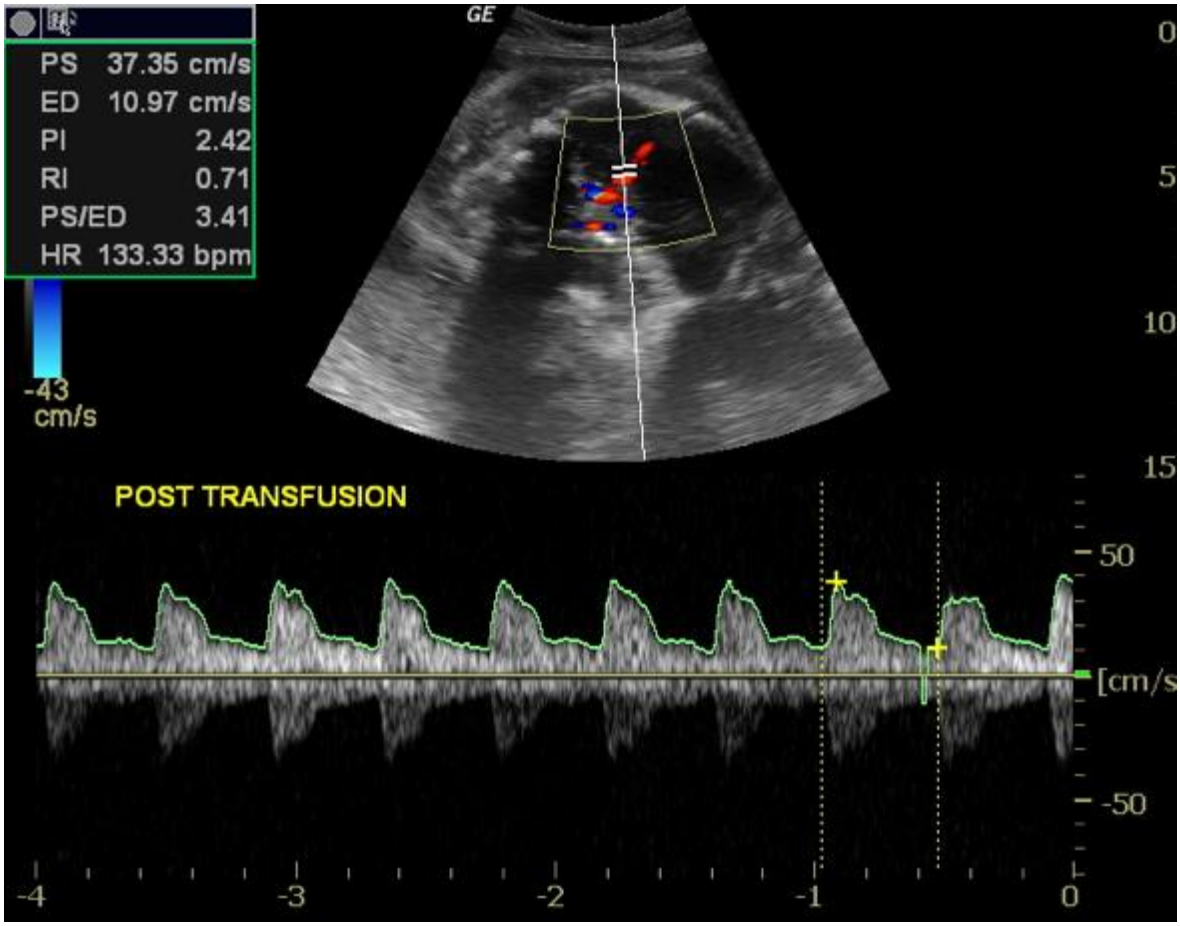
Figure 1 Scalp oedema

Intrauterine transfusion was performed where 90 ml of fresh O negative double packed blood transfused into the umbilical vein.

	HEMOGLOBIN	HEMATOCRIT	MCA-PSV
DONOR BLOOD	24.2	75.7	
CORD BLOOD	7.3	22.8	50 CM/SEC



Colour Doppler two days post procedure showed an middle cerebral artery – peak systolic velocity in zone B (37.3 cm/sec) with an impression of resolving hydrops. The next day patient complained of abdominal discomfort. On general examination mother had mild anemia with a pulse rate of 92/min and blood pressure of 120/80 mm Hg. On per abdomen examination uterus was 32 weeks with breech presentation with fetal heart rate dropping to 60 beats per minute. Ultrasound revealed single viable breech with increased liquor (21 cm) with an estimated weight of 2.7 kg with no retroplacental collection with middle cerebral artery – peak systolic velocity in zone A. An emergency lower segment caesarean section was done at 31.5 weeks of gestation. Intraoperatively liquor was brownish tinged mixed with blood. She delivered a male child, 2.6 kg with APGAR score of 5/10, 6/10. The neonate did not cry immediately after birth and had to be intubated. Heart rate was 180 beats per minute and respiratory rate was 40 beats per minute with shallow breathing and jerky respiration. Neonate was put on ventilator, kept nil by mouth on intravenous fluids and intravenous antibiotics. Cord blood showed haemoglobin of 8 gm%, O positive blood group and bilirubin of 5mg/dl. An exchange transfusion was performed. Neonatal death occurred within six hours of life due to respiratory distress syndrome. Lactation suppression and injection Anti - D 300 microgram was given to the mother and she had an uneventful recovery.



DISCUSSION

The relative incidence of hydrops fetalis has changed dramatically in the past 20 years due to prevention of immune related hydrops fetalis secondary to Rhesus isoimmunisation by Rh anti D prophylaxis. Maternal causes of non immune hydrops are idiopathic, alpha thalassemia, TORCHS infection, twin pregnancies, thyrotoxicosis, diabetes mellitus, preeclampsia, anemia, hypoprotenemia. Fetal causes are cardiovascular diseases leading to low or high output cardiac failure, chromosomal abnormalities, thoracic masses, intrauterine Infections, renal malformations, placental abnormalities, metabolic conditions[1]. Lysosomal storage disorders must be considered when dealing with recurrent hydrops. These include mucopolysaccharidosis 7, Niemann–Pick disease, galactosialidosis, mucolipidosis & type 2 gaucher disease. Mechanism by which these lysosomal storage disorders present as non-immune hydrops is still unknown[2,3]. Other rare cause of recurrent fatal fetal hydrops is nucleotide substitution in the erythrocyte Beta spectrin gene[4]. In utero treatment includes tertiary care management, intraperitoneal and intrauterine transfusions, fetal thoracocentesis or pericardiocentesis, transplacental drug therapy for fetal dysrhythmias and treatment of polyhydramnios. The management at birth includes aggressive resuscitation, appropriate fluid therapy, diuresis, dialysis, treatment of cardiac failure, partial or total exchange transfusions[1,5]. Though rare, obstetricians must keep in mind the possibility of a neonate with non immune hydrops fetalis in a Rhesus non immunised mother. It is also important to screen for metabolic disorders when other common etiologies have been excluded.

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