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A Case of Resolving Non-Immune Hydrops

S. H. Dodampahala[1], Dr. M. S. Eranda Karunadasa[2]

[1]MBBS,MRCOG, MD,FRCSE, MISUOG, Consultant Obstetrician and Gynecologist, Professorial unit, De Soyza Hospital for Women, National Hospital of Sri Lanka, Associate Professor, Obstetrics and Gynecology Department, Faculty of Medicine, University of Colombo, Sri Lanka

[2]MBBS, Research associate, Obstetrics and Gynecology Department, Faculty of Medicine, University of Colombo, Sri Lanka

Abstract: ***Mrs RP a 36 year mother presented in her second pregnancy at 12 weeks of gestation. Her previous delivery was uncomplicated and baby weighed 3.3kg. The booking oral glucose tolerance test was abnormal showing impaired glucose tolerance. She was controlled with the diet. She was immune to Rubella. A booking ultrasound scan confirmed correct dates with a nuchal translucency of 2.4 mm. The anomaly scan at a POA of 23 weeks revealed fetal ascites, pleural effusion and pericardial effusion. There were no fetal structural abnormality. Her blood group was B(+)ve, husband was AB(+)ve and there was no history suggestive of maternal viral infection, haemoglobinopathy, chronic disorders or medication in the mother. The TORCH screen did not reveal any intrauterine infection. Hydrops progressed with evidence of skin oedema at 28 weeks. Fetal anaemia was suspected as there was increased peak systolic velocity in the middle cerebral vessels. A cordoscentesis was performed at a POA of 28 weeks. The fluid bilirubin, insulin levels and cytology was normal (46 XX karyotype). It showed Hb of 12.8 g/dl which was slightly below the normal. The platelet count was normal. At 33 week scan showed resolution of the edema and hydrops. Her fetal middle cerebral Doppler showed normal velocities. Her pregnancy was continued up to 38 weeks and the 3.8 Kg weight baby was delivered by an elective caesarean section. The recovery was uneventful. Discussion: This is a case of non-immune hydrops. Non immune hydrops is defined as the presence of excess extra-cellular fluid in two or more sites without any identifiable circulating antibody to red cell antigens. Generalized skin thickening of more than 5mm and or two or more of the following Pericardial effusion, Pleural Effusion, Ascites and Placental Enlargement. The incidence of 5 to 8 per 10000 represents the figure of live born hydropic neonates admitted in neonatology units. Actual incidence is much higher, since majority die either in-utero or the pregnancies are terminated electively. Most of the causes have been excluded in our series by performing detailed Ultrasound Anomaly Scan and Fetal Echocardiography, Doppler Blood Flow Studies, Liquor Volume Assessment (AFI), Placental Thickness & Echogenicity. However Parvo virus B19 anti body states could not be assessed due to non availability of the test in Sri Lanka. Management: As a rule of thumb exclusion of major structural, genetic/metabolic and chromosomal abnormalities is required before embarking on prenatal therapy which can be transplacental or direct fetal therapy. Invasive procedures are required in determining the cause as well as in the palliative therapy. Since our patient did not have a definitive pathology and showed signs of improvement she was managed expectantly with ultrasound supervision. Mother was explained and counseled about the outcome, prognosis, risk of invasive procedure and the rationale of our management. Conclusion: Our case can be considered as a case of idiopathic hydrops which has settled in the third trimester managed expectantly. A detailed accurate assessment and follow-up by ultrasound scanning and invasive blood sampling which has given all the strength to manage and counsel the patient expectantly. Successful therapy can be administered in some cases, yet majority are associated with a poor prognosis.***

Keywords: Non-Immune Hydrops, pregnancy, Gynecology

1. A Case of Resolving Non-Immune Hydrops

Mrs RP a 36 year old mother presented in her second pregnancy with a previous uncomplicated vaginal delivery with a 3.3 kg baby. She presented to university obstetrics and gynaecology unit, Colombo at 12 weeks of gestation. An oral glucose tolerance test was performed and which detected an impaired glucose tolerance. She was controlled with the diet. Index pregnancy was a planned pregnancy and she had not used pre-conceptional folic acid. She was immune to Rubella. A booking ultrasound scan did not reveal any abnormality and the nuchal translucency was 2.4 mm which was slightly increased, but within normal limits. She attended the antenatal clinic regularly and received appropriate care.

Her routine anomally scan preformed at a POA of 23 weeks revealed collection of the fluid in the peritoneal and pleural cavities. The heart appeared anatomically normal with a pericardial effusion. The detailed anomaly scan did not reveal any fetal structural abnormality. Her blood group was B(+)ve, husband was AB(+)ve and there was no history suggestive of maternal viral infection in the first trimester. There was no history of haemoglobinopathy, chronic disorders or medication in the mother. The blood sugar estimations were normal and TORCH screen did not reveal any intrauterine infection.

Hydrops progressed with evidence of skin oedema at 28 weeks. Fetal anaemia was suspected as there was increased peak systolic velocity in the middle cerebral vessels. A cordocentesis was performed at a POA of 28 weeks although it was not the optimum timing for it. The amniotic fluid collected during the procedure was sent for viral studies, insulin levels, cytology. The fluid bilirubin, insulin levels and cytology was normal. The cordocentesis analysis showed Hb of 12.8 g/dl which was slightly below the normal. The platelet count was normal. Chromosomal analysis of the cord blood showed a 46 XX karyotype. Sex determination by ultrasound confirm a female fetus.

At 33 week scan showed resolution of the edema and hydrops. Her fetal middle cerebral Doppler showed normal velocities. The severity of hydrops became less after 34 weeks. Her pregnancy was continued up to 38 weeks and the baby was delivered by an elective caesarean section. The recovery was uneventful and the birth weight was 3.8 Kg.The neonatal assessment showed marked edema of the

lower limbs and low set ears. There were no other external or internal abnormalities as detected by neonatal examination, ultrasonography and 2 D Echocardiography.

2. Discussion

This is a case of non-immune hydrops. Non immune hydrops is defined as the presence of excess extra-cellular fluid in two or more sites without any identifiable circulating antibody to red cell antigens. Generalized skin thickening of more than 5mm and or two or more of the following Pericardial effusion, Pleural Effusion, Ascites , Placental Enlargement. The incidence of 5 to 8 per 10000 represents the figure of live born hydropic neonates admitted in neonatology units. Actual incidence is much higher, since majority die either in-utero or the pregnancies are terminated electively.

It is an end result of an array of disorders of the fetus, umbilical cord and placenta that leads to deranged fluid homeostasis. A wide range of fetal organs are involved - No common mechanism is responsible for the signs of hydrops.

The commonest causes of hydrops like:

1. Anemia, eg.Alpha (α) Thalasaemia, Secondary to Feto-maternal Hemorrhage, Twin-twin transfusion, Other Hemoglobinopathies
2. Cardiac Failure eg. Disorders of cardiac function / Structure include Cardiomyopathies, Tachyarythmias, Bradycardias (Congenital heart block), Obstructive left heart disease, Ebstien's anomaly, Atrial isomerism
3. Reduction in Osmotic Pressure (Hypoproteinaemia).
4. Obstruction to venous return, Congenital cystic adenomatoid malformation of lung
5. Impaired lymphatic drainage, Cystic hygroma, Karyotypic abnormalities (45XO), Connective tissue malformation
6. Fetal Infections, eg. TORCH, Parvo Virus B19
7. Genetic disorders/Metabolic disorders - Gaucher disease, Hurler disease, hypothyroidism, hyperthyroidism, mucolipidosis, and mucopolysaccharidosis
8. Skeletal dysplasias - Achondrogenesis, achondroplasia, asphyxiating thoracic dystrophy, lethal osteoporosis, Noonan syndrome, short rib–polydactyly syndrome, and thanatophoric dysplasia
9. Fetal hypokinesis - Arthrogryposis, congenital myotonic dystrophy, Neu-Laxova syndrome, and Pena Shokeir syndrome
10. Idiopathic disorders - Recurrent isolated hydrops
11. Maternal disorders - Graves disease, severe anemia, severe diabetes mellitus, and severe hypoproteinemia
12. Placental disorders - Chorioangioma, chorionic vein thrombosis, cord torsion (knot or tumor), umbilical artery aneurysm, and venous thrombosis

Most of the above causes have been excluded in our series by performing detailed Ultrasound Anomaly Scan and Fetal Echocardiography, Doppler Blood Flow Studies, Liquor Volume Assessment (AFI), Placental Thickness & Echogenicity.

We have obtained a Fetal Blood Sample from placental Insertion of Umbilical Cord (Procedure related fetal loss is 25% in hydropic fetus) for Full Blood Counts, Karyotype, Blood Gas Analysis, Virology screening, Serum Protien Evaluation and these investigation did not show any abnormality except mild fetal anaemia with a Hb of 12.8 g/dl. However Parvo virus B19 anti body states could not be assessed due to non availability of the test in Sri Lanka. Generally Parvo virus infection of the fetus causes severe bone marrow suppression hence pancytopenia showing marked reduction in all cell lines. This feature was not seen in our fetal blood sample.

Management Options: As a rule of thumb exclusion of major structural, genetic/metabolic and chromosomal abnormalities is required before embarking on prenatal therapy which can be transplacental or direct fetal therapy. Invasive procedures are required in determining the cause as well as in the palliative therapy. Since our patient did not have a definitive pathology and showed signs of improvement she was managed expectantly with ultrasound supervision. At all times mother was explained and counseled about the outcome, prognosis, risk of invasive procedure and the rationale of our management.

The common associated maternal complications like polyhydramnios, pre-mature labour, pre-eclampsia oligohydramnios, problems with third stage, morbidly adherent placenta was not seen in our patient.

- Obstetric management
- Rarely straightforward
- Temptation to deliver the sick fetus before term should be avoided
- Common maternal complications should also be considered like associated polyhydramnios.
- Amniocentesis before delivery makes the delivery easy and less traumatic for both mother & Fetus, and facilitates resuscitation
- Most of the times such fetuses are delivered with elective cesarean section.
- No evidence that mode of delivery has a marked effect on outcome.

Pediatric team was informed about the babies condition a week prior to the delivery and necessary fetal monitoring was done usinf fetal movement chart, CTG, Bio-physical profile. During the delivery by elective caesarian section the pediatric team was kept ready as resuscitation is often difficult and adequate senior assistance must be available. Even though it has been documented that high incidence of third stage complications are recognized we have not encountered any third stage complication due to large but normal placenta and active management of the third stage.

Prognosis in hydrops is generally very poor with very high peri-natal mortality but in the presence of idiopathic and cardiac arrhythmias perinatal mortality can be reduced with close observation and fetal therapy.

3. Conclusion

Our case can be considered as a case of idiopathic hydrops which has settled in the third trimester managed expectantly. A detailed accurate assessment and follow-up by ultrasound scanning and invasive blood sampling which has given all the strength to manage and counsel the patient expectantly. Successful therapy can be administered in some cases, yet majority are associated with a poor prognosis. An appropriate prenatal investigations must be performed to make a correct diagnosis to identify an effected fetus in whom a good outcome may be anticipated.

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3. Dr.Basky Thilanganathan, The Director, Fetal Medicine Unit, King's college, University of London.

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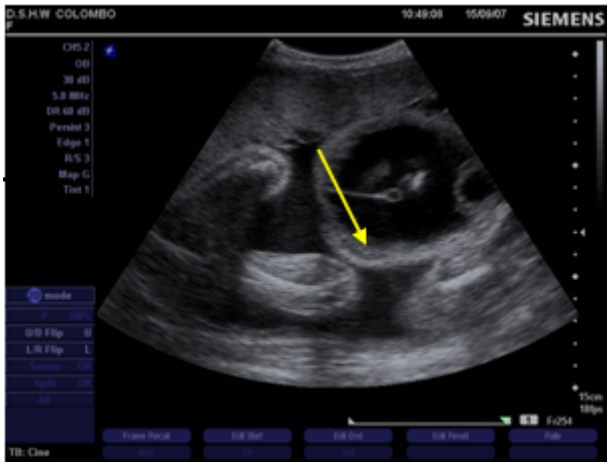

Figure 1: 23 weeks scan showing the coronal section of the fetal abdomen with ascites and falciform ligament

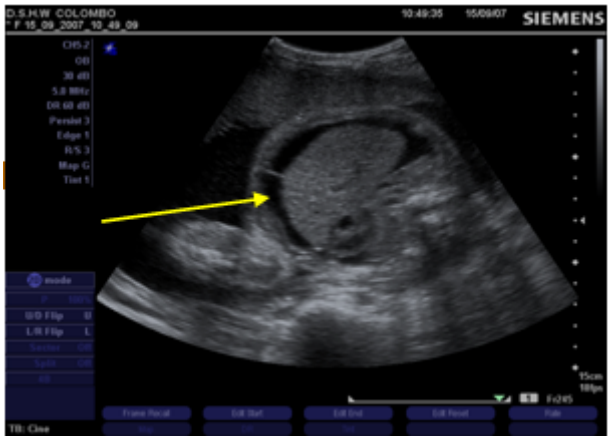

Figure 2: 23 weeks scan showing the Coronal section of the fetal abdomen with ascites and liver

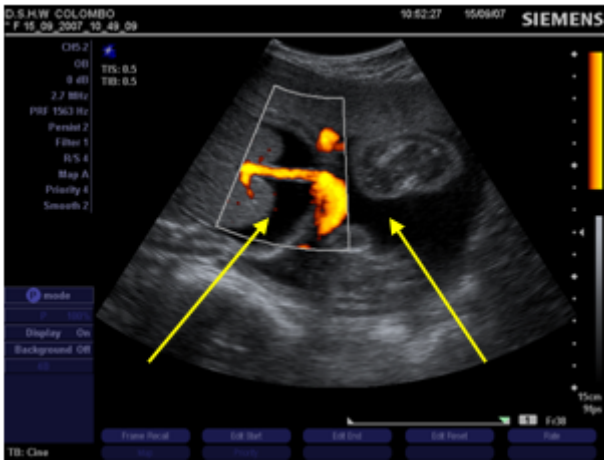

Figure 3: 23 weeks scan showing the transverse section of the fetal abdomen with ascites and hydrothorax

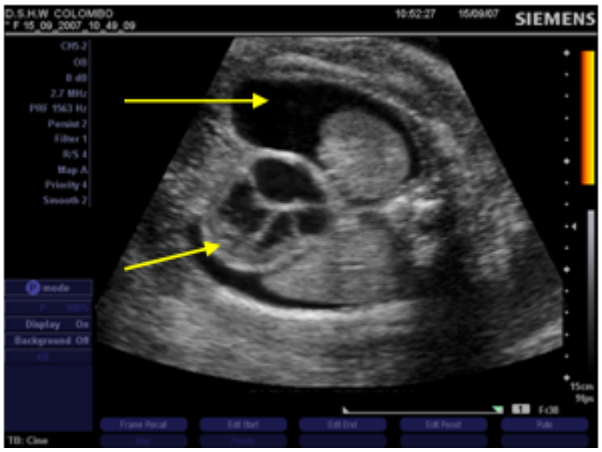

Figure 4: 23 weeks scan showing the cross section of the thorax showing hydrothorax and pericardial effusion

Figure 5: Neonate after birth receiving special baby care, note the leg oedema

Figure 6: Neonate after birth receiving special baby care, note low set ears