Section: Anatomy

Case Report

HYDROPS FETALIS WITH CYSTIC HYGROMA – A CASE REPORT

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Abstract

Background: Hydrops Fetalis (HF) is an excess accumulation of fluid in serous cavities and/or edema of soft tissues in the fetus. Depending on the severity and cause of hydrops, there may be edema of fetus and placenta, ascites, pleural effusions and/or pericardial effusions.1 Most cases of hydrops are caused by severe erythroblastosis fetalis secondary to Rh isoimmunization & is called immune hydrops fetalis (IHF). Cases of HF caused by other conditions such as Down syndrome, Noonan syndrome and Osteogenesis Imperfecta syndrome type 2 are known as Non Immune Hydrops Fetalis (NIHF).1 Here is a case of an 18 week Fetus with Non immune HF and Cystic Hygroma (CH). **Materials and Methods:** A 30 yr old primigravida was referred to MIMS, Mandya after the USG 2nd trimester revealed diagnosis of Cystic Hygroma with hydrops. The parents were counseled and they opted for Medical termination of pregnancy. After taking consent labour was induced & the dead female fetus was expelled and sent to anatomy dept for studying. Gross features, microscopic details of organs & microscopic structure of cystic hygroma were studied on autopsy and findings are presented. **Result:** 30 yr old primigravida was referred to MIMS, Mandya after the USG 2nd trimester revealed diagnosis of Cystic Hygroma with hydrops. The parents were counseled and they opted for Medical termination of pregnancy. After taking consent labour was induced & the dead female fetus was expelled and sent to anatomy dept for studying. Gross features, microscopic details of organs & microscopic structure of cystic hygroma were studied and findings are presented. **Discussion:** The incidence of NIHF is approximately 1 in 4000 neonates worldwide. A high mortality rate is observed in a combination of CH and NIHF which has been linked to increased incidence of fetal chromosomal anomalies. Trisomy 21 is one of the most common associated abnormality seen sonographically in the 1st trimester. In the 2nd trimester, Turner's syndrome followed by Trisomy 18 are most common unless proved otherwise according to the literature. **Conclusion:** This case report has highlighted the association between Cystic Hygroma with NIHF and need for genetic evaluation of the fetus. Karyotyping of parents and Flourescence in Situ Hybridisation (FISH) of fetal tissue to establish or exclude aneuploidy.

INTRODUCTION

Hydrops Fetalis (HF) is defined as an overabundance of fluid accumulating in serous cavities and/or fetal tissue edema. Ascites, pleural effusions, pericardial effusions, edema of the fetus and placenta, and other conditions may occur depending on the severity and etiology of hydrops.[1,2]

A thin-walled, multiseptate cystic structure with thin walls that is posterior to the fetal head and neck and is eccentrically positioned with regard to the fetus's long axis and does not have a vertebral column defect is known as a cystic hygroma.[3,4]

CASE REPORT

30yrs old primigravida with B +ve blood group at 18 weeks presented for MTP (Medical Termination of Pregnancy) due to Hydrops fetalis with Cystic Hygroma was detected on routine obstetric sonography at Mandya Institute of Medical Sciences district hospital. No contributing history of

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consanguinity or history of similar disorders in the family were noted. After taking consent from the patient, delivery was induced & fetus was handed over to our department.

A large cystic hygroma (fluid collection on the back of neck) with many loci was seen.

- There was considerable edema over the hands and ankles (puffy hands and feet) in addition to severe widespread edema.
- Fetal ascites and a B/L minor pleural effusion were seen. Widely spaced nipples.
- A fetal autopsy was performed, organs (exhibited normal histological features) and tissue from the hygroma were sent for histological analysis which showed large, irregular dilated lymphatic channels lined by attenuated bland endothelial cells with variable thick collagenous wall and lymphoid aggregates.

Though parents were counseled, and referred for further investigations like Karyotyping, FISH etc we lost the follow up of the parents as they were not available and non cooperative.

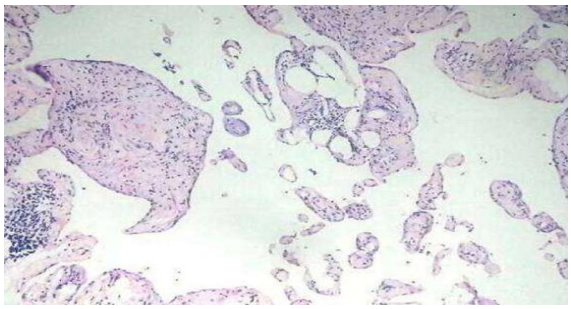

Figure 1: The microphotograph shows large, irregular dilated lymphatic channels lined by attenuated bland endothelial cells with variable thick collagenous wall and lymphoid aggregates

Figure 2: Hydrops Fetalis

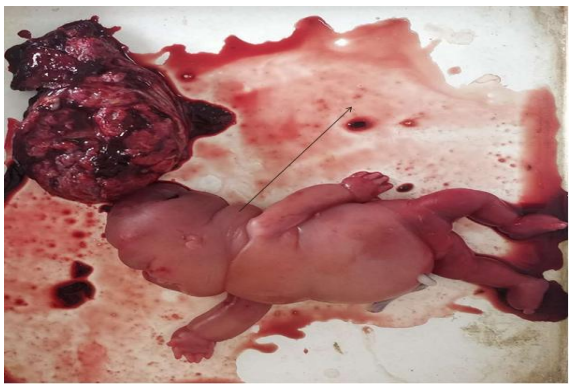

Figure 3: Cystic hygroma

DISCUSSION

Prenatal ultrasound scans, particularly in the second trimester, that show both hydrops fetalis and cystic hygroma have been linked to serious outcomes such abortion, intrauterine fetal death, and early newborn death.[2]

As there was no evidence of Rh incompatibility, NIHF was made a likely diagnosis. In newborns, the incidence is roughly 1 in 4000. It is separated into two groups: immune and non-immune. When red cell alloimmunization is present, it is referred to as immune hydrops fetalis. If not, it's referred to as non-immune hydrops fetalis. One of the most frequent anomalies detected sonographically in the first trimester is a cystic hygroma. The most common trisomies in fetuses with first-trimester cystic hygroma were 21, 18, and 13. Premature birth is the fate of nearly all fetuses with hydrops and cystic hygroma.[5] Cystic hygromas that do not have hydrops typically regress entirely.[6]

Around the sixth week of embryonic life, mesoblasts begin to develop the primitive lymph sacs; the main pair is located in the neck, between the jugular and subclavian veins.[7]

Any disease that accelerates the rate at which fluid leaves the vascular compartment or delays the lymphatic return to the circulation is the primary cause of hydrops fetalis which was evident in microscopic details.

CONCLUSION

It is likely a case of Turner syndrome in utero with characteristics such as inflated hands and feet, widely separated nipples, and fluid collection on the back of the neck during the second trimester. In the second trimester, hydrops fetalis and cystic hygroma have been linked to worse neonatal outcomes.

This case report has highlighted the association between Cystic Hygroma with hydrops and need for genetic evaluation of the fetus. Karyotyping of parents and Flourescence In Situ Hybridisation (FISH) of fetal tissue to establish or exclude aneuploidy.

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