

Non-Immune Hydrops Fetalis with Cystic Hygroma

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ABSTRACT

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Most common cause for Hydrops fetalis-HF is immune mediated i.e. Rh incompatibility. Cases of non immune Hydrops fetalis-NIHF also occur with an incidence of 1/3500 cases and the causes are manifold. Here we present a case of non immune foetal Hydrops in 20 weeks old fetus for which induction of abortion was done after Ultra sonographic evaluation and autopsy examination was done.

Keywords: Hydrops fetalis- HF, Cystic hygroma

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CASE REPORT

Ultra sonographic evaluation of a 25 year old primigravida as part of antenatal check up revealed Hydrops fetalis in a 20 week old fetus, for which induction of abortion was done. The foetus was sent for autopsy examination with a diagnosis of Non immune hydropsfetalis-NIHF with multiple anomalies.

External examination showed a female fetus with generalized oedema and swelling around the neck. It weighed 480 grams with a head circumference of 21 cm. No obvious external anomalies were noted. Skin and subcutaneous tissue markedly oedematous, multi loculated cystic spaces were seen on both sides of neck. On opening the pleural and peritoneal cavities were filled with clear fluid. The thymus was conspicuously absent. All other internal organs were normal. There were no obvious cardiac anomalies. The placenta received along measured 10x9x3cm with cord of 15cm long and both grossly and histologically appeared normal. Karyotyping done in this case revealed 47XX+16 (trisomy16). Anatomical diagnoses of hydropsfetalis with Cystic hygroma and Thymic aplasia was given

DISCUSSION

Hydropsfetalis is Latin for edema of fetus. This was described first by Ballantyn² in 1892. Hydropsfetalis means severe oedema of trunk, extremities and head with effusion in all serous cavities. The basic problem is imbalance in fluid homeostasis, which is either Immune or Nonimmune related. Until later half

of 20th century, the most common cause for this was alloimmune hemolytic disease or Rh isoimmunisation, therefore are labeled immune Hydrops fetalis(IHF). But cases of non immune Hydrops (NIHF) also occur with an incidence of 1/3500 case. In 1970 major cause for IHF was conquered with use of Immunoglobulin prophylaxis in high risk mothers.

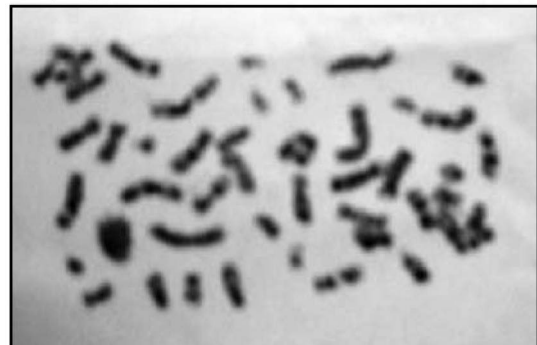


Figure 1. Karyotyping revealed 46XX +16 (Trisomy)



Figure 2. Sonogram showing fetal ascites

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Figure 3. Hydrops baby

Three major causes⁶ of NIHF includes cardiovascular defects, chromosomal anomalies and fetal anemia. Out of which the most common cause for NIHF is cardiac anomalies with primary myocardial failure, high output cardiac failure with resultant decreased colloid osmotic pressure, increased hydrostatic pressure and increased vascular permeability. Cardiac malformations, arrhythmias and cardiomyopathies are also recorded. Second most common cause is chromosomal anomalies such as Turners-45X (42%), Trisomy-21(34%) Trisomy-18(9%), other trisomies such as Trisomy 13, 15 and 162 as well as Tetraploidy. The pulmonary causes⁵ include pulmonary sequestration, Right diaphragmatic hernia, congenital cystic adenomatoid malformation and enterogenous cysts. Fetal anaemias include alpha thalassemias, Hemoglobinopathies and fetomaternal transfusion. Infections as cause include Parvo virus, Treponema pallidum and Cytomegalovirus.



Figure 4. Hydrops Baby with general edema and cystic hygroma

Sonography is very helpful in the diagnosis as it was in this case. Sonographic criterias⁷ were developed after a study of 26 cases as to anaemia related or not. Fetuses without anaemia as a cause for HF showed pleural

effusion-87%, marked edema 62.5%.A combination of pleural effusion and marked edema was evident in fetuses without anaemia in56.3% .Thickened placenta seen with anaemia associated HF.

In conclusion causal diagnosis of HF is often difficult to determine at autopsy especially if there has been prolonged fetal death. Only strictly structural causes could be identified at autopsy. Attempts to diagnose malformations and other genetic disease through investigation will be necessary for assessing risk of recurrence

END NOTE

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REFERENCES

1. Hydrops fetalis with polymalformative syndrome [Internet]. [cited 2016 Apr 29].
2. Pediatric Hydrops Fetalis: Background, Pathophysiology, Epidemiology. 2016 Mar 24 [cited 2016 Apr 29].
3. Hydrops fetalis. In: Wikipedia, the free encyclopedia [Internet]. 2016 [cited 2016 Apr 29].
4. Children's Hospital of Wisconsin [Internet]. [cited 2016 Apr 29].
5. Pediatric pathology Dehner-Vol 1,2 nd edition p 146-148,354
6. Pathological basis of disease-Robins and Cotran- 8th Ed p459-462
7. Sono graphic evaluation of HF-Saltzman DH, Frigoletto F. D etal-OB&Gyn-1989 July-74(1)106-11 .