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Fetal Cystic hygroma with hydrops fetalis: a rare case report in Somalia

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Abstract

Fetal cystic hygroma represents a condition of early lymphatic obstruction that looks like fluid-filled, membranous cysts. It can relate with 90% nonimmune hydrops fetalis and chromosomal abnormality 50–80%, and it is caused by communication failure in between jugular veins and lymphatic sacs. In Somalia there was poor antenatal care in pregnant women so you could not detect actual cases of fetal hydrops, we present rare case in Somalia a 20 weeks and four days being pregnant with hydrops fetalis, cystic hygroma and with Rh positive parents and no structural anomalies we could not suggestive any chromosomal abnormalities because there were no available tests in Somalia, she delivered normal vaginal delivery and she was well and discharged the hospital. The patient was satisfied with the overall intervention and outcome; we have written informed consent from the parents was obtained for this publication, Termination of pregnancy might be considered if there is fetal hydrops with cystic hygroma.

Introduction

Fetal cystic hygroma is a condition of early lymphatic obstruction that looks like fluid-filled, membranous cysts, lined by true epithelium in the anterolateral or occipitocervical site. It can relate with nonimmune hydrops fetalis 90% and chromosomal abnormality 50–80%, Cystic hygroma is characterized an important marker for aneuploidy generally including trisomy's but is an extremely common in Turner syndrome [1], unfortunately in Somalia we couldn't do genetic test because of lack of instruments and facilities. Subsequently the start of ultrasonography, we were seen more cases of cystic hygroma and fetal hydrops separately, but this first case was seen cystic hygroma and fetal hydrops together. Fetal hydrops and cystic hygroma can be diagnosed in the first trimester and second trimester of gestation and it has a high mortality.

The cystic hygroma could be small in size and transient or large, multiseptated and or without septation. Hydrops fetalis is a pathological condition of the excessive fetal fluid accumulation which can express as ascites, pleural effusion, and pericardial effusion and/or generalized subcutaneous edema.

The fetal cystic hygroma caused by communication failure in between the jugular veins and the jugular lymphatic sacs that develops at 40th day of gestation. If that communication between the lymphatic and the venous system does not occur will advances a progressive peripheral lymphoedema and hydrops and finally leading to early intrauterine fetal death [2].

Fetal cystic hygroma is same with cystic lymphangioma, which is also known as a macrocytic lymphatic malformation and was first pronounced in 1828 by Redenbacher. The survival rate of fetuses with cystic hygroma is only 2-6%. The incidence of cystic hygroma is likely to be 1 case per 6000-16000 live births3.

In this study, we report fetal cystic hygroma with hydrops fetalis a rare case in Somalia at 20weeks and four days live fetus were not seen before, written informed consent from the parents was obtained for this publication.

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25 years old G1P0 A1 with 20weeks and 4 days of gestation was diagnosed prenatally during ultrasonography for the fetal with a large cystic hygroma without septation that located in the cervical area and associated with bilateral pleural effusion and ascites with a variable heart rate (Figure1).

Family history revealed no specific disease or congenital malformation. There was history of consanguineous marriage, the husband is her cousin and had abortus in the last pregnancy and Rh-positive pregnancy she doesn't has any chronic diseases. Now she is pregnant 20 weeks and four days, presented to Mogadishu Somalia Turkish Training and Research Hospital (MSTERH) the due to past history of Abortus baby.

The pregnancy was terminated with the consent of the parents after explaining about the situation and poor outcome of pregnancy. The pregnancy was terminated after induction with misoprostol tablets with a maximum dose of 1200 mcg and delivered normal. Examination of fetus exposed a cystic structure extending posterolateral along the neck and it was also associated with ascites and fetal edema (Figure2) and the placenta was looked little bit large with edematous (Figure3), we were discharged.

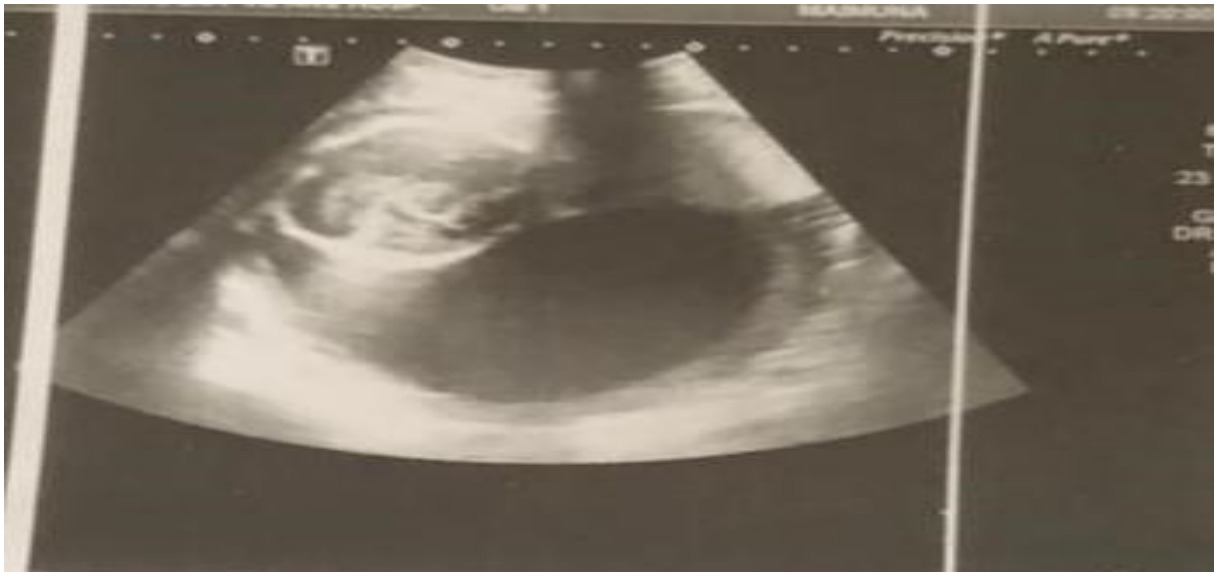

Figure 1: USG showing cystic hygroma without septate hygroma.

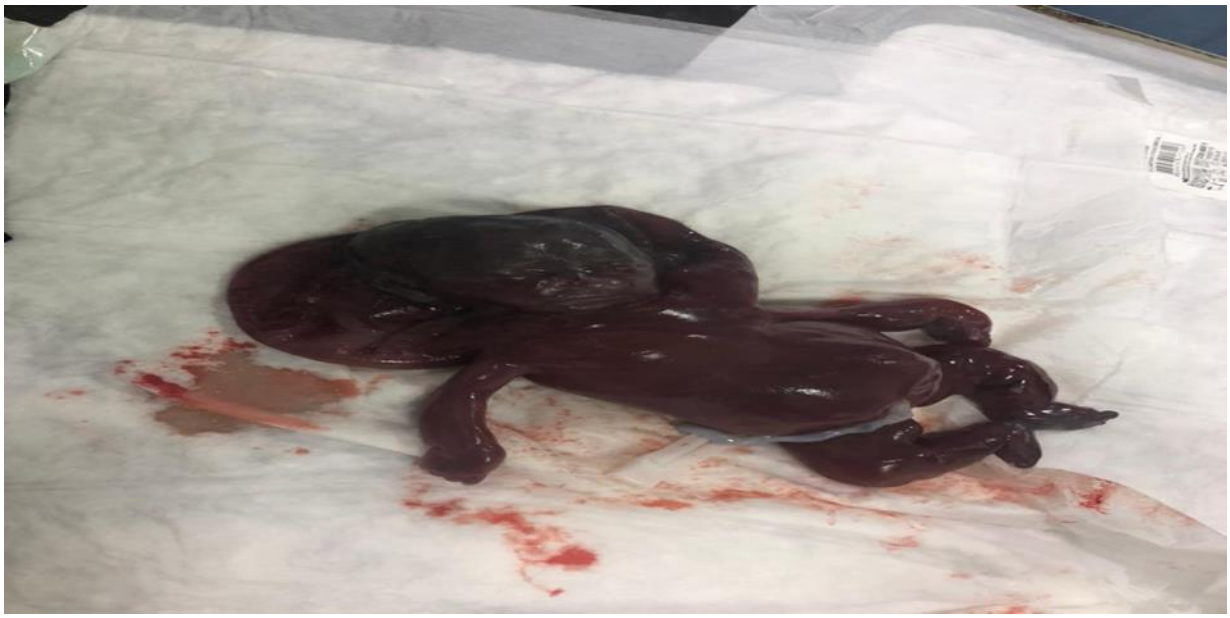

Figure 2: Death fetus with ruptured cystic hygroma with hydrops fetalis

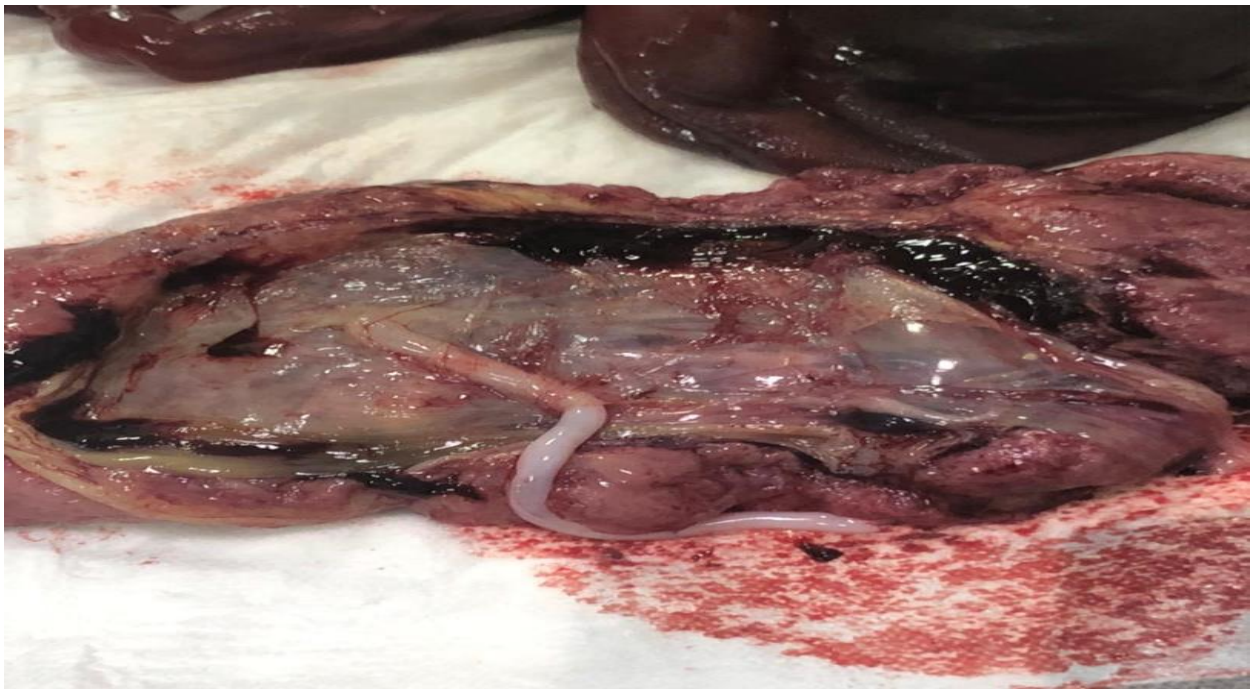

Figure 3: Large Edematous placenta with hydrops fetal.

Discussion

Hydrops fetalis refers to fluid accumulation in serous cavities and/or edema of smooth tissues withinside the fetus. It is characterized as nonimmune if there no indication of a feto-maternal blood organization incompatibility [4].

Fetal cystic hygroma is group of disorders that initiated from lymphatic obstructions, and it could congenital or develop at any time during a person's life. Almost all fetuses with cystic hygroma and hydrops die antenatally [5]. Fetal cystic hygroma is fluid filled mass with thin wall in occipitocervical area and the fetus can progress to hydrops and ultimately fetal death.

They are characterized by single or multiple congenital cysts of the lymphatic system most commonly found within the soft tissues of the neck [6]. the 80% cystic hygroma occurs in the neck that is called nuchal cystic hygroma. Other locations are axilla and 10% of cases in the mediastinum and only 1% confined to the chest.

Congenital cystic hygromas can resolve to webbed neck, edema, and low posterior hairline. In other instances, the hygroma can progress in size to become larger than the fetus. Cystic hygroma is powerfully associated with Turner's syndrome (predominantly a 45 XO karyotype), trisomy 21 (Down syndrome), trisomy 18 (Edwards syndrome), and 45 X / 46 XY (Noonan syndrome). Fetal cystic hygroma associated with NIHF has a high incidence of aneuploidy, particularly Turner syndrome or trisomy 21. About 30 to 70% of fetuses with cystic hygroma have chromosomal abnormalities [7]. So Genetic counseling are required to exclude chromosomal abnormality or not, unfortunately we don't have any genetic testing or counseling.

Conclusion

We have to do routine ultrasound during antenatal care to detect any abnormality. Fetuses with cystic hygroma and hydrops can have poor prognosis and mostly those dead after birth. Fetal death has been related to chronic fetal hypoxemia secondary to compression of the thoracic structures by the generalized edema. When hydrops is present along with cystic hygroma the mortality rate is near 100%. Termination of pregnancy might be considered if there is fetal hydrops with cystic hygroma, especially when it is related with chromosomal anomalies or other structural abnormalities.

Cystic hygromas without hydrops usually regress completely8. fortunately, fetal with cystic hygroma alone can be survival in the latter life it needs evaluation and close follow up.

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