Original Research Paper

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ANTENATAL DIAGNOSIS AND MANAGEMENT OF CYSTIC HYGROMA WITH HYDROPS FETALIS

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ABSTRACT

Cystic hygroma is a congenital thin-walled cysts that contain lymphatic fluids.Ultrasonography has helped in detecting more and more cases of cystic hygroma with fetal hydrops. Fetal hydrops and cystic hygroma can be diagnosed in first and second trimester of gestation and is associated with higher incidence of aneuploidy, and has higher mortality rate. Mortality rate of cystic hygroma diagnosed before 30weeks is 93% and 84% of those are associated with progressive non-immune hydrops.We present you the case series of 3 cases of cystic hygroma progressing to hydrops fetalis

KEYWORDS

non immune hydrops, cystic hygroma, chromosomal abnormalities, aneuploidy

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1. INTRODUCTION:

Cystic hygroma is a congenital abnormality ofvascular lymphatic system.It can be associated with **aneuploidy(50%), structural anomalies(33.8%).Some of the these fetuses develop hydrops fetalis leading to intrauterine fetal demise.**(1)

Their birth prevalence is 1 in 5000. Cystic hygroma is of two types based on septations- septated and non-septated Small cystic hygroma usually regress spontaneously whereas large cystic hygroma are associated with hydrops fetalis.

When diagnosed in-utero,survival rate of fetuses affected with cystic hygroma is only 2-6%. When hydrops is present with cystic hygroma, the mortality rate is near 100%.

2. Case Report 1:

Patient Mrs X, 24 year old, primigravida with 18weeks and 2days gestation presented to us for a normal antenatal check up.There was no history of hypertension and diabetes. She had Rh positive blood group and had non- consanguineous marriage.Gestational age of the fetus was determined by fetal biometry and it was corresponding with the menstrual history of the patient.

The ultrasound examination done at 12weeks had not picked up any abnormality.

Anomaly scan was done to find congenital anomalies and it was suggestive of a large anechoic cystic collection at the back of the neck with multiple septations – suggestive of cystic hygroma, presence of skin edema, bilateral pleural effusion, ascites and mild placentomegaly- suggestive of hydrops. Figure 1,2

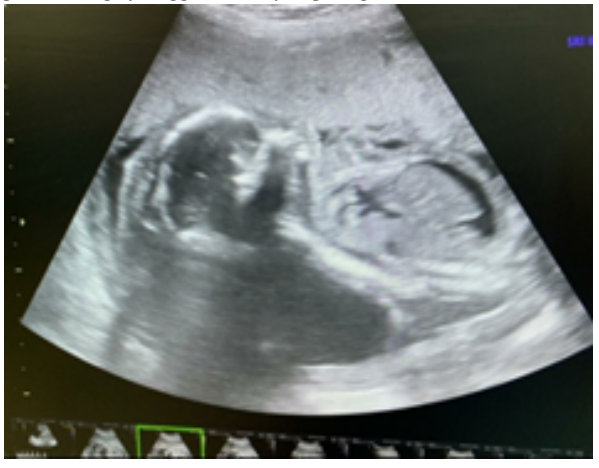


Figure1: USG showing large anechoic cystic collection at back of neck

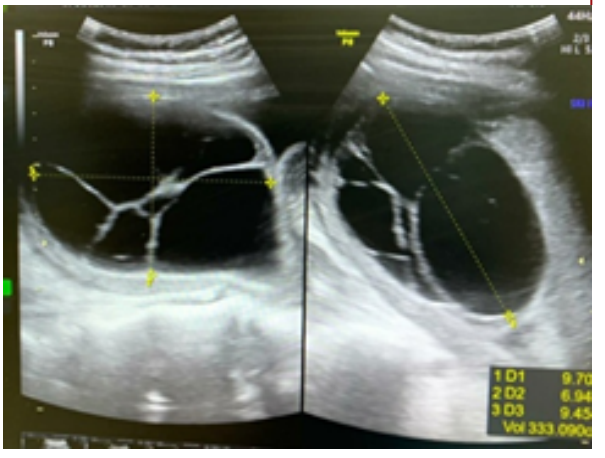


Figure 2: USG showing septated cystic hygroma

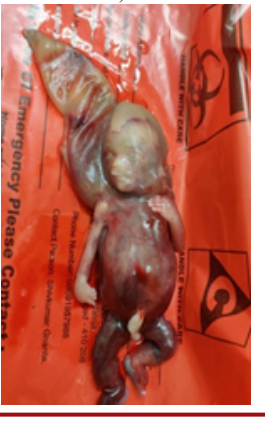
Prognosis of the fetus and possibility of chromosomal disorder, structural disorder, risk of infection were explained to the patient.

Patient was advised for amniocentesis for fetal karyotyping. However the patient declined amniocentesis and opted for termination of pregnancy.

The patient was induced with:

Tab Mifepristone 200mg oral followed by Tab Misoprostol 200mcg oral after 48hours.

Patient aborted. Figure 3 (the time interval between last dose of tab misoprostol and abortion is 4 hours)



Volume - 4 | Issue - 4 | December - 2020 | ISSN No. Online : 2663 - 1199 | ISSN No. Print : 2663 - 1180

Figure 3: Aborted fetus showing cystic hygroma with hydrops fetalis

3. CASE REPORT 2:

Patient Mrs. Y, 28years old, gravida 2 para 1 live 1 with 19weeks and 4days of gestation came to MGM hospital for women at Kalamboli Navi Mumbai. She was Rh positive and had non- consanguineous marriage.

The1st trimester scan at 12weeks 2 days detected nuchal translucency of 10mm(**increased**).Gestational age of the fetus was determined by fetal biometry parameters and it was corresponding with the menstrual history of the patient. The patient was kept under regular surveillance. Fetalanomaly scan done at 18 weeks revealed bilateral cystic lesion behind the neck suggestive of cystic hygroma. The fetus also had bilateral pleural effusion, pericardial effusion, ascites, subcutaneous edema in limbs, the features suggestive of hydrops fetalis. Figure 4,5

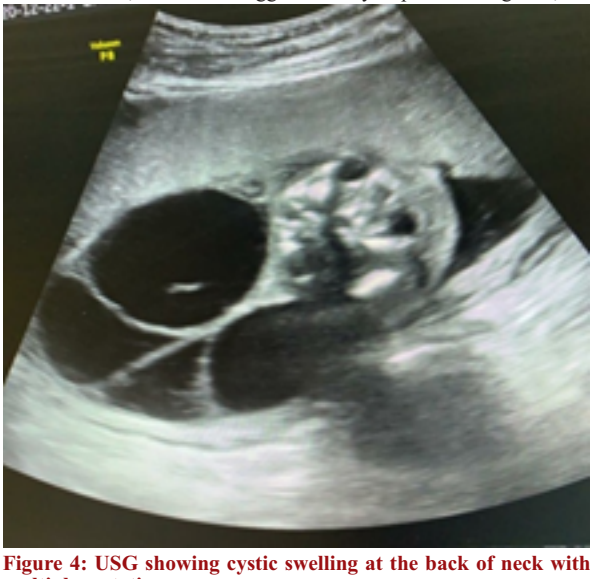


Figure 4: USG showing cystic swelling at the back of neck with multiple septations

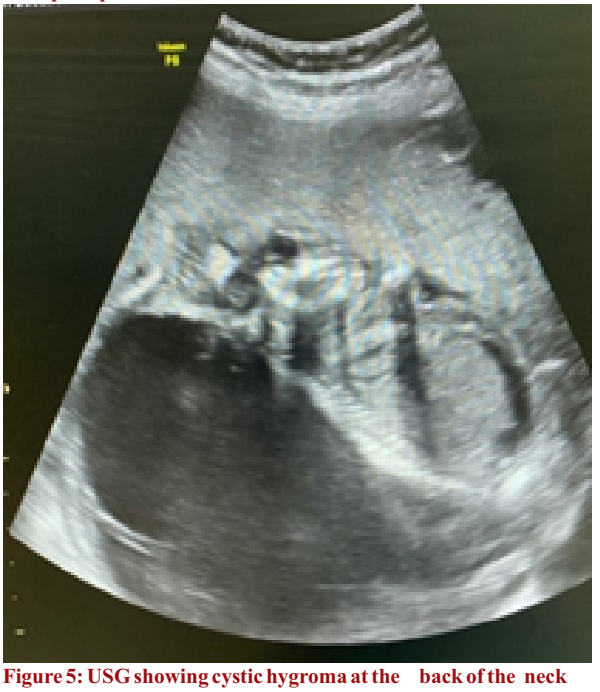


Figure 5: USG showing cystic hygroma at the back of the neck

Prognosis of the fetus and possibility of chromosomal disorder, structural disorder, risk of infection were explained to the patient.Patient was advised for fetal karyotyping. However she opted for termination of pregnancy rather than fetal karyotyping.

Patient was induced with:

Tab Mifepristone 200mg oral followed by Tab Misoprostol 200mcg oral after 48hours followed by Tab Misoprostol 200mcg oral after 6hours.

Patient aborted. Figure 6 (the time interval between the last dose of Tab Misoprostol and abortion is 6hours)

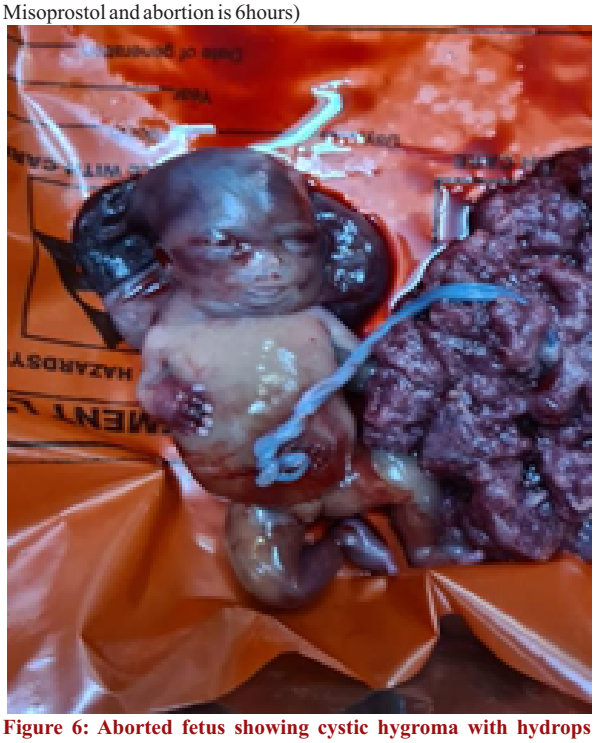


Figure 6: Aborted fetus showing cystic hygroma with hydrops fetalis along with placenta

3. Case Report 3:

Patient Mrs Z, 35years old, gravida 2 para 1 live 1 with 29weeks and 4days of gestation, unregistered and unbooked, came with complaints of pain abdomen since 1day. She had Rh positive blood group and non consanguineous marriage. She had no previous history of congenital anomaly. On evaluation, she had scan of 28 weeks 1day gestation showing bilateral cystic swelling behind the neck suggestive of cystic hygroma, presence of subcutaneous edema involving skull, neck and abdomen, suggestive of hydrops fetalis. She had no history of hypertension and diabetes.

On examination

- General condition fair
- Pulse rate- 84/min
- Blood pressure- 110/70mm/hg
- No pallor No icterus No edema

Systemic examination- normal

Per abdomen examination-

- Uterus 28weeks size
- Longitudinal lie
- Cephalic presentation
- Fetal heart sound present/100 beats/minute
- Uterine contractions -2-3 in 10min lasting for 20 sec
- Per speculum examination- cervix and vagina -healthy
- Per vagina examination- Cervix was 5cm dilated, 40% effaced, station -1, bulging membranes
- Patient and relatives were explained regarding poor prognosis of baby and was observed for spontaneous progression of labour
- Patient delivered vaginally but the baby died after few hours. Figure 7

Volume - 4 | Issue - 4 | December - 2020 | ISSN No. Online : 2663 - 1199 | ISSN No. Print : 2663 - 1180

Figure 7: Aborted fetus showing mild cystic hygroma at back of neck with airway abnormalities

3. DISCUSSION

- Cystic hygroma is one of the most common abnormalities seen in sonography. They arise from **failure of primitive lymphatic tree to connect to the venous system**. Blind lymphatic pouch results in dilation of the lymphatic sac.(2)
- It is mainly located at the cervicofacial and axillary regions (3)
- Its incidence is 1 in 100 pregnancies in first trimester.(4)
- The prevalence of the potential causes of NIHF depends on gestational age at the time of presentation.(5)
- They are highly associated with chromosomal abnormalities especially aneuploidy, cardiac anomailies, skeletal abnormalities, fetal hydrops and intrauterine fetal demise.(1)
- In first trimester, it is most commonly associated with trisomy 21 followed by Turner syndrome (45 X) and trisomy 18 whereas in second trimester, most commonly it is associated with Turner syndrome (45X).
- Small cystic hygroma usually regress spontaneously whereas large cystic hygroma are associated with hydrops fetalis.
- Hydrops fetalis is defined as the abnormal accumulation of fluid in at least two different fetal compartments.(6) Hydrops fetalis are of two types - immune and non-immune based on presence or absence of Rhesus isoimmunisation and histologic evidence of eyrthroblastosis.
- Hydrops is detected sonographically as two effusions or one effusion plus anasarca.
- Edema may be prominent around the scalp, or trunk and extremities. As hydrops progresses in severity, it can be associated with placentomegaly and hydraminos.
- The incidence of nonimmune hydrops fetalis (NIHF) is estimated at 1 in 3000 pregnancies.(7) Overall, prognosis is poor with perinatal mortality rate of 52% to 98%.(8)
- Cystic hygroma and hydrops fetalis in the second trimester have been associated with poor prognosis.

4. CONCLUSION

The association of cystic hygroma and hydrops fetalis in second trimester have been associated with adverse perinatal outcome. Care must be taken in fetus for early diagnosis of cystic hygroma so that early termination of pregnancy can be performed, if patient desires.

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