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A RARE CASE REPORT OF CYSTIC HYGROMA

Dr. Rita D — Prof And HOD, Department of Obstetrics and Gyneacology, Navodaya medical college and research centre.

Dr. Kapu Shanthi Reddy* — Junior resident, Department of Obstetrics and Gyneacology, Navodaya medical college and research centre. *Corresponding Author

ABSTRACT Cystic hygromas are malformations of the lymphatics system that appear as fluid-filled, membranous cysts, lined by true epithelium in the anterolateral or occipito-cervical area. They result from the jugular lymphatic obstruction sequence. Most of the cystic hygromas are associated with chromosomal anomalies. When diagnosed in-utero, the survival rate of foetuses affected with cystic hygroma is only 2-6% . The incidence of cystic hygroma is estimated to be 1 case per 6000-16000 live births. A case of 22 years old , primigravida 13 weeks period of gestation with no complaints on her first antenatal checkup , was diagnosed prenatally during her first antenatal scan, with a large cystic hygroma with septation. The pregnancy was terminated with the consent of the parents.

KEYWORDS : Cystic hygroma, Hydrops fetalis, Chromosomal abnormalities

INTRODUCTION

Cystic hygroma is a benign congenital malformation of the lymphatic system that has its genesis in the lack of development of communication between the lymphatic and venous systems. The cyst may be unilocular or multilocular and could be of variable size but is characteristically brilliantly transilluminant. The incidence of cystic hygroma is approximately 1/6000 live births[1]

Most of the cystic hygromas are associated with chromosomal anomalies. When diagnosed in-utero, the survival rate of foetuses affected with cystic hygroma is only 2-6%.[2]

A cystic hygroma in a developing baby can progress to hydrops and eventually foetal death.

Case Report

A 22yr old , primigravida presented to our OPD with history of 3 months of amenorrhoea with no complaints on her first antenatal checkup.

Detailed history was noted, non consanguineous marriage , no significant family history of congenital anomalies

O/E

- PR -82bpm
- BP- 116/72 mmHg
- CVS/RS/CNS - NAD
- Per abdomen – uterus just palpable

UPT was positive

Dating scan was done - A single live fetus of 13 weeks period of gestation detected as cystic hygroma likely lymphatic malformation.

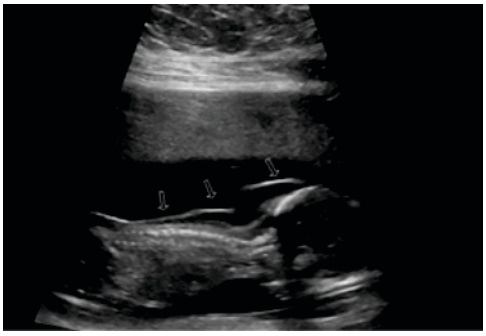

Fig 1 – USG showing cystic hygroma

Anc Profile

Hb - 12.1g%
Blood group – A POSITIVE
Total count – 1O, OOO mm^3
Platelet count –2.76 lakh/mm^3
Blood group – A POSITIVE
Urine routine – normal
RBS – 65 mg/dl
TSH – 1.14 mIU/L
HIV- NON REACTIVE
HBSAG- NEGATIVE
VDRL - NON REACTIVE

Management

- The couple is counselled about the diagnosis ,its prognosis and the need for termination of pregnancy .
- Termination done by Tab Mifepristone 200mg p/o followed by foleys induction and Tab misoprostol 400mcg 2 hrs apart 2 doses . Inj Oxytocin 10units in 1 pint RL was infused during expulsion . She expelled A single dead male fetus of weight 180 gm .
- O/E of the fetus – cystic swelling present over the nape of the neck.
- Check scan done after 24 hrs showed RPOC and check curettage was done.

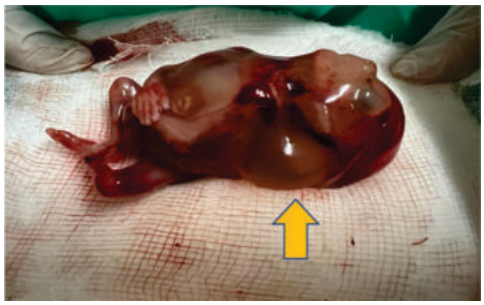

Fig 2 - Expelled fetus with cystic hygroma

DISCUSSION

A cystic hygroma can present as congenital or develop at any time during a person's life. Some cases of congenital cystic hygromas resolve leading to webbed neck, edema, low posterior hair-line. In other instances the hygroma can progress in size to become larger than the fetus.

Cystic hygromas may be associated with Turner syndrome (most common), Noonan syndrome, trisomies, fetal alcohol syndrome, chromosomal aneuploidy, cardiac anomalies and fetal hydrops.[3]

Imaging Routine antenatal ultrasound can detect cystic hygroma from 12 weeks gestational age. If a cystic hygroma is seen, the scan should also carefully assess for associated structural abnormalities, as well as monitor fetal growth and well being throughout pregnancy. High-resolution transvaginal ultrasound or 3D ultrasound can provide more detail than routine ultrasound. Fetal echocardiography is recommended at 18–22 weeks gestational age if a cystic hygroma persists. Fast-spin magnetic resonance imaging (MRI) may clarify the situation if the diagnosis is uncertain. It also helps delineate the extent of the cystic hygroma and its relationship to the surrounding structures..

Combined First Trimester Screening Test An ultrasound to determine the nuchal translucency thickness and maternal blood for the Triple test (alpha-fetoprotein, unconjugated oestriol and human chorionic gonadotrophin) assesses for the risk of aneuploidy.

An amniocentesis will provide cells for a fetal karyotype and FISH (fluorescence in situ hybridisation) testing, particularly to test for abnormalities of chromosomes 13, 18, 21, X and Y.[4] The management of cystic hygromas is preferably surgical, although a careful "wait and see" policy may be indicated in few asymptomatic cases, as spontaneous regression has been reported.

Indications for surgery in pediatric cases include significant cosmetic deformity, obstructive symptoms, bleeding and recurrent infections. Other treatment modalities include aspiration, radiation, and injection of sclerosing agents, in particular the agent OK-432, derived from a strain of streptococcus pyogenes, which has been used successfully, especially in macrocystic lymphangiomas and in patients who are at increased anesthetic risk.[5]

The recurrence risk for cystic hygroma due to aneuploidy is usually very low. Cases of cystic hygroma with a normal karyotype can occur as a familial condition with autosomal recessive inheritance (25% recurrence risk).

CONCLUSION

- Fetuses with cystic hygroma are at high risk for adverse outcomes .
- Due to its extremely poor prognosis , termination should be considered when early diagnosis is made associated with other chromosomal abnormalities .

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