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CASE STUDY

CYSTIC HYGROMA ASSOCIATED WITH NON-IMMUNE FETAL HYDROPS: ANTENATAL DIAGNOSIS OF A RARE ENTITY

***Dr. Abhinav Aggarwal and Dr. Aakriti Kapoor Aggarwal**

A-1/ 237, First Floor, Janak Puri, New Delhi -110058, India

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ABSTRACT

Cystic hygromas are manifestations of early lymphatic failure. They appear as simple or multi-septated fluid collections, most commonly at nuchal region. Prenatal diagnosis of cystic hygroma can be made within 2nd trimester of pregnancy. Hydrops fetalis is defined as accumulation of fluid in serous cavities and/ or edema of soft tissues in the fetus. Incidence of fetal hydrops is approximately 1 in 2500 to 3500 neonates with incidence co-existence of hydrops and cystic hygroma even rarer. We here present a case of a fetus of 14 weeks gestational age with diagnosis of large cystic hygroma complicating to fetal hydrops.

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INTRODUCTION

Hydrops fetalis is derived from a latin word meaning edema of the fetus and was first described by Ballantype in 1892 (Abrams *et al*., 2007). Hydrops fetalis is defined as accumulation of fluid in serous cavities and/ or edema of soft tissues in the fetus (Bijma *et al*., 2004). Incidence of fetal hydrops is approximately 1 in 2500 to 3500 neonates (Smith *et al*., 1960). Cystic hygromas are manifestations of obstruction to lymphatic system. Their appearance can vary from increased nuchal translucency to thin-walled cystic masses, which can become larger than fetal head (Nadel *et al*., 1993). Coexistence of cystic hygromas and fetal hydrops is rarely encountered and is associated with higher incidence of morbidity and mortality (Srivastava and Gupta, 2006). Increased incidence of aneuploidy has also been reported in such cases (Chen, 2001). We hereby present a case of a fetus of gestational age of 14 weeks who was diagnosed with a large cystic hygroma associated with hydrops fetalis.

Case report

A 24- year-old Rh positive, primigravida was referred to our department for routine sonographic examination of pregnancy. Her last menstrual period (LMP) was not correctly known and she had not undergone an ultrasonography examination previously during the course of pregnancy. She had no complaints.

Trans-abdominal ultrasonography examinationrevealed a live intrauterine fetus of gestational age of 14W3D with well corresponding biparietal diameter, head circumference and femur length. However, a large septated cystic lesion of size approximately 4cm x 3cm x 2cm was seen along posterior aspect of fetus extending from nuchal region superiorly upto lower aspect of fetal abdomen inferiorly. Diffuse subcutaneous edema was noted over fetal thorax and abdomen. Bilateral pleural effusion was also noted in the fetus. Polyhydramnios was not visualized in the patient. No pericardial or pleural effusion was noted at the time of scan. No other abnormality was detected. Placenta appeared normal for gestational age.

A diagnosis of large cystic hygroma associated with fetal hydrops was made.Considering the poor prognosis the patient was advised for further investigation through karyotyping. Owing to cultural beliefsthe patient refused and was lost on follow up.

DISCUSSION

Cystic hygromas can appear as simple or multi-septated fluid collections most commonly in nuchal region as a result of

Corresponding author: Dr. Abhinav Aggarwal,
A-1/ 237, First Floor, Janak Puri, New Delhi -110058, India.

failure of communication of lymphatic system with venous nuchal system resulting in loss of homeostasis (Bronshtein *et al*., 1989; Ali *et al*., 2012). Fetal hydrops was previously considered to exist only in Rh negative pregnancies, but in recent years has been diagnosed in Rh positive mothers and termed as non-immune fetal hydrops (Ali *et al*., 2012). Fetal hydrops is usually characterized by fluid accumulations in various body cavities like pleural, pericardial and peritoneal and soft tissues with a wall thickness of > 5mm (Has, 2001). In our case also we saw a large multi-septated cystic hygroma in a Rh positive female. This recent upturn in diagnosis of non-immune hydrops fetalis can be attributed to vast improvement in diagnostic capabilities. Studies have shown that multi-septated cystic hygromas have much poorerprognosis as compared with simple cystic hygromas. One of the reasons attributed to it is resolution of cystic hygromas in cases of simple cysts. Incidence of aneuploidy has also been reported at a much higher rate in multi-septated cystic hygromas as compared to their simple counterparts (Bronshtein *et al*., 1989). Our case had a large multi-septated cystic hygroma which was a sign of poor prognosis of the disease.

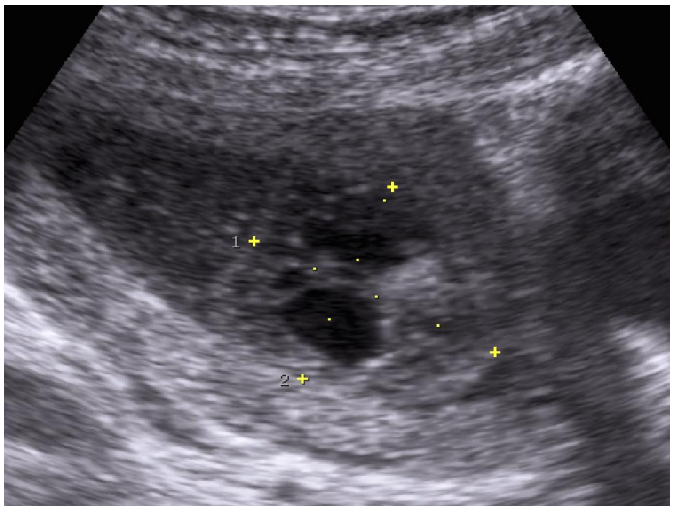

Figure 1. Large multi-septated cystic hygroma

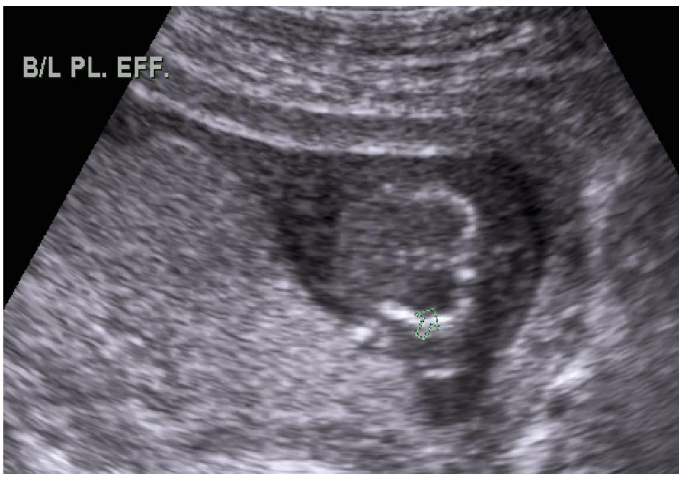

Figure 2a. Diffuse subcutaneous edema along with caudal extension of cystic hygroma in the fetal thoracic region, Pleural effusion is also noted

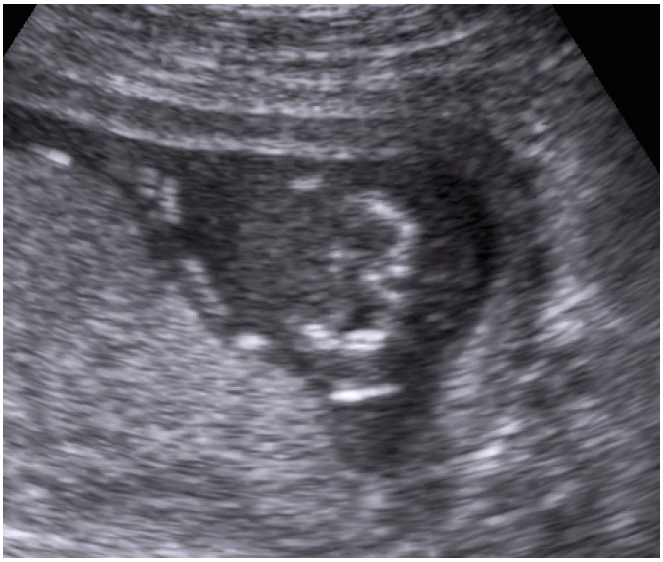

Figure 2b. Diffuse subcutaneous edema along with caudal extension of cystic hygroma in the fetal thoracic region, Bilateral pleural effusion is also noted

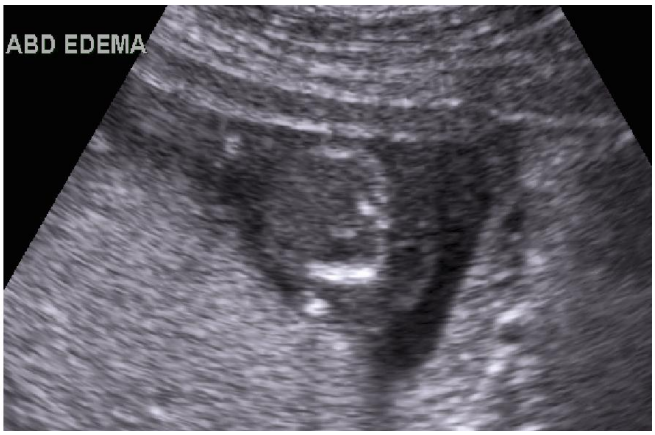

Figure 3a. Diffuse subcutaneous edema along with caudal extension of cystic hygroma in the fetal abdominal region

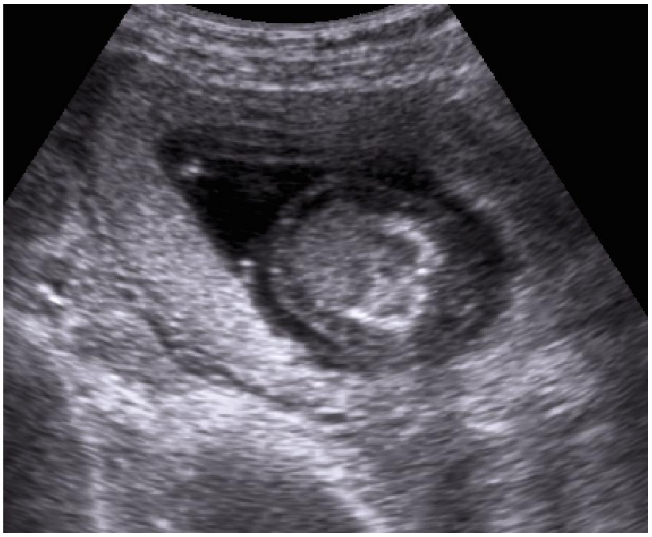

Figure 3b. Diffuse subcutaneous edema along with caudal extension of cystic hygroma in the fetal abdominal region

Common genetic abnormalities associated with cystic hygromas include Turner's syndrome (predominantly 45 XO karyotype), Down syndrome (trisomy 21), Edwards syndrome (trisomy 18) and Noonan syndrome (45X/ 46XY) along with other structural and chromosomal abnormalities (Nadel *et al*., 1993; Has, 2001). Although karyotyping was not possible in our case, we strongly believe that karyotyping must be done in every case of cystic hygroma not only for diagnostic purpose but also for further increase in knowledge about the entity.

Other abnormalities associated with fetal hydrops are polyhydramnios, thickened placenta and hepatosplenomegaly (Ali *et al*., 2012). We did not find any of these findings in our case probably since ours was an early diagnosis at 14 weeks of gestational age considering transabdominal approach of examination. Although there are studies where fetal hydrops was diagnosed at 8-12 weeks, most of these studied used transvaginal approach (Bronshtein *et al*., 1989). This makes our case one of the rarest in terms of early diagnosis. Although bilateral pleural effusion was noted in fetus we did not find any evidence of pericardial or peritoneal effusion at the time of scan, which might also be due to relatively early nature of diagnosis.

Although pure cystic hygromas may resolve spontaneously, those associated with hydrops lead to antenatal demise of the fetus (Kiyota *et al*., 2009; Beke *et al*., 2009). This makes detailed genetic counseling and further sonographic follow up extremely necessary in all cases of antenataly diagnosed fetal hydrops and cystic hygroma. We hereby conclude by saying that increasing evidence suggests association in cystic hygroma (especially multi-septated) and fetal hydrops (especially non-immune), which warrant early diagnosis & management and regular follow up.

REFRENCES

Abrams ME *et al*. Hydrops fetalis: a retrospective review of cases reported to a large national database and identification of risk factors associated with death. *Pediatrics*, 2007; 120(1): 84-9.

Ali MK, Abdelbadee AY, Shazly SA, Othman ER. Hydrops fetalis with cystic hygroma: A case report. *Middle East Fertility Society Journal,* 2012; 17: 134-135.

Beke A, Joo JG, Csaba A, Lazzar L, Ban Z, Papp C *et al*. Incidence of chromosomal abnormalities in the presence of fetal subcutaneous edema, such as nuchal oedema, cystic hygroma and non-immune hydrops. *Fetal Diagn Ther.,* 2009; 25(1): 83-92.

Bijma HH *et al*. Ultrasound diagnosis of fetal anomalies: an analysis of perinatal management of 318 consecutive pregnancies in a multidisciplinary setting. *Prenatal Diagn*., 2004; 24(11): 890-5.

Bronshtein M, Rottem S, Yoffe N, Blumenfeld Z. First-trimester and early second-trimester diagnosis of nuchal cystic hygroma by transvaginal sonography: diverse prognosis of the septated from the nonseptated lesion. *Am J Obstet Gynaecol*., 1989 Jul; 161(1): 78-82.

Chen CP. Prenatal diagnosis of partial trisomy 3q (3q22___> qter) and monosomy 6q(6q25.3___>qter) in a fetus sonographic findings of cystic hygromata colli and unilateral pleural effusion. *Prenat Diagn*., 2001; 21:73.

Has R. Non-immune hydrops fetalis in the first trimester: a review of 30 cases. *Clin Exp Obstet Gynaecol*., 2001; 28(3): 187-90.

Kiyota A, Tsukimori K, Yumoto Y, Hojo S, Morokuma S, Fukushima K *et al*. Spontaneous resolution of cystic hygroma and hydrops in a fetus with Noonan's syndrome. *Fetal Diagn Ther*., 2009; 24(4): 499-502.

Nadel A, Bromley B, Benacerraf B. Nuchal thickening or cystic hygromas in first – and early second trimester fetuses: prognosis and outcome. *Obstet Gynecol*., 1993; 82: 43-8.

Smith DW *et al*. A new autosomal trisomy syndrome multiple congenital anomalies caused by an extra chromosome. *J Pediatr.,* 1960; 57: 338-45.

Srivastava S, Gupta A. Fetal cystic hygroma with nonimmune hydrops fetalis and prenatal diagnosis of Tutner syndrome. *J Obstet Gynecol India* 2006 May/June; 56(3): 260-261.

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