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CASE REPORT

Cystic Hygroma: A Grave and Thought Provoking Entity

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Introduction

Cystic hygroma is a venolymphatic malformation due to malfunction in lymph drainage from head into the jugular vein resulting in its accumulation in the jugular lymphatic sac. At birth the prevalence is 1 in 5000. In 70 percent cases, it is associated with aneuploidy. Small, nonseptated cystic hygroma with normal fetal karyotype and echocardiogram carry good prognosis, whereas large, septated cystic hygromas are usually associated with hydrops fetalis and carry a poor prognosis. There are no guidelines regarding mode of delivery.

In 2013, a large cystic hygroma was delivered vaginally but prognosis of baby could not be evaluated due to lost to follow-up.

Case Report

19 year old primigravida visited our tertiary care center first time at 19 weeks 2 days period of gestation. Ultrasonography at a private clinic at 18 weeks 6 days period of gestation revealed 3.5*3.2 cm septated cystic hygroma with increased vascularity on right side of neck. We re-evaluated the fetus in utero sonographically for any other congenital anomaly. No other anomaly was present. Pediatric surgery consultation, echocardiography and karyotyping was advised, but she was lost to follow-up due to her financial constraints. Next ultrasonography done at 34 weeks gestation, outside, revealed that cystic hygroma had increased to 7.7*4.9 cm (Fig. 1). She reported to our hospital in active labor at 38 weeks period of gestation with 5.5 cm cervical dilation. Repeated ultrasonography revealed that neck swelling had increased to 11.2*8.2 cm. Amniotic fluid index and growth of baby was normal. After consultation with pediatric surgeon, vaginal delivery was contemplated, keeping in view the need for aspiration of the cyst in second stage of labor. Cesarean section was kept as backup plan. Continuous intrapartum fetal monitoring was done. Liberal episiotomy was made after full dilation. Vaginal delivery was conducted successfully; intrapartum aspiration was not required. She delivered 2.6 kg alive male baby who cried immediately after birth. On examination by pediatrician, APGAR score was 7/10 and 9/10 at 1 min and 5 min, respectively. Baby appeared appropriate for his gestational age. He had a large 13*8 cm soft fluctuant cystic non-pulsatile trans-illuminating swelling with variable consistency covering half of neck circumference (Fig. 2). It increased in size on crying. No clinical features of Noonan syndrome, Multiple-pterygium syndrome, Fryns syndrome, Neu-Laxova syndrome or hydrops were detected. Due to large size of hygroma, baby had respiratory distress. He was resuscitated and shifted to NICU**.** Baby was intubated on day one. Day 2, ultrasonography whole abdomen, cranium and 2D ECHO were advised to rule out other associated anomalies, but parents did not give consent for investigations. Forty ml of straw colored fluid was aspirated from the cyst on day two. 0.6 units of bleomycin was injected into the cyst on day four after aspiration of 30 ml of straw colored fluid. Baby was continued on nasogastric feed. Baby was stable with minimal ventilator support. Two attempts of extubation made on day four and day seven but failed and the baby needed re-intubation. Main concern was collapsible airway. After third attempt of extubation on day eleven, baby had an episode of vomiting. At that time, baby desaturated and needed re-intubation.

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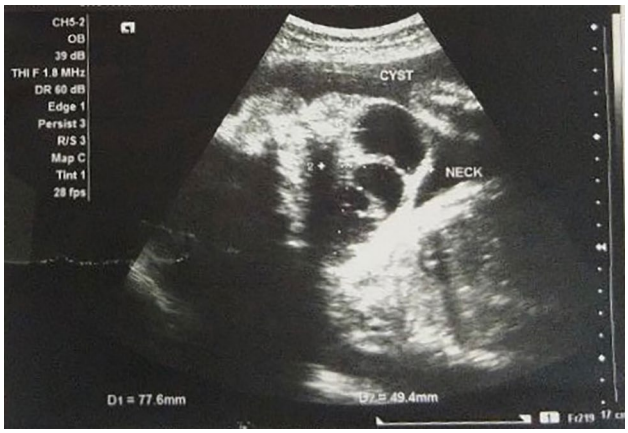

Fig. 1 Ultrasonogram showing cystic hygroma at 34 weeks period of gestation

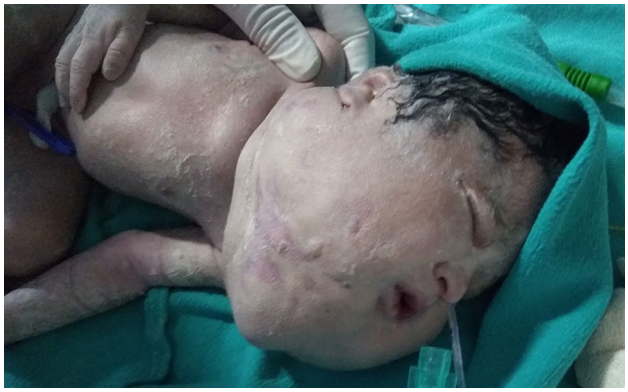

Fig. 2 Neonate immediately after delivery showing cystic hygroma (anterolateral view)

Following day, the baby was found to have cyanosis along with bradycardia, saturation dropped probably due to aspiration of gastric contents after vomiting. Resuscitation was tried with three cycles of CPR but could not be revived. Parents did not give consent for autopsy.

Discussion

Cystic hygroma is painless trans-illuminating swelling developing due to lymphovascular malformation. 61% of cystic hygroma are associated with aneuploidy, trisomy 21 being most common if cystic hygroma is diagnosed in the first trimester and turner syndrome in case of second trimester diagnosis [1]. In the absence of aneuploidy, there is high risk of association with cardiac anomalies like coarctation of aorta and hypoplastic left heart. Cystic hygroma can be a part of genetic syndrome like Noonan syndrome. There is 18% chance of live birth in all cases of cystic hygroma, 10% in case of septated cystic hygroma with normal karyotype and morphology. Septated cystic hygroma has five times chance of aneuploidy, twelve times associated cardiac malformations, six times a chance of fetal or neonatal death in comparison with non-septated cystic hygroma.

Fetal cystic hygroma can be diagnosed by ultrasonography in the first trimester. Parents should be counseled and importance of regular follow-up emphasized. Once the fetus is diagnosed with cystic hygroma, the karyotype of baby should be determined and a detailed anomaly screening should be performed. Comparative genomics hybridization, a type of chromosomal microarray analysis, can be used instead of normal karyotyping though the cost is a prohibiting factor. It can detect additional clinically significant abnormalities in approximately five to eight percent of fetus with normal conventional karyotyping and ultrasound abnormality [2]. In case of euploid karyotype with normal anomaly scan, parents should be offered a choice to continue with expectant management. Choice of termination of pregnancy should be offered in case of aneuploid child with structural malformation with septated cystic hygroma. Fetal echocardiography should be done to rule out associated cardiac anomalies.

There is no consensus regarding time and route of delivery. In case of small, non-septated cystic hygroma, vaginal route of delivery should be preferred, whereas in case of large septated cystic hygroma, vaginal delivery with aspiration of the cyst during the second stage of labor can be attempted or elective cesarean section can be planned, depending on the survival possibilities of the baby after birth and NICU facilities. In case of large septated cystic hygroma with antenatally detected airway obstruction, cesarean section along with EXIT (Ex Utero Intrapartum Treatment) can be planned.

Management of pediatric airway is difficult and in cases of cystic hygroma, due to disturbed anatomy, it becomes more difficult. Airway obstruction should be ruled out prenatally by MRI, so that mode of delivery can be planned to have a better outcome.

Treatment: mainstay of treatment includes sclerotherapy (bleomycin, OK-432) and surgical resection along with supportive management. Bleomycin is given in a dose of 0.5 mg/kg body weight intralesionally after aspiration of the contents of cystic hygroma. It should not exceed beyond 10 units at a time [3]. Main concern is pulmonary toxicity related to bleomycin therapy which increases with a total dose exceeding 400 units or a single dose exceeding 30 mg/m^2 of body surface area given intra-venous. Sclerotherapy targets specific cysts in each session necessitating a minimum of 2 sessions for every cyst. Combination of sclerotherapy with sirolimus provides better results. The dosage and the duration of treatment with sirolimus differs in various studies as no evidence-based guidelines exist for proper dosing and duration of treatment [4]. In most studies, Sirolimus

is given in a dose 0.4 to 0.8 mg/m^2 2 times/day orally with its blood levels to be maintained between 10 and 15 ng/mL.

Till now, a maximum of 15*30 cm size cystic hygroma has been reported in the literature. The baby was delivered by cesarean section and was managed by surgical treatment for cystic hygroma. Another fetus with 14*11 cm large axillo-thoraco-abdominal cystic hygroma was delivered at 38 weeks period of gestation vaginally, and was treated surgically on day 4 of neonatal life.

Declaration

Conflict of interest The authors declare that they have no conflict of interest.

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