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CASE REPORT

Can Large Fetal Cystic Hygromas Be Delivered Vaginally?

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Introduction

Fetal cystic hygroma is a rare developmental congenital anomaly of the lymphatic system, characterized by the formation of a multilocular, variably sized cystic mass. The incidence of cystic hygroma is estimated to be 1:6,000 pregnancies, but it is a relatively common anomaly in miscarried fetuses, with a frequency of 1:875 [1]. Among the cystic hygroma, the septate cystic hygroma carry poor perinatal outcome and management difficulties after birth; hence, the mode of delivery of cystic hygroma babies needs special concern. There are four major concerns in such patients:

(1) What should be the antenatal management protocol?
(2) How should the delivery of the baby be planned, i.e. vaginal or cesarean?
(3) What should be the intrapartum management of the baby in case there is mass compression over the cervical structures like trachea?
(4) What should be the treatment for the baby once the delivery is completed?

We, hereby, report a case of large cervico-facial cystic hygroma, although planned for cesarean but delivered vaginally without any intrapartum intervention.

Case Report

A 30-year-old woman as second gravida with one living issue was referred to tertiary institute because of a big, thick multicystic mass in neck region detected by routine ultrasonography (USG) performed at a private clinic at 34 weeks' gestation. This ultrasound showed fetal growth parameters corresponding to gestational age. A targeted sonographic examination, performed in our hospital, at 35 weeks' gestation, revealed a live fetus with a septate cystic structure projecting from the dorsal aspect of the neck, measuring 12.51 × 7.26 cm (Fig. 1). There was no evidence of fetus hydrops, or any other congenital malformation. Fetal parameters were corresponding to the calculated gestational age. Amniotic fluid index was normal, and fetal echocardiography was normal. She was advised for fetal karyotyping, but was unwilling for any invasive procedure. Her antepartum period was uneventful. Pediatric surgery, neonatology, and genetic counseling were done, and neonatal survival

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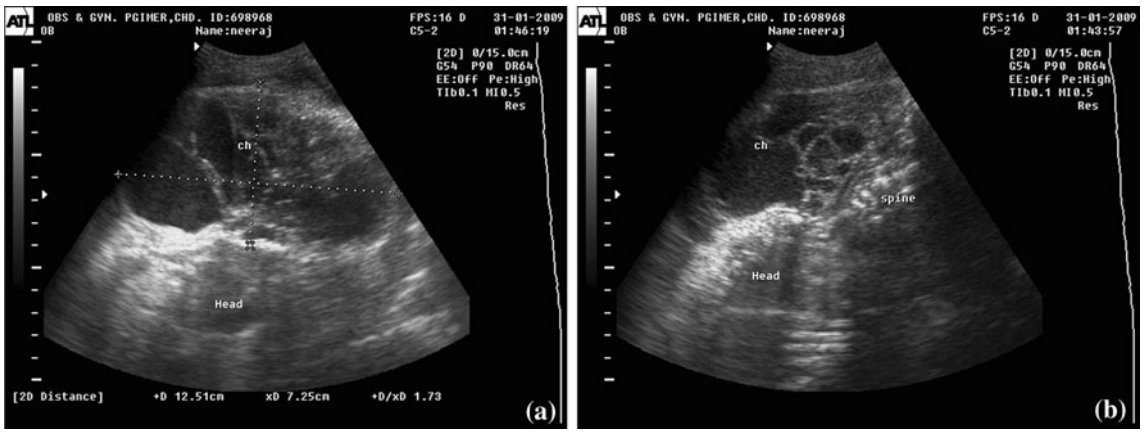

Fig. 1 Antenatal sonogram showing cystic hygroma in **a** sagittal plane, **b** coronal plane

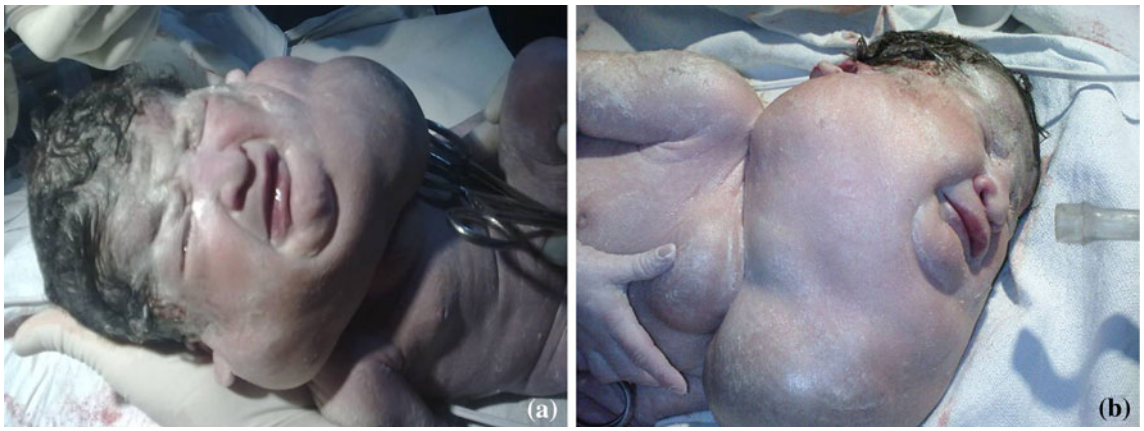

Fig. 2 Neonate immediately after delivery (**a** anterior view, **b** lateral view)

prognosis was discussed with the parents. She was initially planned for cesarean section at 38–39 weeks period of gestation for huge fetal cystic hygroma as per reviews in the literature. Later, she presented in labor ward in advanced stage of labor at 38 weeks. The decision of cesarean section was reviewed during her active labor, in agreement with pediatric surgeon. Parents were re-counseled regarding the mode of delivery, and they opted for vaginal delivery. Delivery was conducted in the presence of pediatric surgeon. Need of aspiration during second stage of labor, if required, was also considered. Patient had normal delivery without any features of obstruction. There was no difficulty in immediate resuscitation of the newborn. She delivered a female newborn baby weighing 2,825 g with Apgar index of 8 at 1 min and 10 at 5 min. The baby did not require any respiratory assistance immediately after delivery; hence, aspiration of the cyst was also not required.

The newborn on examination had a large mass in the posterolateral aspect of left side of neck, extending to anterolateral aspect of right side of neck as well as anterior chest wall just above the sternum (Fig. 2). The mass was cystic in consistency, fluctuant, and transilluminant. No other obvious malformations were found on detailed evaluation. On day 2 of life, the baby had mild inspiratory stridor but did not require intubation for airway. Subsequently, the baby was shifted to neonatal surgical intensive care unit and required intubation for respiratory distress. Roughly, 40 ml of fluid was aspirated from the cystic hygroma, and intracystic injection bleomycin 0.2 U/kg was given as sclerosing agent. The baby was followed up a week later, and there was a significant reduction in hygroma size. Unfortunately, parents could not afford further treatment, and the baby was lost to follow up.

Discussion

Various theories have been proposed for the formation of cystic hygroma. The first theory [2] suggests that an early jugular–lymphatic obstructive sequence could cause

hydrops fetalis, pterygium colli, and cystic hygroma. This obstruction impedes communication between the jugular–lymphatic sacs and the internal jugular vein. Second theory suggests that cystic hygroma is caused by an abnormal embryonic sequestration of lymphatic tissue and its subsequent failure to join normal lymphatic channels [3]. It usually affects the head and neck (approximately 75 %), with a left-sided predilection. Within the neck, the posterior triangle tends to be the most frequently affected. The prenatal diagnosis of cystic hygroma by ultrasound is well documented in the literature. The incidence varies with gestational age.

Till date, largest cystic hygroma of size 15 × 30 cm is reported in the literature, which was delivered by cesarean section. This baby underwent mechanical ventilation in immediate neonatal period and underwent surgical treatment for cystic hygroma.

Antenatal Protocol

Fetuses with cystic hygroma are at high risk for adverse perinatal outcome. This malformation can be seen on antenatal USG from the end of the first trimester of pregnancy. Prenatal diagnosis including invasive procedures to know karyotype of the fetus should be offered. In cases with normal karyotypes, detailed sonography beside the exclusion of hydrops fetalis should be concentrated on fetal heart defects, skeletal, urogenital, and craniofacial anomalies. These anomalies might cause severe morbidity and mortality; hence, they affect the prognosis.

As there is no consensus regarding mode of delivery, it is prudent to review the factors affecting the perinatal outcome and mode of delivery. As fetal lymphangiomas are associated with high morbidity and mortality if airway compression occurs, it is of paramount importance to identify airway obstruction prenatally. Prenatal ultrasound can reveal an airway obstruction (by secondary signs such as polyhydramnios, decreased swallowing, or tongue protrusion) [4]. However, assessment of the extent of the mass and the direct visualization of both the larynx and trachea are difficult on USG. Fetal MR imaging can critically estimate the severity of airway obstruction and can help in planning an approach for possible tracheostomy and resection of the mass after delivery. The use of MR imaging provides excellent contrast resolution and allows for assessment of the extent of airway displacement or compression. MR imaging also allows for enhanced evaluation of possible extension into the thorax cavity and the relation of the mass to the neck vessels [5]. Hence, well-evaluated cystic hygroma cases with no other congenital malformations, and chromosomal anomaly should undergo USG for primary evaluation regarding decision of mode of delivery. If large size of the hygroma is suspected or sign of obstruction is noticed, then it should get MR imaging done and subsequently should be planned for cesarean section.

There are no data regarding benefits derived from cesarean delivery compared with vaginal delivery. In the presence of structural anomalies, fetal hydrops, aneuploidy, or a large multiloculated lesion, the route of delivery opted should ensure the safest manner for the gravida. In cases with isolated, small, non-septated lesions with no hydrops, the vaginal route seems acceptable. Large cystic hygromas that may threaten the airway must be delivered early in life, and cesarean section may offer optimal conditions for airway management.

Mode of delivery is a matter of concern in this group of women, especially in developing countries like India. In our opinion, in developing countries like India where infectious morbidity after cesarean is very high and confirmatory prenatal diagnosis is usually not available, aspiration of the cyst as and when required to facilitate vaginal delivery can be opted as preferred mode of delivery. Theoretically, it is unusual for a compressible cystic structure to cause obstruction during labor and can be aspirated during the second stage of labor or immediately after delivery as per the need. It would be more convenient to do cyst aspiration and endotracheal intubation as and when required in the presence of pediatric surgeon than more cumbersome intrapartum treatment during cesarean, as has been advised in ex utero intrapartum treatment (EXIT) procedure [6]. In the case of group of patients, who present unevaluated, late, or in an advanced stage of labor, with fetal cystic hygroma with undiagnosed congenital malformations, cyst aspiration and subsequent vaginal delivery procedure is an option. Also, cystic hygroma presenting late, unevaluated, but having no other obvious congenital malformations, as in our patient, can be offered vaginal birth accompanied with cyst aspiration, if required. By this approach, mother can avoid surgery-related complications, and, moreover, it is justifiable to have vaginal delivery if there is undiagnosed congenital malformations in the same baby. Antepartum cyst aspiration has also been proposed to facilitate vaginal delivery, by some authors, but it may further increase the size of the cyst because of hemorrhage.

During cesarean delivery, to treat suspected airway obstruction, EXIT procedure was devised as the therapeutic option of operating on placental support [6]. A few authors reported the advantage of EXIT along with routine rigid bronchoscopy to secure the neonate's airway without preliminary attempts at endotracheal intubation for a group of fetuses with antenatally detected airway obstruction.

The management of cystic hygromas is preferably surgical. Other treatment modalities include aspiration, radiation, and injection of sclerosing agents. Particularly,

OK-432, has been used successfully, especially in macrocystic lymphangiomas and in patients who are at increased anesthetic risk. Bleomycin has also been used successfully as sclerosing agents. Commonly, these infants develop neural paresis or paralysis after excision of massive cervical lymphangiomas as these congenital abnormalities tend to distort normal anatomy, surround, or displace neurovascular structures making their identification quite challenging intraoperatively.

In conclusion, in either scenario, cesarean delivery or vaginal birth, perinatal coordination between various medical teams particularly radiology, obstetrics, and pediatric surgery is necessary for the successful delivery and preventing infantile respiratory obstruction. Subsequent treatment in neonatal period should be individualized depending upon the cystic hygroma size, presentation, and complexity.

Conflict of interest The authors have no conflicts of interest to declare.

Ethical standards This case report was approved by the departmental screening committee. Parents of the case gave informed consent for publishing photographs, clinical history, and management of the same. The parents were assured that anonymity will be preserved. This case report does not violate the policies and/or procedures established by the journal.

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