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BRIEF COMMUNICATION

Management of Isolated Fetal Lymphangiomas Following Prenatal Diagnosis: Case Series

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Abstract Lymphangiomas are benign cystic lesions caused by the failure of lymphatics to communicate with the lymphatic-venous drainage system. They have an incidence at birth of about 1 in 6000 and can occur at almost any location, most commonly the soft-tissues of the neck. Prenatal diagnosis is by ultrasound, where they appear as cystic masses of variable size, often with septations inside and absent flow on color doppler. Antenatal management involves serial monitoring of the fetus by ultrasound with attention to possible pressure effects and airway obstruction depending upon the size and location of the lymphangioma. The mode of delivery must be decided based on the possibility of obstructive labor or the need for resuscitation. Rarely, a large mass over the anterior neck causing airway obstruction may need an EXIT procedure. Large cystic hygromas need surgical excision while small to medium size lesions can be managed by conservative measures including sclerotherapy. We report three cases of isolated fetal cystic hygromas diagnosed prenatally at various gestations. Two were of moderate size over the anterolateral aspect of the neck and one case had a huge lymphangioma involving the entire right side of the chest wall.

Keywords Fetal lymphangiomas · Fetal cystic hygromas · Fetal neck masses · Fetal thoracic lesions

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Introduction

Lymphangiomas are benign malformations with an incidence of 1 in 6000 at birth [1]. They are composed of dilated cystic lymphatic sacs due to failure to communicate with the lymphatic-venous system. Common sites are soft tissues of the neck (75%), axilla (20%), thorax/mediastinum (1%), retroperitoneal/intraabdominal (2%), and lower extremities (2%) [2, 3]. Lesions vary from tiny blebs to large masses and those presenting over the neck are commonly called cystic hygromas. In the 1st or 2nd trimester, they usually involve the posterior triangle of the neck, often have associated chromosomal abnormalities (60%) or structural malformations and high mortality. Isolated cystic hygromas, on the other hand, are often detected in the third trimester, usually involve anterior or anterolateral cervical triangle, and have an excellent prognosis (Table 1) [4–7]. Described here is a series of three fetal lymphangiomas prenatally diagnosed and managed.

Table 1 Summary of differences between Cystic hygromas presenting in early and late gestation [2–7, 16, 17]

Cystic hygromas of 1st and early 2nd trimester	Cystic hygroma/lymphangioma in 3rd trimester
Early gestational age at presentation	Isolated cystic hygroma usually presents during the third trimester
Usually located in the posterior triangle	Often with previously normal sonographic studies earlier in gestation
Often associated with chromosomal abnormalities in up to 60%, structural anomalies, hydrops fetalis, a high incidence of intrauterine death, and rare postnatal survival	Usually located anteriorly or anterolaterally in the anterior cervical triangle. Other locations are the axilla (20%), retroperitoneal/ intraabdominal organs (2%), limbs/bones (2%), and mediastinum (1%) [23]
Associated abnormalities: [16, 17]. Chromosomal: Most commonly Turner syndrome (45–50%), trisomies 13, 18, 21. Single gene disorders: Noonan, multiple pterygium, Robert syndrome, and polysplenia etc.	Association with chromosomal and structural abnormalities is unusual
High mortality rate	Prognosis is good

Case 1 (Fig. 1a, b, c, d)

A primigravida of 18 years showed a large multi-septate cystic lesion of size > 10 × 12 cm over the right antero-lateral chest wall of the fetus at 24 weeks of gestation on prenatal ultrasound. As there were no other major malformations, signs of hydrops, karyotype abnormality, or pressure effects, the prognosis appeared to be good. The fetus was monitored by serial ultrasound during which there was no undue enlargement or compression of adjacent structures. Since the mass was huge and obstruction was anticipated if allowed for vaginal delivery, a cesarean section was done at term. The baby was a male weighing 3.88 kg at birth with a huge, cystic mass of size 18 × 14 cm over the right lateral chest wall, axilla, and extending over the back. The skin over the mass was intact, there were no intra-thoracic or intra-abdominal extensions or attachment to the underlying bone. The baby was followed up with postnatal ultrasound and intermittent aspirations. By one year, three aspirations were done to bring down the size along with 3 cycles of Bleomycin sclerotherapy. At 16 months of age, surgical excision with grafting was done and the lesion healed with no major disfigurement.

Case 2 (Fig. 2a, b, c, d)

The fetus of this second gravida aged 28 years, showed an isolated cystic swelling of 4 × 3.5 cm over the right cheek extending down to the upper neck at 33 weeks of gestation. Pregnancy was followed up with serial ultrasounds and a male baby delivered at term by a spontaneous vaginal delivery. Ultrasound of the neonate showed a septated anechoic lesion of 2.8 × 3.8 cm over the right cheek and neck, closely abutting the right parotid. The cyst was confined to the subcutaneous plane and there was no evidence of pressure effects on adjacent structures. The baby was followed up postnatally with conservative management with propranolol from day 15 up to 4 months with subsequent bleomycin for 4 cycles.

Case 3

A 26 years second gravida presented with a fetus of 24 weeks showing an isolated cystic, septated lesion of 4.3 × 3 cm over the left anterolateral compartment of the neck. Pregnancy was serially monitored with ultrasounds until 39 weeks. There were no changes in size or appearance of the mass. The baby which was delivered by cesarean at term had a diffuse 5 × 6 cm cystic mass over left anterolateral neck. Postnatal management was done with Propranolol sclerotherapy.

Discussion

The sonographic appearance of cystic hygromas is fluid-filled cystic spaces divided by fine septae. Differentials to be considered when these involve the neck are encephalocele, cystic teratoma, and goiter. MRI can aid in defining the size, extent, and infiltration into adjacent structures [8]. All cases described here presented in the late 2nd or 3rd trimester. Once a cystic hygroma is detected, a search for signs of nonimmune hydrops and associated anomalies should be done. None of our cases had hydrops or associated malformation. Although karyotyping is strongly recommended in the 1st trimester, testing 2nd-trimester lesions in other areas may not be emphasised. We did fetal karyotyping by amniocentesis in Case 1 which was reported as normal.

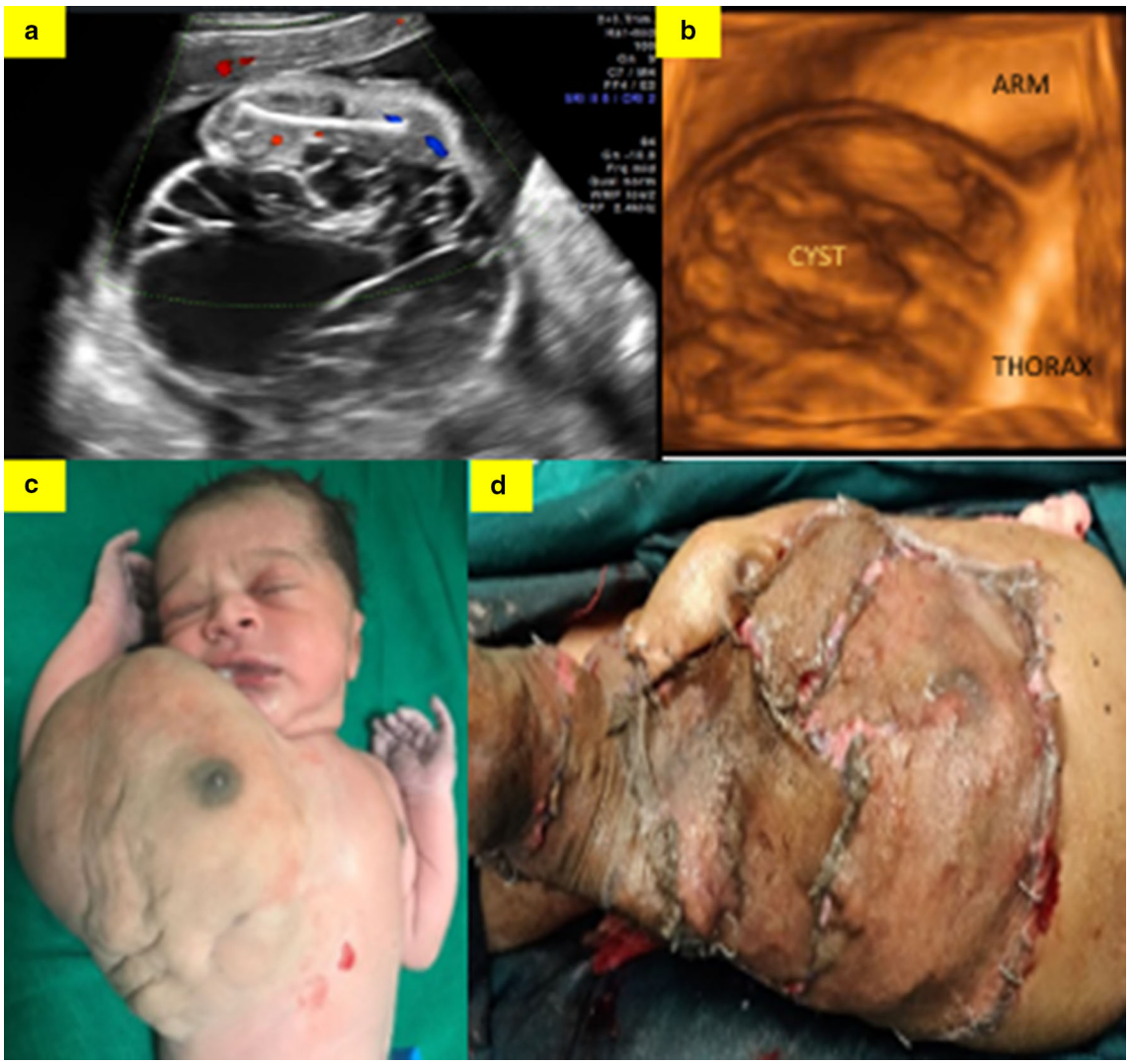

Fig. 1 Case 1 Ultrasound scan showing large cyst with multiple septations over the right thorax extending up to the axilla and lower costal margin and wedged between the right arm and lateral chest wall. There were no solid components, no vascularity on color doppler Doppler, no involvement of underlying bone, no effusions and no intrathoracic or intra-abdominal extensions. b 3-D scan showing the origin of the cystic mass from the lateral thoracic wall below the right arm with no attachment to underlying bones including ribs, spine and long bones. c Postnatal appearance of the lymphangioma which was irregular in shape with a smooth surface, cystic in consistency and was freely mobile. d After surgical excision

Isolated cystic hygromas are followed up by serial ultrasound to note for changes and pressure effects. Delivery by cesarean section is recommended in huge masses likely to cause obstructive labor. If there are concerns about airway compromise at birth, consideration should be given for an EXIT procedure [9]. In case 1, as the lesion was huge, the baby was delivered by ceserean section. Case 2 delivered vaginally and Case 3 had ceserean.

In utero drainage by ultrasound-guided cyst aspiration has been described albeit with a lack of benefit and risk of repeated aspirations. Postnatal treatment depends on site, size, progression, and disability/disfigurement caused. Surgical excision is the standard treatment and long-term outcome depends on the completeness of resection. Injury to cranial nerves and Horner syndrome are reported complications. Involvement of larynx, trachea, or floor of the mouth may necessitate tracheostomy/tube placement [10, 11]. Small to moderate lesions can be managed with sclerosing agents like OK-432, propranolol, sirolimus and bleomycin or with Co2 laser and radiotherapy [12–15]. In case 1, aspirations were done thrice postnatally along with 3 cycles of Bleomycin followed ultimately with surgical excision. Case 2 received propranolol followed by bleomycin and Case 3 had propranolol cycles.

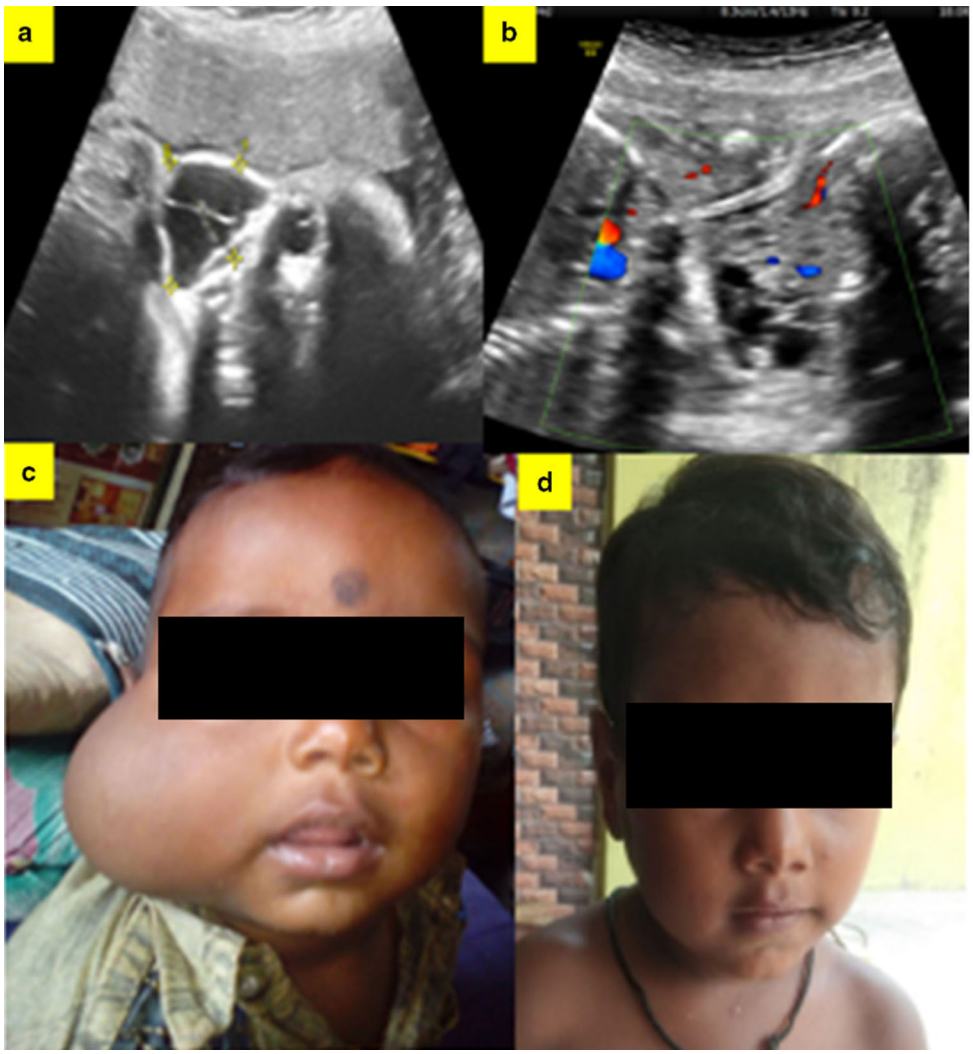

Fig. 2 Case 2 An ultrasound appearance of cystic hygroma over cheek with thin septae, no solid components. There were no deeper extensions or encroachment to the underlying bone. b No vascularity on color doppler. c Postnatal appearance. d At 3 years of age after treatment with sclerosing agents

Conclusion

Isolated fetal lymphangiomas have an excellent prognosis and should be followed antenatally with serial ultrasounds. Route of delivery is guided by location, size and pressure effects on adjacent structures. Small to medium lesions can be managed conservatively by postnatal sclerotherapy, while large masses require surgical excision.

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