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**Case Report**

# Fetal thoraco-abdominal lymphangioma: a case report

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## ABSTRACT

Lymphangiomas are rare congenital malformations of the lymphatic system. Authors present a case with giant, septated, axillary thoraco-abdominal lymphangioma. Diagnosis was made at 19 weeks' gestation by antenatal ultrasonography. The case underwent prenatal expectation treatment.

**Keywords:** Antenatal ultrasonography, Congenital malformations, Fetal lymphangiomas

## INTRODUCTION

Lymphangiomas are defined as rare congenital malformations of the lymphatic system.[1] Despite their benign histological features, lymphangiomas tend to grow rapidly and invade into the surrounding tissues such as muscle and bone.[2] They can be located in different anatomic regions; the neck being the most common accounting for 75% of the cases. These are known as cystic hygromas. The remaining cases are located at the axillary region, extremities, anterior abdominal wall and other parts of the body.[3]

Although cystic hygromas are common and associated with chromosomal abnormalities and poor outcome, the true incidence of fetal axillary lymphangioma is unknown and there are only few case reports in the literature. It develops late in the first trimester or early in the second trimester. It can occur in healthy children and in children with chromosome abnormalities or malformations.[4] Prenatal diagnosis is important as it permits planned delivery, allows postnatal resuscitation, and improves prognosis.[5]

Authors report a rare live born case with giant, septated, axillary thoraco-abdominal lymphangioma who underwent prenatal expectation treatment.

## CASE REPORT

A 25-year-old gravida 2 woman was referred to our clinic at 12 weeks gestation for pregnancy control. The patient did not have any antenatal care until admission to the clinic. She did not take folic acid supplements neither before the pregnancy nor during the pregnancy. She was non-smoker and non-drinker. There was no systemic illness and operation in her past medical history. At the second visit, ultrasonographic examination showed a 19 weeks gestation, singleton, alive fetus who had a mass derived from the left axillary region which was extending to the anterior and posterior thoracic wall with fluid-filled cavities about 12 cm in size. Fetal biometry was appropriate for gestational age. Placenta and amniotic fluid were normal. No other fetal anomalies were identified. Color velocity imaging demonstrated no flow through the mass. There was a multilocular, subcutaneous cystic mass measuring 3.9×3.2×3.1 cm, located on the left chest wall and left upper arm of the fetus laterally

extending to the abdomen (Figure 1). Prenatal screening for Down's syndrome trans-nucallucency was done and showed that the fetus was normal. After prenatal counseling, the parents refused amniocentesis and opted to continue the pregnancy.

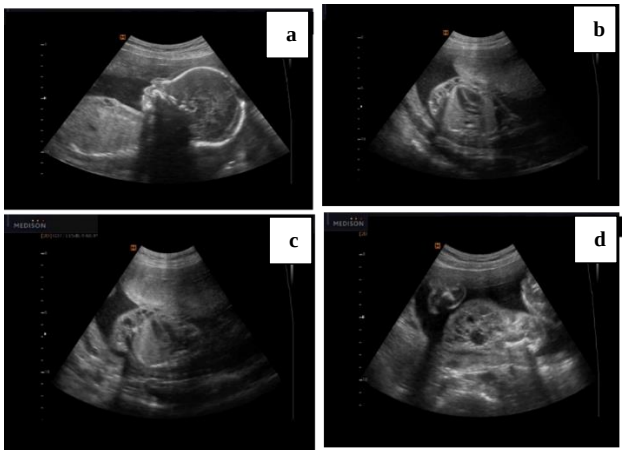


**Figure 1: Ultrasound at 19 weeks gestation showing subcutaneous cystic mass.**

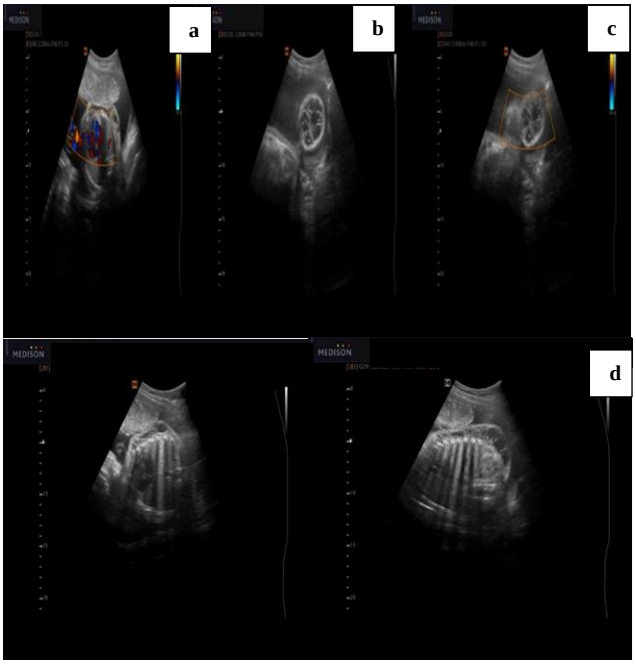


**Figure 2: Ultrasound at 40 weeks gestation showing the cystic mass.**

The pregnant woman and her husband denied any family history of genetic disorders, tumors, or unusual lymphatic or skin lesions. Prenatal care began at 12 weeks gestation; however, the nuchal translucency measurement was performed. Serial sonographic studies showed appropriate fetal growth, while the volume of the cyst did not increase. The cyst was lucent without internal echoes, but substantial intervening solid septations were evident. Color velocity imaging demonstrated no blood flow through the mass. At 40 weeks gestation ultrasound showed that the cystic mass had reached 6 ×7.2×70 cm (Figure 2). With the diagnosis of fetal thoraco- abdominal wall lymphangioma, elective cesarean section was performed at 40 weeks gestation, due to concerns about dystocia and fetal trauma. A 4.250 kg male infant was delivered with Apgar scores of 6 and 9 at 1 and 5 minutes respectively. At delivery, ultrasound examination confirmed the diagnosis of the mass. The baby had a 14×9×9 cm, soft cystic mass in the left anterior chest wall area, which extended to the left upper arm (4×3×3 cm). No other structural anomalies were visually identified. Umbilical cord blood taken at delivery revealed a normal karyotype (46, XY). Chest computed tomography performed on postnatal day 1 showed a 4.2×4.2×4.1 cm, well-marginated cystic mass with multiple septation. This was found at the left lateral chest wall and extended to the left upper arm, but not the upper neck (Figure 3).

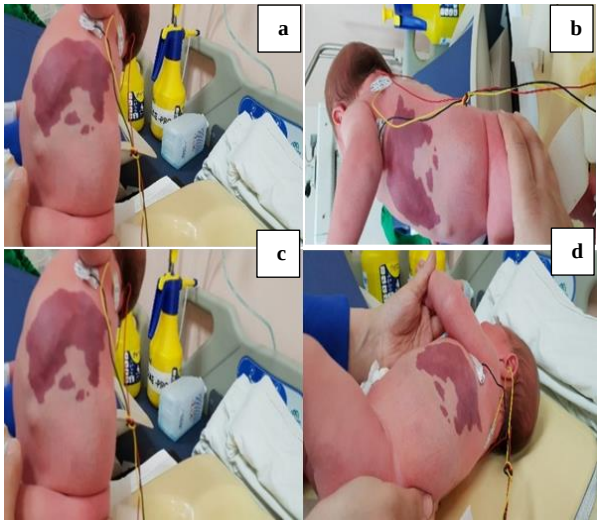


**Figure 3: Well-marginated cystic mass with multiple septation at the left lateral chest wall that extended to the left upper arm.**

The mother's postoperative course was uncomplicated. Her vital signs, routine blood and urine tests were found normal. She was discharged 3 days after the cesarean delivery. At 3 days of age, the infant underwent circumcision.

## DISCUSSION

Lymphangioma may result if a defect or obstruction occurs during the development of the lymphatic system which occurs by the end of the 5 weeks' gestation.[6] Two main views concerning the embryonic development were suggested. The first suggested that the lymphatic system develops as diverticula of the endothelium of the veins. The second claimed that the lymphatic channels develop from clefts in the mesenchymal tissue.[6] The insufficient drainage of lymphatics into the venous system is the leading mechanism of cystic hygromas at the fetal neck. However, lymphangiomas may occur in other parts of the fetal body such as axillary, thorax and extremities.[7]

*Lymphangiomas can be classified into three types*

1. Simple lymphangiomas, made up of lymphatic capillaries
2. Cavernous lymphangiomas, consisting of larger lymphatic vessels with a fibrous adventitia
3. Cystic lymphangiomasor hygromas, comprising multiple cysts of varying size.

All types may coexist in the same lymphangioma.[8]

The diagnosis of lymphangioma is made by ultrasonographic examination. The cyst may be loculated, septate or of complex structure.[9] Its size varies from millimeters to enormous cysts.[2] In the ultrasonographic scan, the fluid in the cyst may be seen either anechoic, internal echoes or fluid-filled levels with variable degrees, which is mainly due to bleeding and fibrin deposition.[10] For a fetal lymphangioma diagnosed during the prenatal period, the overall prognosis is poor with a mortality rate ranging from 50% to 100%.[11] Karyotypic abnormalities and different malformation could happen in more than 50% of patients affected by nuchal cystic hygromas.[11] In the present case, there was no chromosomal abnormality. Fetal sonography imaging identified the lymphangioma and defined its anatomical location and extent. Sonography has been considered as the imaging modality of choice for the prenatal assessment of fetal abnormalities since it is accurate, safe, easy to use and has low cost. However, some of its disadvantages include operator-dependency and limited field of view.[12] Magnetic resonance imaging (MRI) has become useful for prenatal diagnosis especially for the assessment of complex fetal abnormalities. MRI allows for more comprehensive imaging of the fetus because of its larger field of view and accurate delineating of the lesion extent.[12]

For the postnatal treatment of lymphangioma, surgical extirpation, with careful preservation of involved structures is the treatment of choice. Large but localized lymphangiomas can be completely excised. However, the surgical treatment of diffuse and multiple lymphangiomas is difficult and associated with high morbidity and mortality.[5] Hence, the injection of a sclerosing agent could be considered appropriate in such cases. The use of intralesional bleomycin, sclerotherapy with OK-432, or percutaneous embolization with Ethibloc was effective.[5]

## CONCLUSION

In conclusion, if lymphangioma is detected during the prenatal period by sonographic examination, detailed evaluation should be performed for potential associated abnormalities. Karyotyping must be offered. This will help in parental counseling and proper management.

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