International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Ghazal K et al. Int J Reprod Contracept Obstet Gynecol. 2023 Oct;12(10):3154-3159
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20232964

Case Series

Prenatal diagnosis of fetal lymphangiomas and outcome in all trimester: case series and literature review

Kariman Ghazal[1,2,3]*, Jihad E. Hassan[1,2,3], Annie Tachdjian[1], Joelle Azzi[4], Kawsar Diab[1]

[1]Department of Obstetrics and Gynaecology, Faculty of Medical Sciences, [4]Department of Pharmacy, Faculty of Public Health, Lebanese University, Beirut, Lebanon
[2]Department of Obstetrics and Gynaecology, Rafik Hariri University Hospital, Beirut, Lebanon
[3]Department of Obstetrics and Gynaecology, Al Zahraa Hospital University Medical Centre, Beirut, Lebanon

Received: 11 August 2023
Revised: 05 September 2023
Accepted: 06 September 2023

***Correspondence:**
Dr. Kariman Ghazal,
E-mail: ghazal_kariman@hotmail.it

ABSTRACT

Lymphangiomas “cystic hygromas" is a lymphatic malformation that can develop in a variety of locations on a developing fetus, most commonly in the neck, axilla, abdominal wall, mediastinal, inguinal, and retroperitoneal regions. Fetal aneuploidy, hydrops fetalis, structural abnormalities, and intrauterine fetal mortality have all been linked to lymphangiomas. We report case series of cystic hygroma in all trimester with there Obstetric outcomes which ultrasound imaging was used to identify these cases.

Keywords: Fetal tumors, Cystic hygroma, Karyotype, Prenatal ultrasound, Cordocentesis, Fetal aneuploidy

INTRODUCTION

Rare congenital abnormalities of the lymphatic system are referred to as lymphangiomas. Despite having benign histological characteristics, lymphangiomas have the propensity to spread quickly and infiltrate nearby tissues including bone and muscle. They can be seen in a variety of anatomical locations, with the neck accounting for 75% of occurrences. The term "cystic hygromas" refers to these. The remaining cases are spread across the body in various locations, including the extremities, anterior abdominal wall, and axillary area. Cystic hygromas are prevalent and linked to chromosomal abnormalities and poor outcomes, although there are few case records for these tumors.[1-3]

It begins to form during the end of the first trimester or the beginning of the second. They are made up of localized lymphatic system abnormality centers. The origin of this anomaly is explained by three different explanations. The first postulates that normal development of the primitive lymph channels is blocked or stopped during embryogenesis, the second that the primitive lymphatic sac does not connect to the venous system, and the third that lymphatic tissue develops in the incorrect location during embryogenesis.[4-6]

According to the extent of the lymphatic cavities contained, lymphangiomas are categorized as microcystic (capillary lymphangiomas), macrocytic (cavernous lymphangiomas), and cystic hygromas. Bill and Summer proposed the idea that lymphangiomas and cystic hygromas are variations of the same entity 1965 and that a tumor's classification depends on where it is found in the head and neck. A lymphangioma presents as a cavernous lymphangioma when it is restricted to relatively dense tissue, like the tongue, but develops as a cystic lesion when it grows in the neck's comparatively lax fascia. On the other hand, 90% or so of the lymphangiomas in the head and neck are cystic hygromas. The axilla, shoulder, chest wall, mediastinum, abdominal wall, and thigh are additional typical locations outside of the head and neck.[7-10]

We provide a case series of cystic hygromas in all trimesters along with their obstetric outcomes. These patients were found via ultrasound imaging.

CASE SERIES

Case 1

Prenatal care

By using ultrasound at 11 weeks, a 24-year-old G1P0 lady showed increased nuchal translucency thickness and cystic hygroma on the foetus (Figure 1). No smoking and no consanguinity.

Outcome

The woman lost her pregnancy ten days later.

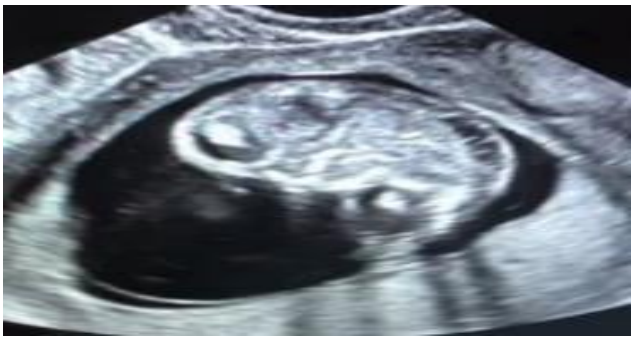

Figure 1: Foetus 11 weeks showed nuchal translucency thickness and cystic hygroma.

Case 2

Prenatal care

A cystic hygroma was detected during the 10-week ultrasound of a 24-year-old G1P0 woman (Figure 2). No smoking and no consanguinity.

Outcome

The patient lost her pregnancy.

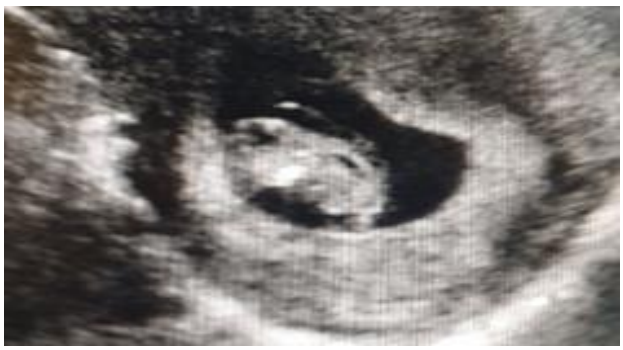

Figure 2: 10-week Foetus with cystic hygroma and diffuse thickness.

Case 3

Prenatal care

11-week ultrasound results of a 25-year-old G1P0 lady with a cystic hygroma and short limbs. No smoking and no consanguinity (Figure 3).

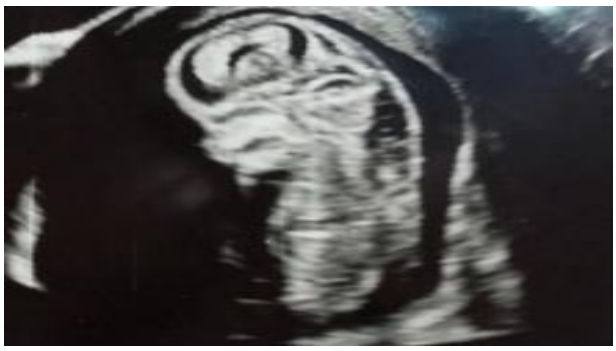

Figure 3: 11-week foetus with translucency thickness by ultrasound.

Outcome

The woman lost her fetus (Figure 4).

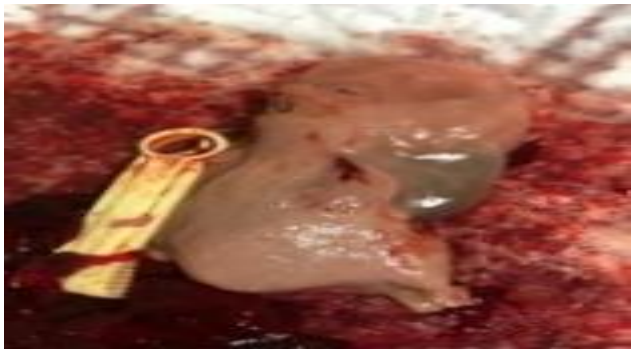

Figure 4: Fetus with short limbs.

Case 4

Prenatal care

A 24-year-old G2P1 woman. The ultrasound images revealed increased nuchal translucency thickness at 12 weeks gestation. The skin is elevated along the back because of subcutaneous oedema. (cystic hygroma) (Figure 5). No smoking and no consanguinity.

Outcome

The woman experienced pregnancy loss.

Case 5

At 12 weeks pregnant, a 25-year-old gravida 2 lady was sent to our clinic for pregnancy control with pelvic

ultrasound (Figure 10a). Neither before nor during her pregnancy did she take folic acid pills. She did not consume alcohol or smoke. Her prior medical history did not include any operations or systemic illnesses.

History

The second visit's ultrasonographic examination revealed a singleton, 19-week-old fetus that was still alive and had a tumor that originated in the left axilla and extended to the anterior and posterior thoracic walls, with cavities filled with fluid that were about 12 cm in diameter. For the gestational age, fetal biometry was appropriate. Amniotic fluid and the placenta were healthy. There were no additional fetal malformations found. No flow through the mass was visible on a color velocity image. On the left chest wall and left upper arm of the fetus, there was a multilocular, subcutaneous cystic mass measuring 3.93×23.1 cm that laterally extended to the belly (Figure 6).

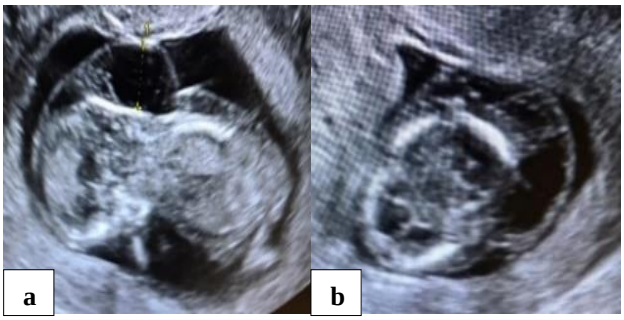

Figure 5: Fetus at 12 weeks skin is elevated along the back because of subcutaneous oedema.

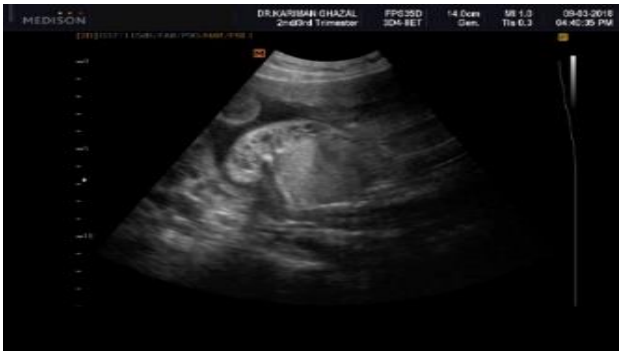

Figure 6: Left chest wall and left upper arm of the fetus, there was a multilocular, subcutaneous cystic mass measuring 3.93×23.1 cm that laterally extended to the belly.

The fetus passed the prenatal test for Down's syndrome trans-nuchal lucency and was found to be healthy. The parents made the decision to carry on with the pregnancy after prenatal counselling and decline an amniocentesis. The expectant mother and her husband denied that any genetic diseases, cancers, or odd lymphatic or skin lesions ran in their family. Serial sonographic examinations revealed adequate fetal growth, but the cyst's volume remained constant. The cyst was translucent and without internal echoes, but there were noticeable thick solid septations in between. Blood flow through the bulk was not visible on color velocity imaging. An ultrasound at 40 weeks of pregnancy revealed a cystic tumor that measured 6×7.2×70 cm (Figure 7).

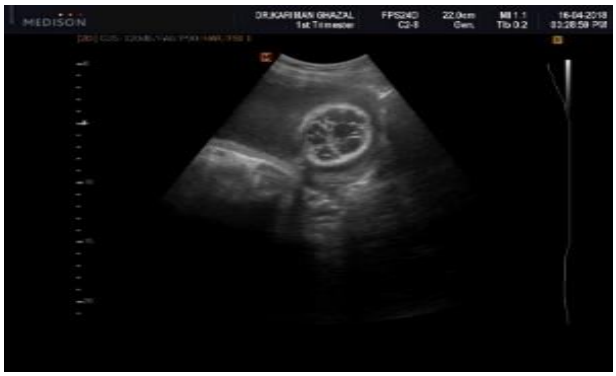

Figure 7: Well-marginated cystic mass with multiple septation at the left lateral chest wall that extended to the left upper arm and abdomen.

Outcome

An elective caesarean surgery was carried out at 40 weeks gestation with the diagnosis of foetal thoraco-abdominal wall lymphangioma.

A 4.250 kg male baby was born with Apgar ratings of 6 and 9, respectively, at 1 and 5 minutes. An ultrasound during birth verified the mass's diagnosis. The infant's left anterior chest wall area contained a soft cystic mass measuring 14 by 9 by 9 cm that also reached the left upper arm (4 by 3 cm). Visual inspection revealed no other structural irregularities. A normal karyotype (46, XY) was found in the umbilical cord blood drawn during delivery. On the first postnatal day, a chest computed tomography revealed a well-marginated, 4×24×24.1 cm cystic mass with numerous septa. The left lateral chest wall was the location of this, and it reached the left upper arm but not the upper neck (Figure 8).

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 caesarean delivery and the newborn underwent circumcision.

Case 6

At 31 weeks of pregnancy, a 30-year-old lady who was gravida 1 para 0 came into our antenatal clinic for a routine scan for the first time due to pelvic abdominal pain. The patient did not get any antenatal treatment. She had a high socioeconomic standing. She did not consume alcohol or smoke. Her prior medical history did not include any operations or systemic illnesses. The patient's vital signs were determined to be normal (Figure 9).

History

A singleton foetus with breech presentation and biometry compatible with gestational age was seen on ultrasound at 31 weeks. A thorough examination of the face led to the identification of a multicystic tumor in the right zygomatic region. The mass is 12 cm by 10×10 cm in size. It was composed of a mixture of solid and anechoic components, thick, avascular septations that were asymmetrical, thin-walled, multiseptated, and solid components that were dispersed throughout the multicystic regions (Figure 9). It spanned the entire fetal neck region, extending from the chin to the front of the neck (Figure 9). The lump was still expanding, causing the fetal neck to extend too far and the face to bulge, according to serial scans of the embryonic neck The mass's edges were clearly defined and showed signs of infiltration of the surrounding tissues (Figure 9).

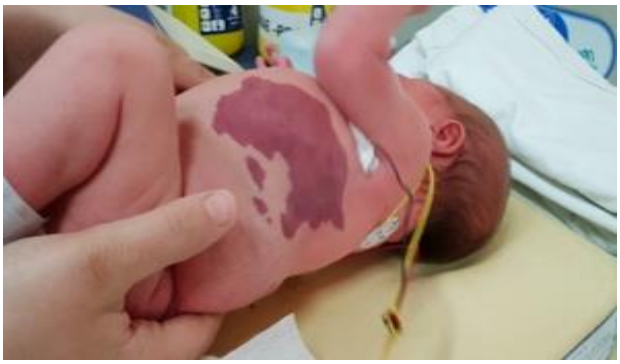

Figure 8: On left lateral chest wall was the location of the mass, and it reached the left upper arm but not the upper neck.

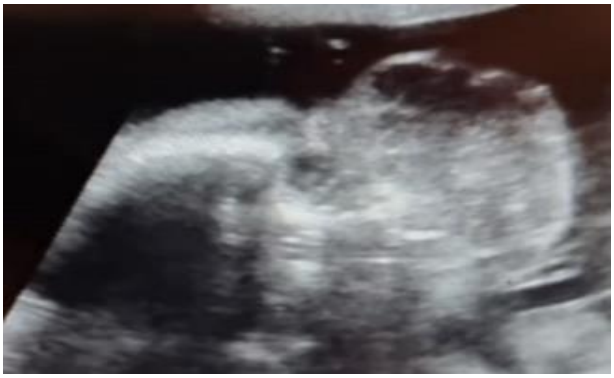

Figure 9: Multicystic tumor in the right zygomatic region of the fetus.

The placenta and amniotic fluid were both in normal range, and the fetal biometry matched the gestational age. There were no additional fetal malformations found. No flow through the mass was visible on a color velocity image.

The head, face, and neck region of the body had normal anatomy. The parents were offered MRI and karyotyping, but they declined.

Outcome

Ten days later, the patient was readmitted to the prenatal clinic once more due to the absence of fetal movements. Fetal demise was discovered during an ultrasonographic evaluation. Although there were signs of hydrops fetalis and the lump appeared to be the same size, the amniotic fluid was confirmed to be normal. The newborn was delivered vaginally after labour was induced with prostaglandin and oxytocin infusion. He had gigantic lymphangiomas on the neck, face, short neck, and hands (Figures 10 and 11).

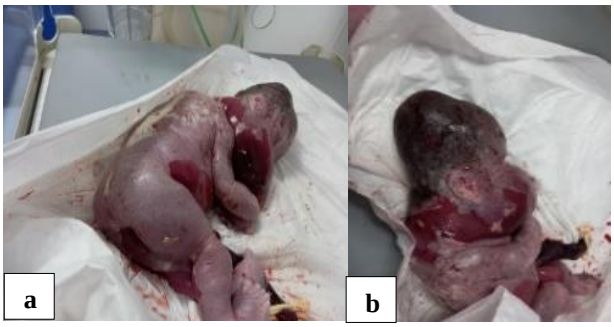

Figure 10: From the neck to the newborn since the anterior zygoma of the face, the mass was present.

Figure 11: The new born macereted with mass on the neck and no cystic harm.

Due to religious beliefs, the parents reject autopsy.

DISCUSSION

A malfunction or obstruction in the lymphatic system's development, which happens towards the end of the fifth week of pregnancy, may lead to a lymphangioma. Cystic hygromas at the fetal neck are mostly caused by inadequate lymphatic drainage into the venous system. The inability of the jugular lymphatic sacs to empty into the internal jugular vein causes the development of a cystic hygroma, which usually occurs between the sixth and ninth weeks of pregnancy. The jugular lymphatic blockage sequence and hydrops fetalis may result from this, and the lymphatic sacs are likely to expand into cystic spaces.[6,11-13,15]

About 75% of the time, it affects the neck, frequently bilaterally and asymmetrically, usually in the posterior and

lateral rather than the anterior regions. CH has been connected to fetal aneuploidy, morphological abnormalities, hydrops fetalis, and intrauterine fetal mortality.[17] When abortions are taken into account, the incidence rate of 1 in 6000 to 1 in 16,000 live births (1/875) is projected to be significantly higher. CH can occur in the extremities but the posterior neck, axilla, mediastinum, groin, and retroperitoneum are responsible with 67%, 75%, 20%, and 1% of cases, respectively.[11,12,14-17]

Fetal CH instances can be categorized into two groups: those identified earlier in pregnancy, which are often lymphangiomas alone, like in our first four cases, and those identified earlier in pregnancy, which are frequently associated with other abnormalities and a poor outcome. These may be specifically related to Noonan syndrome, Turner syndrome, cardiac problems, and fetal hydrops. When a woman is severely disabled, hydrops and progressive peripheral lymphedema (PLE) develop, which can cause an early miscarriage.[12,15,17-20]

In later gestation CH, which most likely reflects a lymphangioma without aneuploidy or other fetal anomalies, has a good prognosis like our 5th case. The baby operated at 2.5 years with exicision of the mass with careful preservation of involved structures. Hence, the injection of a sclerosing agent could be considered appropriate in this case. The use of intralesional bleomycin, sclerotherapy with OK-432, or percutaneous embolization with ethibloc was effective.[5,18,19]

The prognosis is thought to be even worse than with non-septated CH when it appears septated.[8,20,21]

Our 6 case presented like cavernous lymphangioma. On the tongue or the floor of the mouth, as in this case, cavernous lymphangiomas of the head and neck is possible for lobulated cystic. Swallowing problems are brought on by lymphangiomas that impact the oral cavity, oropharynx, and/or hypopharynx. Tongue might result in macroglossia, which can obstruct airway and difficulty to swallowing and death specially after delivery.[16,21]

A lymphangioma is diagnosed with an ultrasonographic examination also indicated its anatomical position and size. The cyst's structure might be complicated, loculated, or septate. Its cysts can range in size from millimeters to huge ones. The fluid in the cyst may seem either anechoic, internal echoes, or fluid-filled levels with varying degrees during an ultrasonographic scan. This is mostly because of hemorrhage and fibrin deposition. The general prognosis is dismal and the death rate for fetal lymphangiomas detected during pregnancy ranges from 50% to 100%.[2,9-11]

The use of magnetic resonance imaging (MRI) for prenatal diagnosis has increased, particularly for the evaluation of complicated fetal disorders.[12]

Although the lymphangiomas can be inherited as an autosomal recessive condition with a 25% recurrence rate when the karyotype is normal, aneuploidy has a very low chance of returning.[21]

CONCLUSION

We learn from this topic role and the importance to the ultrasound for diagnosis this defect for good treatment that the importance of multidisciplinary approach. A knowledgeable and conscientious sonographer is required for the prenatal diagnosis of lymphangiomas. A thorough and methodical study of the fetus is also necessary because of the connection with other defects. The obstetrician will be better equipped to advise the patient on the baby's prognosis, the delivery technique, and any extra testing like karyotyping and MRI. A multidisciplinary approach involving the obstetrician, neonatologist, and pediatric surgeon is crucial for the best outcome.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Chiaverini C, Benoit B, Bongain A, Chevallier A, Lacour JP. Prenatal ultrasonographic detection of an axillo-thoracic lymphangioma: an ethical dilemma. Prenat Diagn. 2003;23:946-8.
2. Goldstein I, Leibovitz Z, Noi-Nizri M. Prenatal diagnosis of fetal chest lymphangioma. J Ultrasound Med. 2006;25:1437-40.
3. Lo Magno E, Ermito S, Dinatale A, Cacciatore A, Pappalardo EM, Militello M, et al. Fetal cystic lymphangioma of the neck: a case report. J Perinat Med. 2009;3:12-4.
4. Lu D, Wang Y, Zeng W, Peng B. Giant fetal lymphangioma at chest wall and prognosis: case report and literature review. Taiwan J Obstet Gynecol. 2015;54:62-5.
5. Deshpande P, Twining P, O'Neill D. Prenatal diagnosis of fetal abdominal lymphangioma by ultrasonography. Ultrasound Obstet Gynecol. 2001;17:445-8.
6. Paladini, M Vassallo, G Sglavo, C Lapadula, M Longo, C Nappi. Cavernous lymphangioma of the face and neck: prenatal diagnosis by three-dimensional ultrasound Ultrasound Obstet Gynecol. 2005;26(3):300-2.
7. Allanson JE. Lymphangiomyoma. In: Stevenson RE, Hall JG, Goodman RM, et al. Human malformations and related anomalies. Vol 2. Oxford: Oxford University Case Report. Rare Tumors. 2011;3:e39.
8. Chervenak FA, Isaacson G, Tortora M. A sonographic study of fetal cystic hygromas. J Clin Ultrasound. 1985;13:311-5.
9. Judith E, Allanson MD. Lymphangioma. In Human malformation and related anomalies, Stevenson RE, Hall JG, Good-man RM (eds). Oxford University Press: New York, NY. 1993;288.

10. Rempen A, Feige A. Differential diagnosis of sonographically detected tumours in the fetal cervical region. Eur J Obstet Gynecol Reprod Biol. 1985;20:89-105.
11. Song TB, Kim CH, Kim SM. Fetal axillary cystic hygroma detected by prenatal ultrasonography: a case report. J Korean Med Sci. 2002;17:400-2.
12. Phillips HE, McGahan JP. Intrauterine fetal cystic hygromas: sonographic detection. AJR Am J Roentgenol. 1981;136:799-802.
13. Garden AS, Benzie RJ, Miskin M, Gardner HA. Fetal cystichygroma colli: antenatal diagnosis, significance, and management. Am J Obstet Gynecol. 1986;154:221-5.
14. Judith E, Allanson MD. Lymphangioma. In Human malformation and related anomalies, Stevenson RE, Hall JG, Good-man RM (eds). Oxford University Press: New York, NY. 1993;288.
15. Teague KE, Eggleston MK, Muffley PE, Gherman RB. Recur-rent fetal cystic hygroma with normal chromosomes: case reportand review of the literature. J Matern Fetal Med. 2000;9:366-9.
16. Bossert T, Gummert JF, Mohr FW. Giant cystic lymphangioma of the mediastinum. Eur J Cardiothorac Surg. 2002;21:340.
17. Nakazato Y, Ohno Y, Nakata Y. Cystic lymphangioma of the mediastinum. Am Heart J. 1995;129:406-9.
18. Langer JC, Fitzgerald PG, Desa D. Cervical cystic hygroma in the fetus: clinical spectrum and outcome. J Pediatr Surg. 1990;25:58-62.
19. Thomas RL. Prenatal diagnosis of giant cystic hygroma: prognosis, counseling, and management: case presentation and review of the recent literature. Prenat Diagn. 1992;12:919-23.
20. Tanriverdi HA, Hendrik HJ, Ertan AK. Hygroma colli cysticum: prenatal diagnosis and prognosis. Am J Perinatol. 2001;18:415-20.
21. Schuster T, Grantzow R, Nicolai T. Lymphangioma colli – A new classification contributing to prognosis. Eur J Pediatr Surg. 2003;13:97-102.

Cite this article as: Ghazal K, Hassan JE, Tachdjian A, Azzi J, Diab K. Prenatal diagnosis of fetal lymphangiomas and outcome in all trimester: case series and literature review. Int J Reprod Contracept Obstet Gynecol 2023;12:3154-9.