Thoraco-lumbar hemangiolymphangioma diagnosed antenatally by ultrasonography*

By Mutya S.A. Salvacion, MD, DPOGS and Cleofe B. Medina, MD, FPOGS, FPSUOG

Department of Obstetrics and Gynecology, Makati Medical Center

ABSTRACT

This is a case of a fetus with a complex cystic structure on the mid-thorax to the lumbar area detected by ultrasonography at 23 weeks age of gestation. There were no other structural abnormalities noted. The fetal Doppler of the middle cerebral and umbilical arteries were normal. The increase in size of the cystic mass, diagnosed as lymphangioma, and the appearance of pleural effusion at 27 weeks age of gestation prompted further surveillance with magnetic resonance imaging. It showed an extensive subcutaneous mass involving the right thoraco- lumbar region, to consider hemangioma. Expectant management, bringing the pregnancy close to term as possible, was planned. However, the progression of the effusion to the bilateral hemithorax and presence of fetal ascites led to the cesarean delivery of a live preterm male with a birthweight of 1,885 grams (4 lbs 1 oz), maturity index of 29 weeks and an Apgar score of 4, 7, 8 at the first, fifth and tenth minute of life. There was a 15 x 13 cm hemangiolymphangioma on the right thoraco-lumbar area. An ultrasound-guided thoracentesis was done to help alleviate fetal distress. The infant was observed in the neonatal intensive care unit and was sent home stable. Presently, the hemangiolymphangioma is gradually resolving.

Keywords: Vascular malformation; lymphangioma; hemangioma; hemangiolymphangioma; antenatal ultrasound; fetal magnetic resonance imaging

INTRODUCTION

Vascular malformations are lesions of abnormal vascular development. One type is hemangiolymphangioma, which is a congenital malformation of the lymphatics and blood vessels. Prenatal diagnosis of this condition is rare. With the help of ultrasonography and magnetic resonance imaging, there is now the possibility to observe its progression in utero and do careful planning on the course of pregnancy. This is a case of a hemangiolymphangioma detected at 23 weeks age of gestation by ultrasonography.

CASE HISTORY

A 38-year-old, Gravida 1 Para 0, who had history of infertility and hypothyroidism (clinically euthyroid), was seen and monitored in our center. The first trimester was unremarkable except for episodes of vaginal spotting and findings of subchorionic hemorrhage on ultrasound. This was treated with progesterone and tocolytics, and eventually resolved. Succeeding scans were unremarkable.

At 23 weeks age of gestation, during a routine congenital anomaly scan, a complex cystic structure was detected on the fetal right thoracic area. Ultrasound showed an avascular, complex, cystic structure, partitioned by thick and thin fibrous bands measuring 13.1 mm thick and located at the mid-thorax to the lumbar area (Fig. 1). Cystic hygroma was considered. The fetal weight was appropriate for gestational age and there were no other gross fetal structural anomalies seen.

At 25 weeks age of gestation, follow-up scan showed the complex cystic structure arising from the right axilla to the right lumbar area, measuring 9.13 x

6.11 x 2.09 cm, to consider soft tissue mass, probably a lymphangioma. There was note of pleural effusion (Fig. 2). The pregnancy was managed expectantly.

At 27 weeks age of gestation, the patient was admitted for tocolysis because of uterine contractions. Fetal surveillance was done. Ultrasound showed the complex mass measuring 7.8 x 2.5 x 2.5 cm at the right thoracic area. There was pleural effusion at the right hemithorax, compressing the lung tissue on the right and pushing the heart more to the left side (Fig. 3). Fetal weight was appropriate for gestational age and the biophysical profile score is 8/8. The case was referred to a neonatologist and a perinatologist who did additional scans. Ultrasound showed the non-vascular solid-cystic mass from the axilla down to the lumbar level, to consider a lymphangioma. There was pleural effusion on the right, with mediastinal shift to the left. The pericardial fluid was within normal. Fetal doppler studies of the middle cerebral and umbilical arteries showed normal blood flow velocities based on Doppler waveforms and pulsatility indices (Fig. 4). The patient was referred to pediatric thoracic and cardiovascular surgery service (TCVS) to check

*Second Place, Philippine Society of Ultrasound in Obstetrics and Gynecology (PSUOG) Interesting Case Paper Contest, October 27, 2018, 3rd Floor Function Room, Dr. Jose Fabella Memorial Hospital, Philippine Blood Center Compound, Diliman, Quezon City

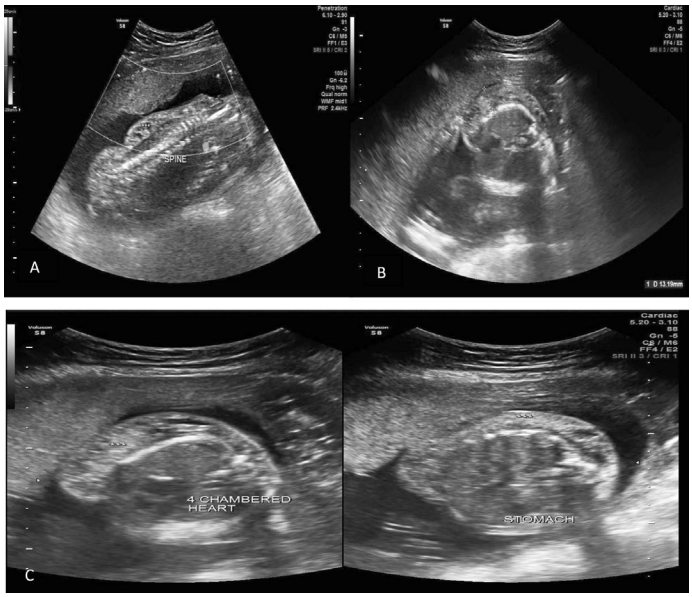

Figure 1. Ultrasound at 23 weeks age of gestation. A. Sagittal view: An avascular, complex mass (labeled with asterisk) at the right thoraco-lumbar region of the fetus; no involvement of the neck. B. Transverse view of the abdomen: cystic structure, partitioned by thick and thin fibrous bands, measuring 13.1 mm, to consider cystic hygroma. C. Transverse view of the fetal chest *(left picture)* and abdomen *(right picture)*, showing the location of the mass (labeled with asterisk), at thoracolumbar region; note that there is no extension to the organs adjacent to it.

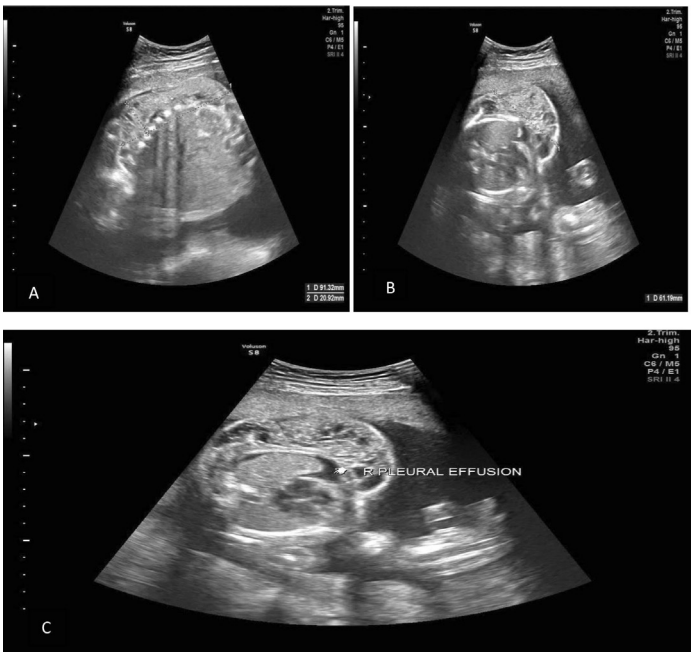

Figure 2. Ultrasound at 25 weeks age of gestation. *A and B.* Soft tissue mass, to consider lymphangioma, arising from the right axilla to the right lumbar area, measuring 9.1 x 6.1 x 2.0 cm. C. Transverse view of the chest, showing the right pleural effusion.

for fetal mass resectability. The pediatric TCVS requested for a magnetic resonance imaging (MRI) with contrast using Gadolinium. However, since no studies suggests the safety of gadolinium use in pregnancy, a plain fetal MRI was done instead. The MRI showed no cystic lesion was identified in the lung. There was a T2-W hyperintense and T1-W isointense subcutaneous mass with internal flow voids within identified in the right anterolateral thoraco-abdominal region, with a primary consideration of a hemangioma (Fig. 5). No intrathoracic, intra-abdominal or spinal involvement seen. The obstetrical plan was to bring the pregnancy close to term as possible. No intrauterine or invasive intervention was warranted at that time. Steroids were administered for fetal lung maturation.

At 29 weeks age of gestation, pelvic ultrasound showed the enlargement of the lymphangioma to 12 x 3.3 x 8.8 cm occupying the right hemithorax, from the axillary to lumbar region to the level of the kidney. No neck involvement noted. Ascites was seen above and below the fetal liver. The cardiac axis is deviated to the left at 54.5 degrees. The right hemithorax is filled with fluid compressing on the right lung tissue, approximately 28.1 mL (47 x 42 x 27 mm). Pleural effusion, approximately 1.86 mL (17.7 x 15.8 x 12.7 mm), was noted at the left hemithorax (Fig. 6). Sonographic fetal weight is large for gestation (1965 grams based on BPD, HC and FL, 99th percentile by Hadlock) and the maternal cervical length is 2.82 cm long. Due to the increasing fetal pleural effusion and ascites, the patient underwent classical cesarean section. A male preterm infant, weighing 1,885 grams (4 lbs 1 oz), with a maturity index of 29 weeks, had an Apgar score of 4, 7, 8 at the first, fifth and tenth minute of life, respectively. There was a 15 x 13 cm cystic mass on right thorax (Fig. 7). The baby was intubated and admitted to the neonatal intensive care unit where he was seen by the pediatric TCVS and pediatric surgery services. Ultrasound-guided right thoracentesis was done for the pleural effusion and approximately 35 mL of bloody fluid was drained. There were no microorganisms seen on the pleural fluid. The baby was referred to the pediatric hematology service and was advised magnetic resonance angiography of the mass when the baby become more stable. Treatment options using Interferon, sclerosing therapy and propanolol were considered but was not advisable at that time because of prematurity. The present plan was to observe and do further work up at the sixth month of life for possible chemotherapeutic options if the hemangiolymphangioma does not resolve. Nutritional building and monitoring was done until the baby was discharged on the third month of life. Presently, both mother and child are doing well. The hemangiolymphangioma is gradually resolving.

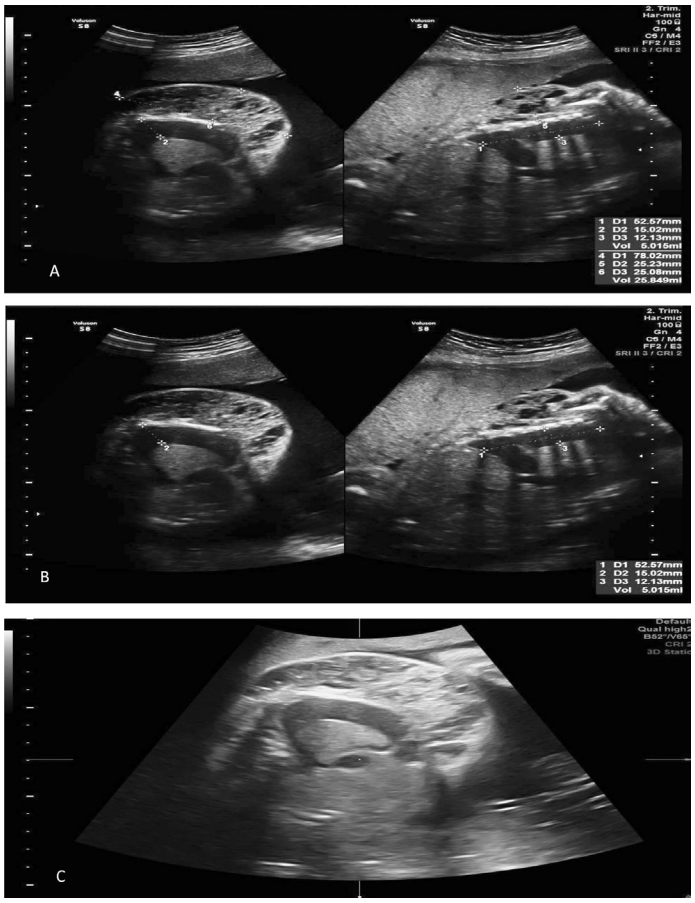

Figure 3. Ultrasound at 27 weeks age of gestation. *A.* Measurement of the thoraco- lumbar cystic mass, 78 x 25.2 x 25 mm. *B. and C.* Pleural effusion at the right hemithorax, pushing the fetal heart to the left.

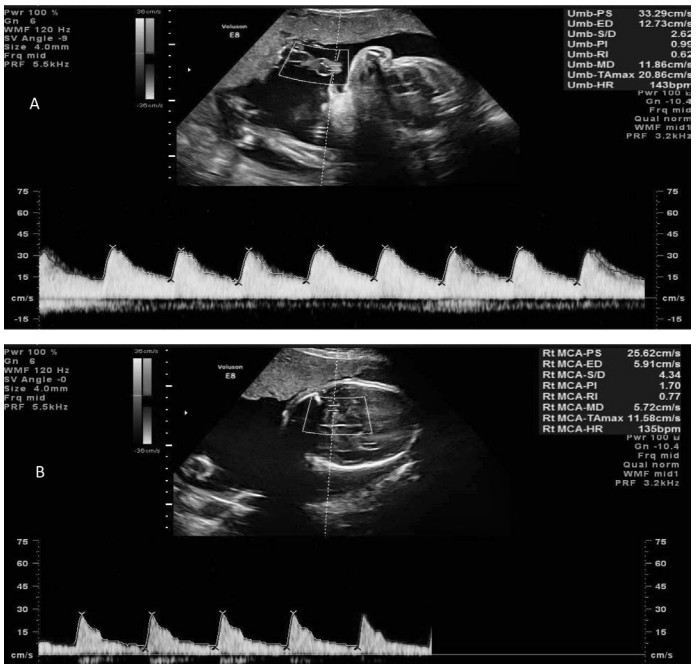

Figure 4. Doppler indices at 27 weeks age of gestation. Normal blood flow velocities on Doppler waveforms and pulsatility index. *A.* Umbilical artery PI = 0.99 (normal values PI = 0.81 – 1.56). *B.* Middle cerebral artery PI = 1.70 (normal values PI = 1.48 – 3.30).

CASE DISCUSSION

Vascular malformations are a heterogenous group of congenital blood vessel disorders. Previously, this was referred to as birthmarks. It is now subcategorized into two groups: vascular tumors, which are proliferative endothelial lesions, include hemangiomas; and vascular malformations, which are developmental aberrations of hematic or lymphatic vessels, include lymphatic malformations. Under this new classification, previous terminology like cystic hygroma is no longer used.[1-3]

Hemangiolymphangioma is a rare congenital malformation of the blood vessels and lymphatics. Under the new system of classification, this is grouped into two categories: hemangioma and vascular malformations. Based on its endothelial characteristics, hemangiolymphangiomas can be a lymphatic-venous or lymphatic-capillary malformation. It may occur anywhere in the body, such as the axilla, extremities, abdominal cavity and urinary bladder.[1,2,4]

The incidence of lymphangioma at birth was reported to be 1:6,000.[5] The prevalence of infantile hemangioma is unknown, but rates as high as 10% have been reported.[6] The incidence of hemagiolymphangioma, which is rarer, were only described on case reports.

In general, vascular anomalies are now being diagnosed prenatally using different imaging modalities. On ultrasound, a hemangioma possesses internal vascularity and may contain calcifications. It can be solid with variable echogenicities. On Doppler ultrasound, it is hypervascular. Lymphatic malformations present as multiseptated cystic masses with no internal vascularity or associated soft tissue mass on ultrasound. On Doppler, it is monophasic with no flow pattern.[7,8] The fluid may be anechoic with variable internal echoes or fluid levels, due to bleeding or fibrin.[9] A hemangiolymphangioma, on prenatal ultrasound, presents as a multilocular mass with both cystic and solid parts. It also has the tendency to grow rapidly and invade adjacent tissues. These features are unusual with pure hemangiomas or lymphangiomas.[10] In the case, the mass was not detected prior to 23 weeks gestation. This might be explained by the nature of the mass to grow rapidly following a normal first trimester scan. The avascular, complex cystic mass progressed and grew to size that caused fetal lung compression and mediastinal shift.

Magnetic resonance imaging (MRI) can help describe the anatomical extent and multicompartmental

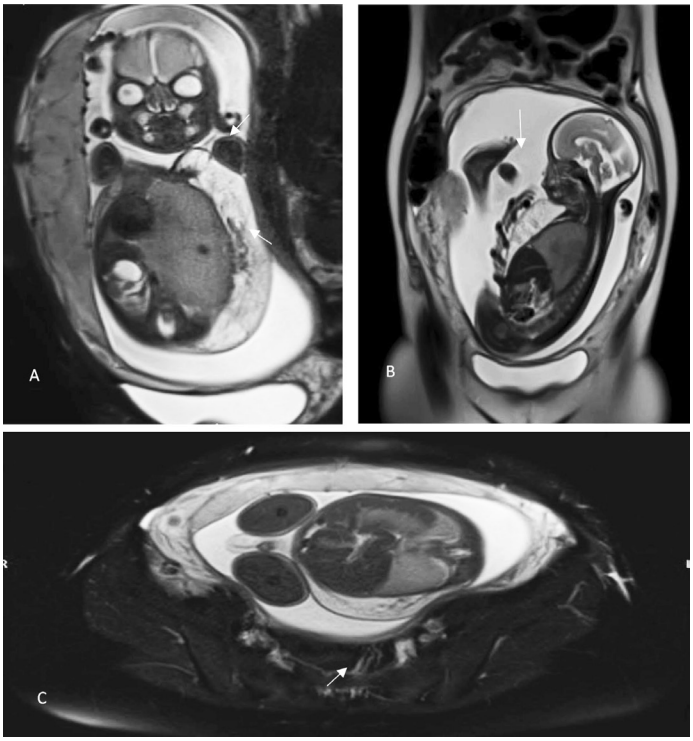

Figure 5. Fetal Magnetic Resonance Imaging (MRI – T2 images) at 27 weeks age of gestation. A & B. Axial view, T2-W hyperintense and T1-W isointense extensive subcutaneous mass (labeled with arrows) with internal flow voids within, involving the right thoracoabdominal region, primary consideration is a hemangioma. The diaphragm appears intact. C. Transverse view at the level of the fetal chest. Subcutaneous mass seen external to the lungs; no cystic lesion identified in the lungs. No intrathoracic, intra-abdominal or spinal axis involvement seen.

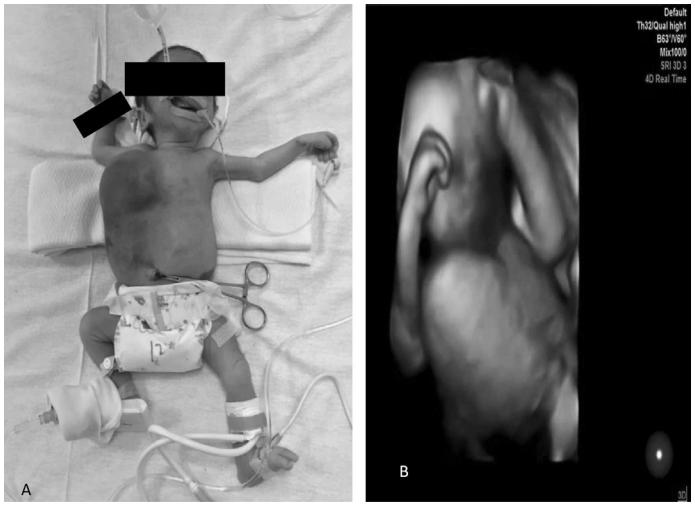

Figure 7. A. Male, preterm, infant with a hemangiolymphangioma at the right thorax, measuring 15 x 13 cm. B. 3D/4D rendering of the mass at 29 weeks age of gestation, for comparison.

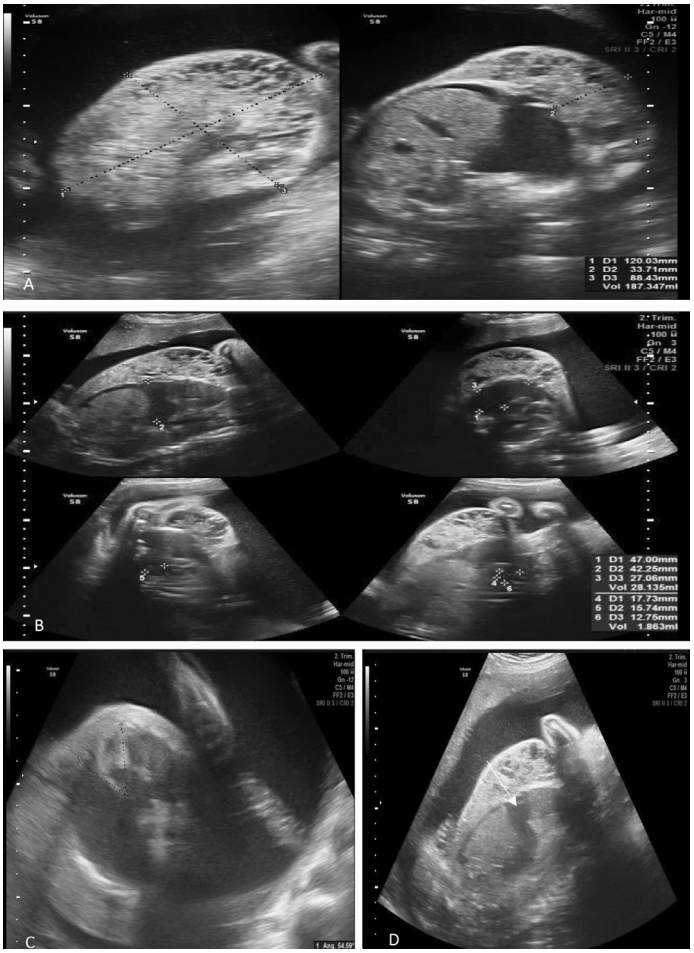

Figure 6. Ultrasound at 29 weeks age of gestation. A. Lymphangioma measures 12.0 x 3.3 x 8.8 cm, involving the right hemithorax from axillary to lumbar region. B. Pleural effusion, approximately 28.1 mL, at the right hemithorax (upper images) and pleural effusion, approximately 1.8 mL, at the left hemithorax (lower images). C. Cardiac axis deviation to the left at 54.5 degrees. D. Fetal ascites (arrow) noted above and below the liver.

involvement. Hemangiomas can show T1 hypointense signals and T2 hyperintense signals with a lobulated mass-like lesion.

Lymphangiomas are hyperintense on the T2-weighted sequences with corresponding T1 hypointense signals. Contrast – enhanced spin echo will show the vascular filling pattern of a hemangioma.[8] In our case, fetal MRI was requested to determine the exact extent and relationship of the mass to the adjacent structures for possible resection. However, the use of contrast was not advisable because of its questionable effect to the fetus. The fetal MRI showed the soft tissue mass external to the fetal lung without invasion of the adjacent organs.

Aside from hemangioma and lymphangioma, another differential diagnosis is Klippel-Trenaunay-Weber Syndrome. This is a congenital anomaly associated with capillary malformations, soft tissue or bony

hyperthrophy and venous malformations.[4,11] It has the same sonologic appearance to the vascular lesions of hemangiolymphangioma but the latter is not associated with asymmetric limb hypertrophy. Our patient had no other skeletal or vascular abnormalities.

In our patient, Kasabach-Merritt syndrome did not occur. This is a condition described to have platelet trapping within the hemangioma, leading to thrombocytopenia, macroangiopathic hemolytic anemia and consumption coagulopathy.[11]

Hemangiolymphangioma is a benign disorder but it has the tendency to invade underlying tissues which can cause severe deformity.[8] Medical and surgical options are available for treatment. Medical options include corticosteroids, interferon and vincristine. Extensive excision is also recommended.[1,10] In our patient, the option for sclerosing therapy, interferon and propanol were considered. However, due to the prematurity and weight, these were postponed until the baby has grown. Nutritional building and careful observation was prioritized before doing a full work up, which includes magnetic resonance angiography of the mass.

CONCLUSION

Thoraco-lumbar hemangiolymphangioma is a rare congenital condition of the blood vessels and lymphatics. With the use of ultrasound and other imaging modalities, vascular malformations are now being diagnosed prenatally with increasing frequency. Ultrasound is used for initial screening and magnetic resonance imaging is used to help localize the mass and delineate the extent of invasion. The combination of these imaging modalities will help observe the character and growth of these vascular malformations in utero, thus, helping in obstetrical planning. Postnatally, these imaging modalities can be used as a guide in the planning of medical management and surgical excision of a hemangiolymphangioma. ■

REFERENCES

1. Richter G, Friedman A. Hemangiomas and Vascular Malformations: Current Theory and Management. *Internal Journal of Pediatrics* Vol 2012, Article ID 645678, 10 pages.

2. Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. *Plast Reconstr Surg.* 1982; 69:412-420.

3. Marler J, Fishman S., et al. Prenatal Diagnosis of Vascular Anomalies. *Journal of Pediatric Surgery*, Vol 37, No 3 (March), 2002: pp 318-326.

4. Senoh D, Hanaoka U. Antenatal ultrasonologic features of fetal giant hemangiolymphangioma. *Ultrasound Obstet Gynecol.* 2001; 17:252-254.

5. DeCou MD, Jones DC, Jacobs HD, et al. Successful Ex Utero Intrapartum Treatment (EXIT) procedure for congenital high airway obstruction syndrome owing laryngeal atresia. *J Pediatr Surg*. 1998; 33(10):1563-5. [PubMed].

6. Munden A, Butschek R, et al. Prospective study of infantile hemangiomas: incidence, clinical characteristics and association with placental anomalies. *Br J Dermatol*. 2014 April; 170(4):907-913.

7. Lowe L, Marchant T, et al. Vascular Malformations: Classifications and Terminology the Radiologist Need to Know. Seminars in Roentgenology. *Semin Roentgenol.* 47 (2):106-117. 4 2012.

8. Masand P. Radiographic findings associated with Vascular Anomalies. *Semin Plast Surg*. 2014; 28:69-78.

9. Rha S.E. et al. Prenatal Sonographic and MR Imaging findings of Extensive fetal lymphangioma: A case report. *Korean J Radiol.* 2003; 4:260-263.

10. Tseng JJ, Chou MM, Ho ESC. Fetal axillary hemangiolymphangioma with secondary intralesional bleeding: serial ultrasound findings. *Ultrasound Obstet Gynecol.* 2002; 19:403-406.

11. Kaddah R, Lofti U. The role of radiology in the planning management of Klippel Trenaunay Syndrome (KTS). *Egyptian Journal of Radiology and Nuclear Medicine*. 2011; 42:201-210.