

THE ROLE OF MAGNETIC RESONANCE IMAGING IN THE PRENATAL MANAGEMENT OF A LYMPHATIC MALFORMATION

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We report the magnetic resonance imaging (MRI) findings in a case of extensive fetal lymphatic malformation involving the upper left arm and axillo-thoraco-abdominal wall found on routine prenatal ultrasound (US) examination at 22 weeks of gestation. MRI clearly reveals the tumor extent and tissue characteristics, and thick-slab T2-weighted MRI has the capacity to provide more information on the cystic lesion on global overview.

Key-words: Fetus, MR – Lymphatic system, abnormalities.

Lymphatic malformation, a hamartoma of the lymphatic vessels, has the potential to infiltrate surrounding structures. The prognosis for this lesion depends on its location, extent, and its association with karyotypic or other structural abnormalities (1-3). Accurate prenatal diagnosis using US complemented by MRI is essential for prenatal counseling of the parents and to guide perinatal and postnatal management. We report the MRI findings in a case of extensive fetal lymphatic malformation involving the upper left arm and axillo-thoraco-abdominal wall that was diagnosed by US and confirmed by MRI at 22 weeks of gestation.

Case report

A 33-year-old pregnant woman, gravida 2, para 1, at 22 weeks of gestation was referred to us for evaluation of a fetal chest mass. The patient and her husband were healthy and denied any family history of genetic disorders or congenital malformations. Prenatal US examination revealed a large, well-circumscribed cystic mass in the thoraco-abdominal-wall measuring 3.1 cm × 4.4 cm × 7.1 cm. The amniotic fluid volume, placenta, and other fetal US biometric data were within normal limits. There was no associated fetal congenital anomaly. MRI revealed a thin-walled, 3.6 cm × 4.8 cm × 7.8 cm homogeneous cystic mass without septation involving an area extending from the upper left arm to the complete left side of the trunk, including the left axilla and the left thoracic and abdominal walls (Fig. 1-3). The patient refused to undergo amniocentesis and decided to terminate her pregnancy because of worry about the poor prognosis predicted on the basis of large lesion size, the possibility of chromosomal abnormalities, unfavorable outcome of the pregnancy and complication of treatment, even though consultation with pediatrician, pediatric sur-

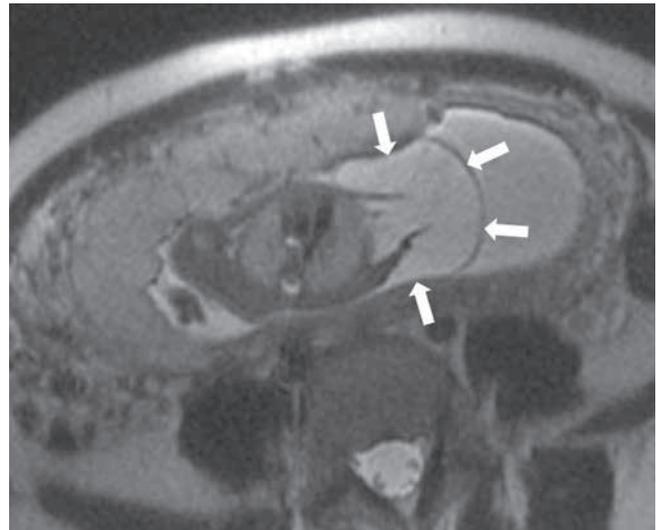


Fig. 1. – Axial MRI showing the thin-walled homogeneous cystic mass, without septation, involving the left axilla and left thoracic wall (arrows).

geon, psychologist as well as well explanation of non-life-threatening benign lesion, response of potential medical and/or surgical treatment, and possibly spontaneous regression during pregnancy. The diagnosis of extensive lymphatic malformation involving the upper left arm and axillo-thoraco-abdominal wall and a normal karyotype of 46, XX were confirmed by subsequent autopsy.

Discussion

Lymphatic malformation was first described in 1828 by Redenbacher and was considered a benign congenital malformation of the lymphatic system rather than a true neoplasm (1-3). Fetal nuchal lymphatic malformations are thought to be etiologically distinct from other lymphatic malformations and are believed to result from inadequate drainage of the lymphatic vessels into the venous system secondary to obstruction. Karyotypic abnormalities

with various malformation syndromes are present in 50-80% of patients. In contrast to nuchal lymphatic malformations, lymphatic malformations in other locations probably develop because of inefficient connections between the lymphatic and venous pathways and carry a low incidence of chromosomal and structural anomalies (4, 5). About 75% of lymphatic malformations occur in the nuchal area, 20% in the axillary region, and 5% in other locations of the body (larynx, mouth, tongue, retroperitoneum, mediastinum, mesentery, spleen, colon, and bones). The prognosis of lymphatic malformation depends on its location, the extent of the lesion, and its association with karyotypic or other structural abnormalities (1-3). Lymphatic malformations with a volume of $\geq 75 \text{ mm}^3$, as determined by US, are associated with the risk of an underlying abnormal fetal karyotype (66.7%; most frequently Turner's syndrome), persistence of the lymphangioma (72.7%), and an unfavorable outcome of the pregnancy (90%) (2). Amniocentesis or karyotype examination is therefore recommended when lymphatic malformation is found in the prenatal

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Fig. 2. — Coronal MRI showing the cystic mass involving the left thoracic and abdominal walls (arrows).



Fig. 3. — 3 Thick-slab T2-weighted MRI showing the extensive cystic mass involving the upper left arm and axillo-thoraco-abdominal wall on global overview.

period (1-3), although a low incidence of chromosomal and structural anomalies is reported for non-cervical lymphatic malformations.

US is the primary imaging modality for prenatal evaluation of lymphatic malformations in the second trimester of pregnancy, and has been reported extensively in the literature (1-4, 6, 7). However, only a few studies of the use of MRI to diagnose fetal lymphatic malformation have been reported (5, 8-10). With recent advances in MRI, this modality has become a useful, complementary tool for prenatal diagnosis of these malformations, particularly for assessment of extension of the lesion, predicting the prognosis, and evaluating other associated abnormalities. On MRI, lymphatic malformations appear as well-defined unilocular or multilocular homogeneous cystic masses, with or without septation. Fetal MRI can be helpful as a complementary tool to US owing to its better tissue contrast, larger field of view, enhanced anatomic evaluation, more detailed demonstration of the tumor extension, as well as allowing differential diagnosis of this type of malformation. In addition, compared to US, thick-slab T2-weighted MRI can provide more information with respect to normal static fluid or cystic abnormalities in the fetus on a global overview (11, 12).

The optimal treatment for lymphatic malformation is complete surgical excision. However, local recurrence is common after surgical

removal. Management of fetal or neonatal lymphatic malformation using sclerosing agents such as bleomycin or OK-432 (low virulence group-A *Streptococcus pyogenes* cultured with penicillin), or embolization with Ethibloc may cause shrinkage of the lesion. Needle aspiration of neonatal lymphatic malformation is usually ineffective and is associated with rapid re-accumulation of fluid and/or development of infection (5, 7, 8).

In conclusion, MRI is a helpful complementary diagnostic tool for the exact evaluation of the extent of fetal lymphatic malformation. This tool is essential for both prenatal counseling of the parents and to guide perinatal and postnatal management.

References

1. Suzuki N., Tsuchida Y., Takahashi A., et al.: Prenatally diagnosed cystic lymphangioma in infants. *J Pediatr Surg*, 1998, 33: 1599-1604.
2. Rosati P., Guariglia L.: Prognostic value of ultrasound findings of fetal cystic hygroma detected in early pregnancy by transvaginal sonography. *Ultrasound Obstet Gynecol*, 2000, 16: 245-250.
3. Ho M., Lee C.C., Lin T.Y.: Prenatal diagnosis of abdominal lymphangioma. *Ultrasound Obstet Gynecol*, 2002, 20: 205-206.
4. Goldstein I., Leibovitz Z., Noi-Nizri M.: Prenatal diagnosis of fetal chest lymphangioma. *J Ultrasound Med*, 2006, 25: 1437-1440.
5. Rasidaki M., Sifakis S., Vardaki E., Koumantakis E.: Prenatal diagnosis of a fetal chest wall cystic lymphangioma using ultrasonography and MRI: a case report with literature review. *Fetal Diagn Ther*, 2005, 20: 504-557.
6. Chiavérini C., Benoit B., Bongain A., Chevallier A., Lacour J.P.: Prenatal ultrasonographic detection of an axillo-thoracic lymphangioma: an ethical dilemma. *Prenat Diagn*, 2003, 23: 946-948.
7. Masood S.N., Masood M.F.: Case report of fetal axillo-thoraco-abdominal cystic hygroma. *Arch Gynecol Obstet*, 2010, 281: 111-115.
8. Rha S.E., Byun J.Y., Kim H.H., et al.: Prenatal sonographic and MR imaging findings of extensive fetal lymphangioma: a case report. *Korean J Radiol*, 2003, 4: 260-263.
9. Chen C.P.: Prenatal imaging of the fetal anterior chest wall cystic hygroma by magnetic resonance imaging. *Prenat Diagn*, 2003, 23: 1099-1100.
10. Hayashi A., Kikuchi A., Matsumoto Y., et al.: Massive cystic lymphangiomas of a fetus. *Congenit Anom (Kyoto)*, 2005, 45: 154-156.
11. Brugger P.C., Mittermayer C., Prayer D.: A new look at the fetus: thick-slab T2-weighted sequences in fetal MRI. *Eur J Radiol*, 2006, 57: 182-186.
12. Chaumoitre K., Wikberg E., Shojai R., et al.: Fetal magnetic resonance hydrography: evaluation of a single-shot thick-slab RARE (rapid acquisition with relaxation enhancement) sequence in fetal thoracoabdominal pathology. *Ultrasound Obstet Gynecol*, 2006, 27: 537-544.