

Prenatal Sonographic and MR Imaging Findings of Extensive Fetal Lymphangioma: A Case Report

Sung Eun Rha, MD¹
Jae Young Byun, MD¹
Hak Hee Kim, MD^{1,2}
Jong-Chul Shin, MD³
Hyun Young Ahn, MD³
Dong-chul Kim, MD⁴
Kyo-Young Lee, MD⁴

We report the imaging findings in a case of fetal lymphangioma involving the retroperitoneum and right lower extremity, and diagnosed by ultrasonography and magnetic resonance (MR) imaging at 26 weeks of gestation. Prenatal ultrasonograms and T2-weighted single-shot fast spin-echo MR images clearly revealed an extensive, multilocular cystic mass with internal hemorrhage in the retroperitoneum extending to the lower extremity.

Index terms :

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Department of ¹Radiology, The Catholic University of Korea, College of Medicine; ²Department of Radiology, University of Ulsan College of Medicine; Departments of ³Obstetrics and Gynecology and ⁴Pathology, The Catholic University of Korea, College of Medicine

Address reprint requests to:

Jae Young Byun, MD, Department of Radiology, Kangnam St. Mary's Hospital College of Medicine, The Catholic University of Korea, 505 Banpo-dong, Seocho-gu, Seoul 137-040, Korea.
Telephone: (822) 590-2785
Fax: (822) 599-6771
e-mail: jybyun@catholic.ac.kr

Lymphangiomas are hamartomas of the lymphatic vessels that have the potential to infiltrate surrounding structures. About 50% are present at birth and up to 90% become evident by the age of two years (1). The prognosis of a lymphangioma depends on the location and extent of the lesion and the presence of other associated abnormalities. Because fetal lymphangiomas are frequently associated with karyotypic or other abnormalities, including skin edema, hydrops fetalis, and polyhydramnios, their prenatal diagnosis is important. The neonatal outcome of a large fetal lymphangioma is generally poor (2). Although spontaneous regression can occur in a fetal lymphangioma with normal chromosomes, large fetal lymphangiomas require a perinatal team approach, with possible management options including prenatal aspiration of the cyst, elective cesarean section, or a delivery mode which will avoid fetal damage (3). Prenatal diagnosis thus permits a planned delivery, and adequate intrapartum and postnatal resuscitation may improve the prognosis. Several case reports regarding the prenatal sonographic findings of fetal abdominal lymphangioma have been published (2), but to the best of our knowledge, only two reports have described the magnetic resonance (MR) imaging finding (4, 5). We report the imaging findings in a case of extensive fetal lymphangioma involving the right side of the retroperitoneum and extending to the right thigh and lower leg, which was correctly diagnosed by sonography and confirmed by MR imaging at 26 weeks of gestation.

CASE REPORT

A 34-year-old woman whose one previous pregnancy had involved cesarian section and the delivery of a healthy male infant at term was referred to our hospital at 26 weeks of gestation on account of earlier sonographic findings which had indicated possible multicystic dysplastic kidney and sacrococcygeal teratoma. Detailed sonography revealed the presence of a large, multiseptated cystic mass at the right side of the abdomen, extending to the right buttock and lower extremity, the transverse diameter of which was much larger than that of the left (Fig. 1A). The mass demonstrated variable echo patterns; color Doppler sonography revealed no internal flow at the time of ex-

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amination. For further evaluation of these complex abnormalities, MR imaging was performed using a 1.5-T superconducting unit (Signa Infinity with twin-speed dual gradient; General Electric Medical Systems, Milwaukee, Wis., U.S.A.) together with a phased array coil. After performing a localized gradient-echo sequence, T2-weighted single-

shot fast spin-echo MR images (TR/TE, 1624/79; bandwidth, 31.2 kHz; field of view, 22 × 22 cm; matrix, 224 × 192; slice thickness, 5 mm; intersection gap, 0 mm; number of excitations, 0.56) were obtained in the axial, coronal, and sagittal planes; the acquisition time for each was less than 1 second. The images revealed a large, multilocular,

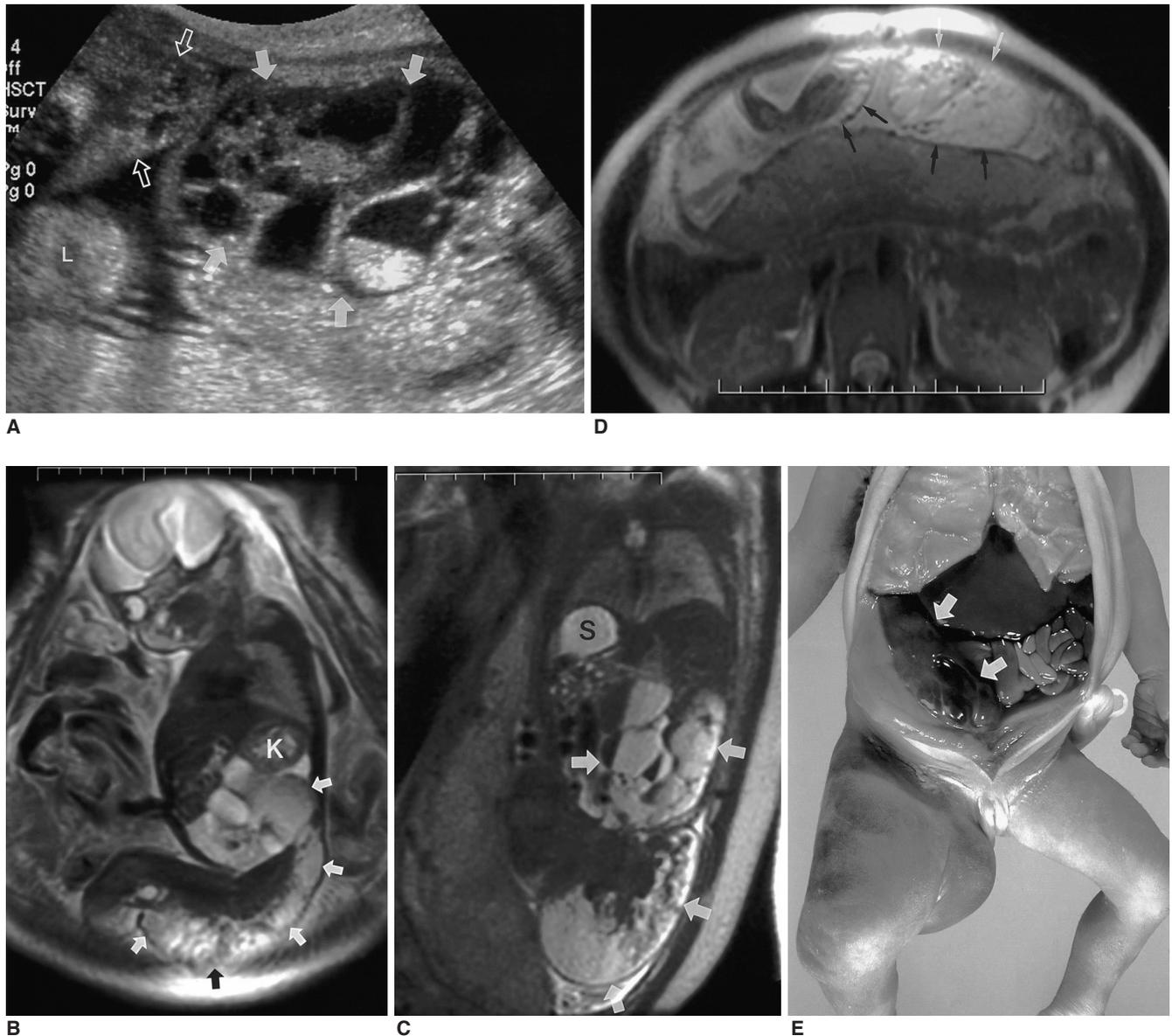


Fig. 1. A 26-week-old female fetus with an extensive lymphangioma.

A. Prenatal ultrasonography shows a large multiseptated cystic mass (arrows) with variable echo patterns, located at the right of the fetal abdomen and also involving the right lower extremity (open arrows). The transverse diameter of this extremity is much greater than that of the left one (L). Color Doppler ultrasonography revealed no flow within the mass at the time of examination (not shown).

B. Sagittal T2-weighted single-shot fast spin-echo MR image (TR/TE, 1624/79) depicts a large intra-abdominal high-signal-intensity mass (arrows) with multiple internal locules, displacing the ipsilateral kidney (K) superiorly. The mass (arrows) extends continuously to the right buttock and right lower extremity.

C. Coronal T2-weighted single-shot fast spin-echo MR image (TR/TE, 1624/79) clearly reveals the multilocular cystic nature of the mass (arrows), each locule of which contains multiple fluid-fluid levels. S=Stomach.

D. T2-weighted single-shot fast spin-echo MR image (TR/TE, 1624/79) obtained parallel to the right lower fetal extremity shows the distal extent of the lymphangioma (arrows). This presents as an extensive subcutaneous cystic mass, with asymmetric limb hypertrophy.

E. Post-mortem photograph depicts a large lymphangioma (arrows) at the right of the abdomen, extending to the right lower extremity.

high-signal mass in the right-side of the abdomen; displacement of the ipsilateral kidney superiorly and bowel loops to the left was noted (Figs. 1B, C). Within the mass, which extended continuously to the right buttock and right lower extremity, presenting as an extensive subcutaneous cystic mass and causing asymmetric limb hypertrophy, multiple fluid-fluid levels were noted (Figs. 1C, D). Cystic lymphangioma was indicated, and after counselling, the parents opted for termination of the pregnancy because of the poor prognosis predicted on the basis of the lesion's large size and infiltrative nature.

Postmortem examination revealed a female fetus weighing 1336 g and with a fetal karyotype of 46, XX. A large, multilocular cystic mass, containing thin-walled cysts of both clear and hemorrhagic fluid, occupied the right side of the retroperitoneum (Fig. 1E), infiltrating the adipose and muscular tissues of the buttock through the retroperitoneal space and extending to the right lower extremity. Histological examination demonstrated the presence of large, endothelial-lined vascular spaces containing lymphocytic aggregates and hemorrhagic fluid, suggesting a diagnosis of lymphangioma. No other significant abnormalities of the internal organs were identified.

DISCUSSION

Lymphangiomas are benign hamartomas of the lymphatic system, consisting of multiple dilated vessels. They arise due to a developmental defect in the lymphatic pathways, which usually develop from the sixth week of gestation, leading to the proximal dilatation of afferent channels, and can be classified histologically as one of three main types: (1) simple lymphangiomas, consisting of lymphatic capillaries; (2) cavernous lymphangiomas, made up of larger lymphatic vessels with a fibrous adventitia; (3) cystic lymphangiomas or cystic hygromas, comprising multiple cysts ranging in size from a few millimeters to several centimeters. All types may coexist within the same lesion. The cyst usually contains serous or chylous fluid, and if complicated, may have bloody or purulent contents (4). Nearly 75% of lesions are located in the head and neck, or axilla, while the other 25% involve the trunk (11%), extremities (11%), mediastinum (1%), or abdomen and genitalia (3%) (6). Abdominal lymphangiomas are reported to occur most commonly in the mesentery of the small bowel, with the retroperitoneum being the second most common site.

For a fetal lymphangioma diagnosed during the prenatal period, the overall prognosis is poor, with a mortality rate ranging from 50 to 100% (7). In 50–80% of patients affected by nuchal cystic hygromas, karyotypic abnormalities and various malformation syndromes are present. Romero

et al. cited a 69% incidence of abnormal karyotypes in affected fetuses, the majority being 45, X (Turner syndrome) (8), but these features might not apply to non-nuchal lymphangiomas diagnosed *in utero*. Because of the paucity of available data, it is uncertain whether abdominal lymphangiomas carry a significant risk for aneuploidy (7). In our case, there was no chromosomal anomaly.

At sonography and MR imaging, cystic lymphangiomas appear as sharply defined, unilocular or multilocular cystic masses, with thin- or thick-walled septa. At sonography, the fluid may be anechoic, with enhanced through-transmission, or there may be variable internal echoes or fluid-fluid levels, due to bleeding and fibrin deposition (9). In our case, fetal sonography and MR imaging accurately identified the lymphangioma and defined its anatomical location and extent, thus providing the information essential for parental counselling. Because of its accuracy and safety, as well as its low cost and real-time capability, and the fact that it is easy to use, sonography has been regarded as the imaging modality of choice for the prenatal assessment of fetal abnormalities. Its inherent disadvantages include, however, operator-dependency and a limited field of view (10). With recent technical advances in ultrafast MR imaging, the modality has become a useful tool for prenatal diagnosis, particularly for the assessment of complex fetal abnormalities. In this case, single-shot fast spin-echo MR imaging clearly demonstrated fetal abnormality and helped make the correct prenatal diagnosis. In addition, MRI allowed more comprehensive imaging of the fetus because of its larger field of view, and accurately delineated the extent of the mass.

To our knowledge, published reports have described three cases in which a fetal abdominal lymphangioma extending to an extremity was diagnosed by prenatal sonography, as in our case (2). This extension reflects the pathway of the normal developing lymphatic system and may be typical of retroperitoneal lymphangiomas. In all three cases, pregnancy was terminated, in view of the poor prognosis. In contrast, postnatal surgical resection or sclerotherapy was successfully performed in four reported cases of fetal abdominal lymphangiomas confined to the abdominal cavity or abdominal wall (2).

For the postnatal treatment of abdominal lymphangioma, the preferred treatment is surgical extirpation, with careful preservation of involved structures. Large but localized lymphangiomas can be excised completely, but the surgical treatment of diffuse and multiple lesions is extremely difficult and is associated with high morbidity and mortality (6). For the treatment of surgically unresectable lesions, the injection of a sclerosing agent is considered appropriate: successful outcomes have been reported following the use

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of intralesional bleomycin, sclerotherapy with OK-432, or percutaneous embolization with Ethibloc (2, 11). The successful intrauterine treatment of a cystic hygroma with OK-432 has also been reported (12).

In conclusion, prenatal MR imaging was valuable in the detection, characterization, and exact evaluation of the extent of an extensive fetal lymphangioma, and complemented the role of sonography.

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