Prenatal Diagnosis of a Large Axillo-thoracic Lymphangioma by Three-dimensional Ultrasonography

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Lymphangiomas consisting of single or multilocular fluid-filled cavities are referred to as cystic hygromas. The cyst usually contains serous or chylous fluid, and if complicated, may have bloody or purulent contents. Postnatal outcome depends mainly on the size and location of the lesion and the absence of blood flow on color Doppler mapping is characteristic of lymphangiomas as opposed to hemangiomas. In the present case, the axillo-thoracic lymphangioma was easily diagnosed on the basis of conventional 2D US and 3D US.

Key Words: Axillo-thoracic lymphangioma, 3D US

Introduction

Lymphangiomas can occur in almost any location in the body, but are most commonly seen in the soft tissue of the neck, axillae, thorax, and lower extremities. The sizes of these tumors vary from a few millimeters to several centimeters. So far, prenatal evaluation for the prognosis of fetal lymphangiomas has been based on 2D US.¹ ² ³ 3D US helped to identify the lesion clearly and completely and make the consultation with pediatric surgeons and parents more comprehensive. We report a case of prenatal assessment of axillo-thoracic lymphangioma by 2D US and 3D US.

Case report

A 26-year-old woman, gravida 2, para 1, was referred at 21 weeks’ gestation to our hospital after prenatal diagnosis of a fetal abdominal mass. At the time of referral, 3D US (ACCUVIX, Medison, Seoul, Korea) confirmed the presence of a large multilocular, septate cystic mass located on the right side of the fetal anterior chest wall, measuring 5.93×4.16 cm. On transverse sections, the lesion extended from the posterior to the anterior axillary line. The fetal shoulder seemed to be involved and it was possible to assess axillary involvement accurately (Fig. 1). Color Doppler imaging did not reveal any blood flow inside the cystic mass. At 28 weeks’ gestation, the patient consented to undergo 3D US examination. It was possible to delineate the shape of fetal skin covering the lesion; the mass did not pass across the fetal ribs. Serial ultrasound studies demonstrated that the right axillary mass increased in size 5.93×4.16 cm at 21
weeks’ gestation to 16.16×11.52 cm at 28 weeks’ gestation. Pleural effusion, ascite and skin edema appeared at 28 weeks (Fig. 2). Emergence cesarean delivery was performed at 28 weeks because of maternal right upper quadrant pain and fetal condition. A male neonate was delivered, weighing 2500 g with Apgar scores of 2 at 1 minute and 2 at 5 minutes. Neonatal physical examination confirmed the presence of a large, soft subcutaneous lesion, which extended from the right lower part of the neck, axilla to the abdomen in the subcutaneous area (Fig. 3), and the hand was spared. The skin had a port-wine discoloration and these soft masses contained macroscopic cysts filled with serous fluid. No communications between the cysts and the thoracic or abdominal cavity existed (Fig. 3). Histological examination demonstrated cystic lymphangioma with dilated communicating lymphatic vessels lined by normal epithelium and cystic areas with rare aggregates of lymphocytes (Fig. 4). The baby was expired due to spontaneous pneumothorax after birth.

We could not perform karyotyping due to disagreement of parents.

Discussion

The lymphatic system of the embryo is supposed to develop around the fifth to sixth week of gestation. Six lymphatic sacs (two jugular sacs draining the head, neck, and arms; two iliac sacs draining the legs and lower trunk; and two sacs draining the gut called the cisterna chill, and the retroperitoneal sac) develop in close proximity to the central venous system. Failure of the lymphatic system to connect and drain into the jugular veins leads to lymphatic fluid stasis, dilated lymphatic vessels and varying fluid-filled mass called the lymphatic obstruction sequence. If the connection between the jugular lymphatic and venous system and an alternative route is established, lymphangiomas might resolve. Lymphangiomas can be classified
into three groups; lymphangioma simplex, consisting of capillary sized channels; cavernous lymphangioma, consisting of dilated lymphatic channels frequently with a fibrous adventitial covering; and cystic lymphangiomas or hygromas, composed of multiple cysts of varying size lined by endothelial cells.\(^5\)

The incidence of cystic hygroma is estimated to be 1:6000 pregnancies but it is a relatively common anomaly in miscarried fetuses, with a frequency of 1:875.\(^6\) Lymphangiomas can occur in a variety of anatomic lesions. About 75% of cystic lymphangiomas occur in the neck, 20% in axillae and 5% in various other parts of the body such as the retroperitoneum, mediastinum, mesentery, spleen, colon, bones, groin, trunk, extremities, larynx, mouth, tongue, and scrotum.\(^7\) There are many papers on cystic hygroma colli but only a few on fetal axillo-thoracic cystic hygroma.

Prenatal diagnosis of a large lymphangioma can be achieved easily by conventional 2D US on the basis of the finding of a multilocular or septate cystic mass, 2D US is
Fig. 4. It shows large, irregularly shaped lymphatic channels in the dermis (A-×40). And they are lined by a single layer of bland endothelial cells (Hematoxylin and eosin, B-×400). Microscopic examination showed variable-sized and dilated lymphatic channels lined by endothelial cells and adjacent lymph tissues.

also effective in identifying predominantly cystic lesions that may benefit from prenatal needle aspiration to reduce their volume before delivery. However, it may be difficult to accurately evaluate the limits of the tumor and its connections with surrounding organs. 3D US had the advantage over conventional imaging of allowing for multiplanar imaging studies. By working on the multiplanar images, we were able to determine the inner limits of the mass and its connections with the bony structures of the thoracic wall. We were able to generate rendered images confirming that the axilla and the arm were involved, the upper limit of the mass ending a few millimeters below the apex of the armpit. In our case, 3D US helped to identify the lesion clearly and completely. Prenatal MRI can confirm ultrasonographic findings, provide detailed fetal anatomical evaluation, and demonstrate the extent and character of lymphangiomas.

The differential diagnosis of prenatally detected cystic heterogeneous masses over the axilla or anterior chest wall include cystic hygroma, simple cysts, branchial cleft cysts, hemangiomas, hemangiolympangioma, lymphoceles, teratomas, hemangioendotheliomas, ectopia cordis, amniotic band syndrome, limb-body wall complex, body stalk anomaly, Parkes-Weber syndrome, Maffucci syndrome, and Klippel-Trenaunay syndrome. Accurate prenatal diagnosis and anatomical evaluation are important as they permit planned delivery and prompt postnatal resuscitation, and allow the option of terminating the pregnancy if a poor outcome is predicted. In our case, we expected the poor prognosis due to 3D US findings of aggressive increasing mass size, pleural effusion, and ascite and skin edema.

For the postnatal treatment of 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 are extremely difficult and are associated with high morbidity and mortality. For the treatment of surgically unresectable lesions, the injection of a sclerosing agent is considered appropriate: successful outcomes have been reported following the use of intraliesional bleomycin, sclerotherapy with OK-432, or percutaneous embolization with Ethibloc. 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.

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.\textsuperscript{14,15}

In conclusion, when an unusually located cystic mass is found in the antenatal screening, the fetus should be screened for other anomalies. Serial sonograms to assess the growth of the mass and monitoring for the development of hydrops should aid in the management of the pregnancy and are useful in the counseling of future pregnancies. Also 3D US might have the advantage of being more widely available, thus enabling, for instant, serial volume measurements that could be of interest in monitoring tumor growth.

References