

# Fetal Mediastinal Lymphangiomas

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**Objective.** The purpose of this series was to evaluate the prenatal sonographic findings and postnatal outcomes in 2 fetuses with mediastinal lymphangiomas. **Methods.** The fetal images were compared with postnatal imaging and surgical findings. **Results.** The 2 fetuses had anechoic mediastinal masses at 25 and 22 weeks, which proved to be lymphangiomas. One, located in the anterior mediastinum, also enveloped the superior vena cava, brachial plexus, phrenic nerve, larynx, and lower parts of the neck vessels and extended into the subcutaneous tissues of the anterior chest wall through an intercostal space. In the second patient, the lymphangioma appeared to be a unilocular cyst, which involved the deep tissues of the neck as well as the posterior and lateral mediastinum. Both required 2 interventions after birth. **Conclusions.** Fetal mediastinal lymphangiomas appeared anechoic and sent extensions into the neck in the first case, around the superior vena cava, through the intercostal spaces to the skin, and around the brachial plexus in the second case, and deviated the trachea in both cases. In 1 case, there was also ectasia of the superior vena cava. This ability to entwine around vital structures can make it difficult to determine the extent of involvement on antenatal sonography and to remove lymphangiomas completely, and recurrence is common. **Key words:** fetal cystic hygroma; lymphangioma; mediastinum; neck; skin; superior vena cava.

## Abbreviations

SVC, superior vena cava

Received August 13, 2007, from the Division of Fetal Imaging, Department of Obstetrics and Gynecology, William Beaumont Hospital, Royal Oak, Michigan USA (C.H.C., W.L., R.A.B.); Department of Obstetrics and Gynecology, Wayne State University, Detroit, Michigan USA (C.H.C., W.L.); Department of Obstetrics and Gynecology, University of Michigan, Ann Arbor, Michigan USA (C.H.C.); Hurley Hospital, Flint, Michigan USA (I.V.); and Saginaw Maternal-Fetal Medicine Specialists, Saginaw, Michigan USA (D.W.). Manuscript accepted for publication September 6, 2007.

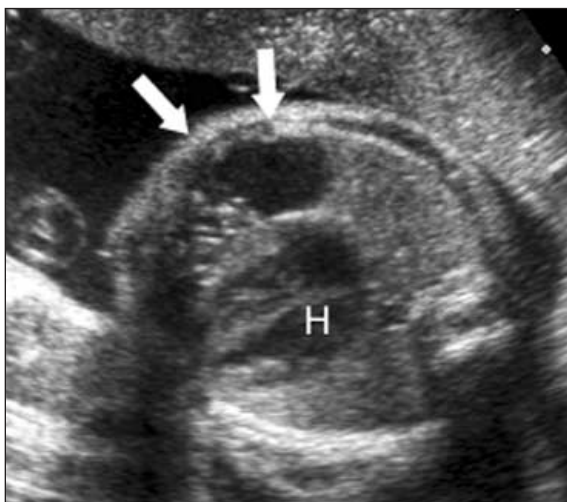
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**F**etal lymphangiomas of the chest are rare occurrences. The purpose of this series was to evaluate the prenatal sonographic findings and postnatal outcomes in 2 fetuses with mediastinal lymphangiomas.

## Case Descriptions

### Case 1

A 22-year-old patient, gravida 3, para 2, was seen for sonographic examination at 25 weeks' gestation. A cystic mass measuring 3.3 × 0.7 cm with some small septations was located between the heart apex and left chest wall (Figure 1) and extended up around the base of the superior vena cava (SVC). The SVC appeared dilated and irregular, but no obstruction could be identified (Figure 2). No flow was seen within the cystic mass, and although there were multiple septations, there were no solid components. The differential diagnosis included a



**Figure 1.** Case 1. Axial view showing a cystic mass between the heart and left chest wall (arrows). Note the septations near the cardiac apex. H indicates heart.

lymphangioma, a pleural cyst, a pulmonary sequestration, and a cystic adenomatoid malformation. The small septations were most consistent with a lymphatic origin. Subsequent 3-dimensional evaluation also showed extension between the ribs into the subcutaneous tissues of the chest. This was confirmed with subsequent 2-dimensional scanning (Figure 3). The most likely diagnosis was then thought to be a lymphangioma. Fetal magnetic resonance imaging added no additional information.

The child was born at 37 weeks by repeat cesarean delivery and had no evidence of SVC syndrome or respiratory difficulty. Magnetic resonance imaging showed a cystic mass adherent

**Figure 2.** Case 1. Sagittal view showing an enlarged and irregular SVC (arrows). Part of the main cystic mass can be seen near the ribs.



anteriorly to the thymus and to the parietal pleura, the pericardium at the base of the heart, and the aorta. It encased the base of the SVC, the bases of the right subclavian artery and vein, and the bases of the right internal jugular vein and carotid artery and extended behind the larynx. It also extended along the right subclavian artery and vein into the right axilla and into the subcutaneous areas above the sternum by 2 slitlike connections. Crepitant tissue could be palpated in those areas.

The child underwent surgery at 6 weeks of age, at which time there were additional findings: some fingerlike projections going into the brachial plexus were also seen, and the mass was noted to surround the phrenic nerve. At 4 months of age, a second operation was necessary because the lymphangioma had reappeared in the mediastinum, and a new area was seen within the right axilla. The axillary portion was removed.

#### Case 2

A 21-year-old patient, gravida 2, para 1, was referred at 22 weeks' gestation. Prenatal sonography showed a unilocular cystic mass in the upper right thorax (Figure 4) with extension into the right neck between the carotid artery and jugular vein (Figure 5). There were no septations and no flow seen within it, and there was no involvement of the subcutaneous tissues or skin. By 30 weeks, it was proportionately larger. Three-dimensional imaging added no new information. Postnatal computed tomography (Figure 6) confirmed the antenatal findings and was consistent with a lym-

**Figure 3.** Case 1. Sagittal view showing cystic areas (arrow) in the skin, which were extensions of the mediastinal lymphangioma.



phangioma. Although the mass deviated the trachea slightly, it did not narrow it, and there were no respiratory problems at birth. Sclerotherapy with doxycycline was performed twice. The fluid reaccumulated after the first attempt. The child remained asymptomatic to age 1 year.

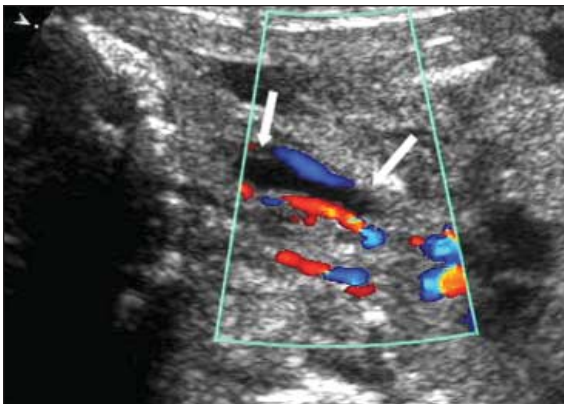
## Discussion

The term “cystic hygroma” usually refers to a lymphatic malformation in the neck. Elsewhere, the same lesion is termed a “lymphatic malformation.” These lesions are most frequently seen on the back of the neck but can be located on the sides of the neck or in the axillae and arms. They have multiple septations and usually are immediately under the skin surface.

**Figure 4.** Case 2. Sagittal view showing the cystic mass extending from the chest into the neck (on the left). L indicates lung.

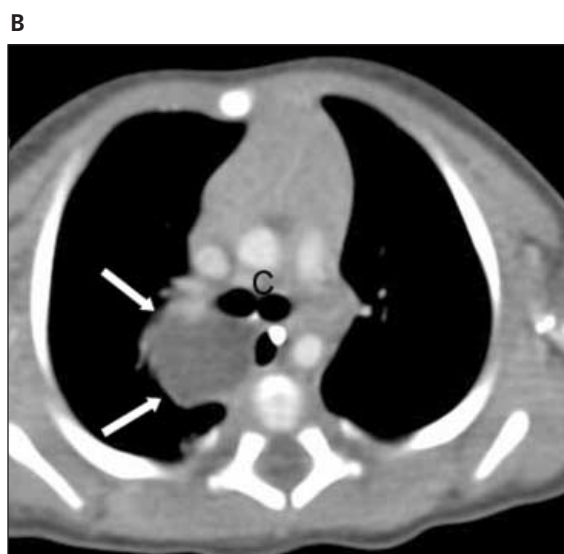


**Figure 5.** Case 2. Coronal view. The neck extension of the lymphangioma (arrows) lies between the jugular and carotid vessels.



Lymphangiomas of the chest in the newborn have been rarely reported.<sup>1-3</sup> Although they were isolated to the chest (anterior mediastinum) and did not extend into the neck, some caused respiratory distress at birth. Lymphangiomas of the fetal thorax have been described in the posterior mediastinum in 2 cases,<sup>4,5</sup> in 1 of which the hygroma continued through the diaphragm into the retroperitoneum; in another case, the mass was in the anterior mediastinum.<sup>6</sup> Pleural effusions developed in the latter.

**Figure 6.** Case 2. Neonatal computed tomography. **A**, Axial view at the level of the mandible showing the mass (arrows) between neck vessels (V). **B**, Axial view at the level of the carina (C), with arrows indicating the mass.



The differential diagnosis includes a bronchogenic cyst, which is usually seen in the upper part of the lung, an esophageal duplication cyst in the posterior mediastinum, a pericardial cyst in the cardiophrenic angle, and a cystic adenomatoid malformation (which usually has no septations and instead has solid components). Extensions such as those seen in 2 cases described here will narrow the diagnosis to lymphangiomas because none of the other masses extend through the intercostal spaces, around the SVC, or into the neck.

Management is difficult because complete resection is usually not achieved, and recurrence is common.<sup>7</sup> Sclerotherapy with doxycycline, bleomycin, or alcohol has been attempted in neck and retroperitoneal lesions but has not been described before in mediastinal lesions.

Both of these cases were unusual in several ways. The neck fluid collections in case 2 were not subcutaneous but rather extended deep into the soft tissues between the neck vessels. The neck and chest cysts were contiguous.

The SVC enlargement in case 1 was thought not to be due to obstruction because flow could be seen through it, and the portion internal to the mass was not smaller than usual; rather, the portion above the mass was larger than expected. Ectasia of the SVC has been reported in adults with cystic hygromas of the chest cavity, in which the SVC was embedded in the mass but obviously not obstructed.<sup>8</sup> It has been postulated that perhaps ectasia of the SVC is part of the underlying malformation. Herniation from the mediastinum through the intercostal spaces, similar to that in case 1, has been reported in a neonate.<sup>2</sup>

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