Lymphangiomas in Children: Correlation of Sonographic and Pathologic Findings

Sun Wha Lee, M.D., Kyung Nam Ryu, M.D., Yup Yoon, M.D., Pil Mun Yu, M.D.*

Department of Radiology, Kyung Hee University Hospital

— Abstract —

The sonographic features of 23 lymphangiomas (19 pediatric patients) were compared with the pathologic findings. Nineteen lymphangiomas appeared as unicameral (n = 2) and multiloculated (n = 17) cystic masses. Remaining lesions were inhomogeneously echogenic mass with small cystic portions (n = 3) and a mixed pattern (n = 1). Fourteen of the multiloculated tumors had thin septa and 6 had solid echogenic foci. The fluid within the majority of the cyst was anechoic in 8 cases and echogenic in 11 cases. Correlation of the sonographic features with the pathologic findings demonstrated that the cystic spaces corresponded to the dilated lymphatic spaces lined with endothelium, separated by septa. Echogenic fluid represented hemorrhage. The echogenic components corresponded to clusters of very smaller dilated lymphatic channels, thick fibro-fatty septa, or blood clot.

The author's experience suggests that the most characteristic sonographic appearance of lymphangioma is a multiloculated cystic mass with thin septa, reflecting the preponderance of fluid-filled spaces. An atypical appearance usually reflects the presence of blood or dominancy of cavernous type. The information obtained with US imaging can help in providing a preoperative diagnosis and in planning surgical resection.

Index Words: lymphatic system, US study 99.1298
children, lymphatic system

INTRODUCTION

Lymphangiomas are rare benign tumors arising from developmental malformation of the lymphatic systems. These lesions may be classified as capillary, cavernous, or cystic, depending on the predominant size of the lymphatic spaces within the tumor, but differentiation of these types is difficult pathologically due to frequent coexistence of three types within a single lesion. Eighty to 90 percent of the cases are detected by the age of 2 years. The greatest majority of lymphangiomas occur in the neck and axilla (1-4).

Recently, ultrasonographic (US) imaging has become established as an important noninvasive method in the evaluation of deep and superficial soft tissue masses. The radiologic findings of lymphangiomas have been well documented in the radiological literature but there is a little information about sonographic-pathologic correlation (4-9). We reviewed the sonographic findings in 23 cases of lymphangiomas in children and com-
pared the US features with the pathologic findings to determine the histologic basis for the sonographic characteristics of these lesions.

MATERIALS AND METHODS

Between March 1988 and June 1992, we performed US imaging on 19 children with 23 pathologically proved lymphangiomas at Kyung Hee University Hospital. There were 17 boys and two girls with ages ranging from 4 days to 11 years (mean age, 4 years). Twenty-two lesions were resected surgically and one lesion was aspirated. The interval between sonography and surgery was less than 5 days in most cases but up to 2 weeks in some cases. Eight patients also underwent computed tomography as part of their diagnostic workup after sonographic examination.

Sonographic examinations were performed by 5.0 MHz (ATL Ultramark-9, USA) or 7.5 MHz (Toshiba SSA-90A, Japan) linear array transducer. Sonograms, medical records and pathologic findings were reviewed retrospectively and we compared the sonographic results with the surgical and pathologic descriptions of the tumors in an attempt to understand the basis of the structural appearances encountered on US imaging. The US images were reviewed with attention to the specific features including pattern of echogenicity of the mass lesion, contents of cystic fluid, solid echogenic focus arising at cystic wall or septa, thickness of internal septations (thin septa:less than 2 mm, thick septa:about 2-4 mm in thickness), margin (well-demarcated or ill-demarcated), extent of tumor, and its relationship to the adjacent structures.

RESULTS

Lymphangiomas occurred in the neck in 10 patients, in the axilla in two patients, in the chest wall, thigh, scrotum, scrotum, and omentum in

Fig. 1. Hemorrhaged lymphangioma arising in Colles fascia of the left scrotum in a 13-month-old boy. a. Transverse sonogram demonstrates a multilocular, septated, and cystic mass crossing the midline (arrows). There are uniform, coarse echoes within multiloculations. Left testis (not shown) displaced up to the left external inguinal ring. T = right testis. b. Histologic section shows a typical lymphangioma with very large lymphatic spaces (L) lined with endothelium, separated by septa containing lymphocytes, fat, blood vessels, nerve, and fibrous tissue. (Hematoxylin and eosin, X40.)
one patients, respectively. Two patients had two separate lesions; face and neck, and thigh and inguinal region, respectively. The remaining one patient had three separate lesions, involving the axilla, anterior chest wall, and back. Lymphangiomas ranged in size from $2 \times 1 \times 0.5\text{cm}$ (a lesion in the face) to $20 \times 18 \times 9\text{cm}$ (a lesion arising in the omentum) and 14 cases were 3 to 6cm in its maximum diameter. Table 1 summarizes the sonographic appearance of lymphangiomas. Nineteen cases appeared as unicameral ($n = 2$) and multiloculated ($n = 7$) fluid-filled cysts and had typical histologic features characterized by cystic spaces of varying size lined with endothelium, separated by septa which was composed of connective tissue, adipocytes, smooth or skeletal muscle, and lymphoid aggregates (Fig. 1b). In eight cases (42%), the features of the fluid within the cysts were those of simple fluid, that is, anechoic with enhanced through-transmission and with a sharply defined far wall (Fig. 2). In remaining

11 cases (58%), combination of the features of simple fluid and diffuse, coarse echoes or dependent echoes were present; these were shown pathologically to be hemorrhage within the ma-
Fig. 3. 3c. shows multi-septated loculations with fluid-fluid levels and small low density of fatty portions between loculi (arrow heads).

Fig. 4. Lymphangioma arising in greater omentum in a 2-year-old boy.

a. Sonogram in the longitudinal plane of the left abdomen reveals multiloculated cystic mass with thick septations. UB = urinary bladder.

b. Sonogram in the transverse plane of the lower abdomen demonstrates echogenic debris and echogenic components in some loculi.
jor portions of lymphangioma (Fig. 1,3). Of the multiloculated cystic masses, septa was thin in 14 cases (Fig. 2) and thick in three cases (Fig. 4). On histologic examination, thicker septa contained inflammatory reaction and/or accumulation of fatty tissue. Six of 19 lesions had echogenic focus arising from cystic walls or septa, which corresponded histologically to portion containing fatty tissue or coagulated blood (Fig. 3,4).

Three lymphangiomas were inhomogeneously echogenic mass containing small cystic spaces. These echogenic components were proven to represent anomalously proliferated very small dilated lymphatic channels with thick fibro-fatty septa on histologic examination (Fig. 5). Remaining one case in the supraclavicular fossa showed a mixed pattern, predominantly cystic on the medial side and inhomogeneously echogenic on the lateral side of the mass lesion (Fig. 6).

Fifteen tumors were round or oval and had well-defined margins that could be clearly distinguished from normal tissue. The remaining eight lesions had ill-defined margin with infiltration into adjacent structures. US demonstrated the extent of tumor and its relationship to adjacent structures. In one infant, ex-

### Table 1. Sonographic Appearance of Lymphangiomas

<table>
<thead>
<tr>
<th>Findings</th>
<th>No. of Cases</th>
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<tr>
<td>Features of mass</td>
<td></td>
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<tr>
<td>mainly cystic mass (n = 19)</td>
<td></td>
</tr>
<tr>
<td>anechoic</td>
<td>8</td>
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<tr>
<td>anechoic &amp; echogenic</td>
<td>8</td>
</tr>
<tr>
<td>fluid-debris level</td>
<td>3</td>
</tr>
<tr>
<td>thin septa</td>
<td>14</td>
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<tr>
<td>thick septa</td>
<td>3</td>
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<tr>
<td>solid echogenic focus</td>
<td>6</td>
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<tr>
<td>mainly solid mass (n = 3)</td>
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<tr>
<td>mixed pattern (n = 1)</td>
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<tr>
<td>Margins</td>
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<tr>
<td>well-defined</td>
<td>15</td>
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<tr>
<td>ill-defined</td>
<td>8</td>
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**Fig. 5.** a. Transverse scan of the right axillary region in a 35-day-old girl shows ill demarcated, inhomogeneous echogenic mass lesion with small cystic spaces.
b. Corresponding CT image demonstrates ill demarcated soft tissue mass lesion in the right axillary region and posterior mediastinum.
c. Histologic section reveals small lymphatic channels (L) surrounded by thick fibro-fatty septa. (Hematoxylin and eosin, X40.)
tension of a cervical lymphangioma into the submandibular gland was detected. Three patients with clinically suspected solitary lesion were found to have multiple lesions. Among 11 cervical lymphangiomas, five cases had contralateral or ipsilateral cervical lymphadenopathy on sonographic examination.

**DISCUSSION**

The lymphatic system arises from five primitive sacs derived from the venous system: paired jugular sacs lateral to the jugular veins and unpaired retroperitoneal sac at the mesenteric root, and paired posterior sacs near the sciatic veins. Extension from these sacs spread peripherally to from the peripheral lymphatic system (3). The exact etiology of lymphangioma is unknown but these lesions are thought to be caused by a growth anomaly or arrest in normal development whereby the lymphatic sacs fail to reunite with peripheral draining channels to flow into the sacs. The lymphatic tissues sequestered in primitive sacs form cysts, from which endothelial fibrillar membranes sprout, penetrate into surrounding

normal tissue, and subsequently canalize to produce more cysts. A lymphangioma is a tumor of the lymphatic vessels consisting of endothelial cells and supporting connective tissue, both of which participate in the neoplastic process. Some observers classify lymphangioma as hamartoma and vascular malformation, rather than true neoplasm, where as lymphangioma is composed of arterial and venous channels, lymphatic vascular spaces, and fibroadipose tissue stroma (1,4).

Histologically, Lymphangiomas have been classified into three types on the basis of the size of the lymphatic channels: (1) simple, made up of capillary-sized, thin-walled lymphatic channels with considerable connective-tissue stroma; (2) cavernous, containing actively growing, dilated lymphatic channels and lymphoid stroma; and (3) cystic lymphangiomas or hygromas, which are macroscopic multilocular cystic masses lined with a single layer of endothelium and containing serous or milky fluid. Separation of these types is difficult pathologically, since three types often coexist within a single lesion (2). The nature of the tissue surrounding a lymphangioma seems to dictate its histologic characteristics. In tissue with high tension such as muscle and dense fibrous tissue, a lymphangioma is usually capillary or cavernous, whereas in loose areolar tissue where the lesion is more expandable, cysts of various sizes develop, resulting in cystic hygroma. The larger the size of lymphangioma, the more cystic portions in the mass. Lesions arising in areas of loose tissue have well-defined borders, while lesions arising in areas of more compact tissue have poorly defined borders (2,10). These observations are consistent with those noted in our cases.

The greatest majority of lymphangiomas occur in the neck (75%) and axilla (20%); rare locations include the mediastinum, retroperitoneum, abdominal viscera, arm, back, parotid, scrotum, inguinal region, and bones (4,11). In our cases 48% occured in the neck. Clinically, 50 to 65% of cases are detected at birth and 90% of these
lesions are detected by the time of the patient is 2 years old, the age range corresponding to the period of greatest lymphatic growth. A few cases, however, have been reported in adults who are primarily in the fourth and fifth decades of life. Most lymphangiomas grow slowly and produce no symptoms, because of their soft yielding consistency and deep-seated location. Sudden enlargement can occur at any time due to infection (usually from the upper respiratory tract) or hemorrhage into the cysts (1,6).

With increasing role of ultrasonography in obstetric management, lymphangiomas are occasionally diagnosed prenatally. The sonographic features of lymphangiomas have been reported to be variable, some appearing as primarily cystic masses with linear septations (6-9) and others appearing more heterogeneous and containing large solid components (12). The sonographic features of lymphangiomas in our series were predominantly those of multiloculated cystic masses. These observations are consistent with those noted in previously published reports (6-8). The fluid content in those reports was usually of uncomplicated clear nature. In our experience, 11 lesions had diffuse, coarse echogenicity or dependent echoes in fluid of the majority of the cysts, reflecting complication by hemorrhage. The thickness and echogenicity of septa varied with the amount of connective tissue, muscle tissue, and adipose tissue present between the cystic spaces. Septa, when present, were usually thin in our cases. Sheth et al. described that solid components of varying sizes arising from the cyst wall or septa were principally caused by clusters of very small (few millimeters) dilated lymphatic channels and infrequently related to coagulated blood, calcified thrombus or focal mesenchymal proliferation near the cyst wall (7). Three cavernous lymphangiomas in our study appeared as echogenic mass with small cystic spaces. The clusters of very small dilated lymphatic channels, too small to be individually resolved with ultrasound is considered to be responsible for the predominantly echogenic nature of the mass. Small cystic portions are believed to be more dilated fluid-filled lymphatic spaces coexisting in a portion of the same lesion.

Differentiation of lymphangiomas from other fluid-filled masses may be possible with US imaging. In the cervical region where lymphangiomas occur most commonly, other congenital cystic lesions to be considered are branchial cleft cyst and thyroglossal duct cyst: the former is found anterolateral to the carotid sheath and anteromedial to the sternocleidomastoid muscle, and the latter is in the midline and can occur anywhere along the course of the duct from the foramen cecum to the pyramidal lobe of the thyroid gland. Both of these lesions also tend to be unilocular, unlike most cystic lymphangiomas. Other lesions included in differential diagnosis are abscess, hematoma, lymphadenopathy, teratoma, and hemangioma (5,7,13,14). Cavernous type lymphangiomas, smaller lesions, and lesions affected by inflammation or hemorrhage appear as echogenic or complex cystic lesions. In these cases, a relatively confident preoperative diagnosis of lymphangioma may be very difficult or differentiation from other diseases by sonography alone may be impossible. However, presence of a few anechoic cysts at the echogenic mass or multiloculated architecture retained in other portion of the tumor can be very helpful in making the correct diagnosis.

Surgical excision is the treatment of choice of lymphangioma and is successful in the vast majority of cases. The deeper and more invasive character of the lesions make complete excision almost impossible and accounts for recurrences. Delineation of the full extent, size, and location of a lesion preoperatively are critical in surgical approach and success. The information obtained with US imaging is helpful in determining the extent of the lesion before surgery and in assessing postoperative complications and recurrences (7,15).
In summary, a relatively confident diagnosis of lymphangioma can be made when the typical pattern of a fluid-filled mass with multiple thin septations is present on US imaging. Atypical and potentially confusing findings may be related to the presence of diffuse hemorrhage into the lesion or predominance of cavernous type. US imaging plays a useful role in the characterization of lymphangiomas and in the determination of tumor extent.

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소아에서의 림프관증: 초음파진단과 병리소견의 비교

경희대학교 의과대학 방사선과학교실, 단국대학교 의과대학 방사선과학교실*

이 선 화·류 경 남·윤 엽·유 필 문*

저자들은 림프관증 23예에서 관찰되는 초음파소견을 병리조직학적 소견과 비교분석하여 림프관증의 특정적인 소견 및 이의 병리학적 근거를 알아보고자하였다. 19예는 낭성 종괴로 17에는 다방성이고 2에는 단방성이었으며 이중 격막이 14예에서 압았고 에코발생의 고형부분이 6예에서 관찰되었다. 낭종내에 에코가 없는 액체가 8예, 에코발생의 액체가 11예였다. 4에는 다수의 작은 낭성부분을 가진 불규칙한 에코 발현의 고형성 종괴였고 나머지 1예에서는 다방성 낭성부분 및 에코발생의 고형성분이 공존하였다. 병리조직학적 비교에서 낭성부분은 격막으로 나누어진 내포세포로 둘러싸인 확장된 림프관에 일치하였고 에코발생의 액체는 출혈에 의한 것이었다. 에코발생의 고형성분은 두터운 섬유지방의 격막을 가진 매우 적은 림프관의 송이들과 대개 관련이 있었다.

림프관증의 특정적인 초음파소견은 압은 격막의 다방성 낭종이며 낭종내의 출혈은 흔한 합병증이었으며, 비정형적 소견은 출혈이나 혈만성성 림프관증의 존재를 의미한다. 이상의 소견으로 초음파 진단은 림프관증의 수술전 진단 및 수술적 치료방침 설정에 매우 유용한 검사임을 알 수 있었다.