Correlative imaging of cystic lymphangiomas: preliminary results.

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Learning objectives

The aim of this study is to illustrate the role of diagnostic imaging techniques in patients with cystic lymphangioma to clearly evaluate the anatomic as well as structural lesion features necessary for patient treatment planning.
Background

Cystic lymphangiomas are rare malformations characterized by cystic structure deriving from detachment of lymph sacs from venous drainage systems preferentially localized in the neck and axillary region while mediastinal and abdominal, both mesenterial and retroperitoneal, locations are very rare approaching 5% of cases [1].

Patients with cystic lymphangioma are often asymptomatic, but because of lesions growth they may present different symptoms depending on lesion location and size [2-4]; thus, lesion detection and characterization are clinically fundamental for patient management. For this purpose, several diagnostic imaging modalities have been proposed showing a predominant role of Ultrasound (US), Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) [5, 6].
Findings and procedure details

We analyzed the imaging results of seven patients with cystic lymphangiomas admitted in our department to evaluate cyst-like tumor masses clinically palpable or detected by ultrasound scan. Patients had different symptoms depending on the size and location of the tumor lesion; the main complaints consisted of dysphagia and dyspnea in cases of mediastinal localization and abdominal pain in case of abdominal lesions. The clinical characteristics of individual patients are illustrated in Table 1.

<table>
<thead>
<tr>
<th>Patients</th>
<th>Sex</th>
<th>Age</th>
<th>Symptoms</th>
<th>Anatomic Site</th>
<th>Fine Needle Aspiration (FNA)/ Surgery (S)</th>
<th>Pathology</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>27</td>
<td>Slight dysphagia</td>
<td>Neck-mediastinal aditus</td>
<td>FNA</td>
<td>Cystic Lymphangioma</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>60</td>
<td>Dyspnea</td>
<td>Right paracardiac region</td>
<td>FNA</td>
<td>Cystic Lymphangioma</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>32</td>
<td>Abdominal pain</td>
<td>Sub-hepatic space</td>
<td>S</td>
<td>Mesenterial Lymphangioma</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>67</td>
<td>Abdominal pain</td>
<td>Right iliac fossa</td>
<td>S</td>
<td>Mesenterial Lymphangioma</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>18</td>
<td>Abdominal pain</td>
<td>Hepatic hilum</td>
<td>S</td>
<td>Cystic Hemolymphangioma</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>60</td>
<td>Intestinal obstruction</td>
<td>Sub-hepatic space</td>
<td>S</td>
<td>Mesenterial cystic lymphangioma</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>47</td>
<td>Abdominal pain</td>
<td>Peritoneal space</td>
<td>S</td>
<td>Mesenterial cystic lymphangioma</td>
</tr>
</tbody>
</table>

Table 1 Patient’s clinical characteristics

Patients underwent initially US and then to CT and/or MRI. Pathology demonstrated mesenterial cystic lymphangiomas in 5 cases undergone surgical resection and chest cystic lymphangiomas in 2 cases undergone fine needle biopsy.
- Ultrasound study was performed using a 7.5-13 MHz linear probe for the evaluation of lesions of the neck and mediastinum and with a 3.5-5 MHz convex probe for the evaluation of abdominal lesions, integrated in both cases with color Doppler analysis.

- CT scan was performed with multi-detector computed tomography (MDCT) using a 64 -rows scanner with a detector configuration of 3 mm × 4 mm, table feed of 9 mm/s, rotation time 0.5 s, beam pitch 1.5, 1.5 mm reconstruction intervals, section thickness of 3 mm, 300 mAs, 120 kVp. A monophasic acquisition was performed 70 s after i.v. bolus (2 cc/s) injection of 150 cc of iodinated non ionic contrast media (Ultravist 370, Bayer Shering Pharma, Berlin, Germany); coronal reformatted images were then obtained with Multiplanar Reconstruction (MPR).

- Magnetic Resonance Imaging (MRI) was performed using a superconductive 1.5 -T magnet; the following sequences were used: axial T1-TSE (TR/TE= 217,8/4,6), axial and coronal T2-TSE-SSh (TR/TE=831/80 ms), axial, coronal and sagittal TRUFI single shot (TR/TE=364/1,3 ms). Subsequently, dynamic post contrast (0,1 mmol/kg Gd-DTPA Multihance, Bracco) T1-FFE sequence was performed with preset scan times and image acquisition in the arterial (30s), portal (60s), equilibrium (180s) and late phases (300s); axial and coronal planes were selected using a slice thickness of 3-5 mm.

The definitive diagnosis of cystic lymphangioma was obtained with abdominal laparotomy in 5 patients with mesenterial location and fine needle aspiration in 2 patients with mediastinal location.

In all patients the imaging findings of US, CT and MRI were concordant consisted of cyst-like tumor mass with different size ranging from 3 to about 10 cm localized in different anatomic sites: mesenteric region (n=5), mediastinal right paracardiac area (n=1) and mediastinal aditus (n=1); the results of imaging studies in all cases are illustrated in Table 2.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Lesion size (cm)</th>
<th>US</th>
<th>CT Pre-contrast density</th>
<th>CT Post-contrast density</th>
<th>MR T1 Signal Intensity</th>
<th>MR T2 Signal Intensity</th>
<th>MR T1 Post-Contrast</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5</td>
<td>Anechoic mass with regular margins and internal septa</td>
<td>Hypodense</td>
<td>Hypodense</td>
<td>Hypointense</td>
<td>Iso-hyperintense with internal septa</td>
<td>Contrast enhancement limited to walls and septa</td>
</tr>
<tr>
<td>2</td>
<td>7</td>
<td>Anechoic mass with</td>
<td>Hypodense</td>
<td>Hypodense</td>
<td>Hypointense</td>
<td>Iso-hyperintense with</td>
<td>Contrast enhancement limited</td>
</tr>
</tbody>
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<th>MR T1 Post-Contrast</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5</td>
<td>Anechoic mass with regular margins and internal septa</td>
<td>Hypodense</td>
<td>Hypodense</td>
<td>Hypointense</td>
<td>Iso-hyperintense with internal septa</td>
<td>Contrast enhancement limited to walls and septa</td>
</tr>
<tr>
<td>2</td>
<td>7</td>
<td>Anechoic mass with</td>
<td>Hypodense</td>
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### Table 2 Imaging findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Table Number</th>
<th>Lesion Description</th>
<th>Density</th>
<th>Signal Intensity</th>
<th>Contrast Enhancement</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>10</td>
<td>Anechoic mass with regular margins and internal septa</td>
<td>Hypodense</td>
<td>Hypointense</td>
<td>Homogeneously hyperintense with internal septa</td>
</tr>
<tr>
<td>4</td>
<td>3,5</td>
<td>Anechoic mass with regular margins</td>
<td>Hypodense</td>
<td>Hypointense</td>
<td>Homogeneously hyperintense with internal septa</td>
</tr>
<tr>
<td>5</td>
<td>10</td>
<td>Anechoic ovalar mass with internal septa and thin walls</td>
<td>Hypodense</td>
<td>Hypointense</td>
<td>Homogeneously hyperintense with internal septa</td>
</tr>
<tr>
<td>6</td>
<td>8</td>
<td>Anechoic mass with regular margins and internal septa</td>
<td>Hypodense</td>
<td>Hypointense</td>
<td>Homogeneously hyperintense with internal septa</td>
</tr>
<tr>
<td>7</td>
<td>15</td>
<td>Anechoic mass with thin and regular walls</td>
<td>Hypodense</td>
<td>Hypointense</td>
<td>Homogeneously hyperintense with internal septa</td>
</tr>
</tbody>
</table>

At US all the lesions were anechoic, with regular borders; 4 of 7 showed fine internal septimations while the remaining 3 presented homogeneous internal structure; color-
Doppler study revealed the presence of only peripheral and weak vascular flow (Fig. 1a and b). At CT the lesions also appeared as cystic masses with homogeneous internal low density and no contrast enhancement after intravenous injection (Fig. 2). MRI studies showed images of high contrast and quality; all masses were hyperintense on T2 weighted sequence, suggesting fluid content, with regular margins, thin walls and internal septa; after gadolinium administration, only walls and internal septa showed increased signal intensity on T1 weighted without internal contrast enhancement (Fig. 3, 4).

Cystic lymphangioma or cystic hygroma is a low-flow vascular malformation, developing where the lymph sacs are separated from the venous drainage system. The most common location is the neck region (75%), especially in the posterior triangle and in the back cervical cavity, but it can be also found in axillary region (20%) and infrequently in the mediastinum, retroperitoneum, mesentery, omentum, colon, pelvis, groin, bone, skin, scrotum and spleen; less than 1% of lymphangiomas are localized intra-abdominally and most of them occur in the mesentery followed by the omentum, mesocolon and retroperitoneum [7]. Mesenteric lymphangiomas may produce complications such as intestinal obstruction or volvulus, and infarction may occur [8]. Even if cystic lymphangioma can show atypical findings, such as heterogeneous content, irregular borders and calcification, the diagnosis is usually obvious in cases with typical features, showing fluid content and regular margins.

Diagnostic imaging has a fundamental role in the detection and characterization of the cystic component either with US and CT as well as with MRI, as reported in our series. In all our cases US has been the technique of first level in order to identify the lesion and to define its eco-structural cystic-type characteristics as well as tumor size. Diagnostic protocol was completed performing CT and MRI scans; both imaging techniques provided accurate images clearly illustrating loco-regional lesion spread; in particular, on TC scan tumor masses showed densitometric characteristics of fluid-type, regular margins and only capsular contrast enhancement. Magnetic resonance imaging, although less available than TC, allowed a clear evaluation of lesion morphology and structure, better showing vessel-like internal septa, wall thickness and fluid content, excluding the presence of mucoid, adipose or solid components.

Since the high contrast resolution of MRI, compared to CT, a better evaluation of internal lesion content, lesion wall thickness and locoregional tumor spread may be obtained using this technique which also provides a pre-operative differential diagnosis from other cystic-like masses [9,10]. In particular, at CT bronchogenic cyst could be misdiagnosed as solid mass because of its proteic content and even if at CT it can appear as a cystic mass with homogeneous attenuation, at T1-weighted MR imaging it could present a variable pattern because of variable content (protein, hemorrhage, mucoid material) with a fluid-fluid level at T2-weighted sequences. Furthermore, there are many other cystic lesions, such as cystic teratoma and epydermoid cyst that contain a certain amount of fat that can be easily detected with MRI [11]; moreover, epydermoid cyst presents a desquamated epithelial keratin content that shows increased signal intensity on Diffusion Weighted Imaging sequences (DWI) on MRI. Cystic lymphangioma may be
indistinguishable on MR imaging from cystic benign mesothelioma because of similar fluid content even if the latter is usually located on the surfaces of the pelvic viscera [12].

US is considered the first level study to investigate a suspected mass suggestive of cystic lymphangioma because of its non-invasiveness, low cost and non-use of ionizing radiation. However, US needs to be integrated with CT and MRI scans because of its non panoramic view and for obtaining more information about structural features and internal as well as peripheral enhancement patterns. CT scan is currently performed with multi-slice technique that allows volumetric acquisition of the selected anatomic region with multi-planar reconstruction using different methods (MPR, MIP, SSD, VR); however, CT imaging is performed with radiation exposure. Conversely, MRI can give specific information about the fluid content even when the CT shows greater intralesional attenuation values for the presence of proteinaceous material; moreover, MRI is helpful in surgical planning for its multiplanarity and high contrast resolution showing lesion loco-regional spread.
Fig. 1: A large (4.5x1.5 cm) capsulated anechoic lesion with regular margins and multiple internal septa (a). Minimal vascular signal of the internal septations was detected at color-Doppler (b). References: Romeo V, Maurea S, Guarino S. A case of lower neck cystic lymphangioma: correlative US, CT and MR imaging findings. Quant Imaging Med Surg. 2013 Aug;3(4): 224-7.

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Fig. 2: CT scans after intravenous administration of iodinated non ionic contrast agent showed a mass with fluid density and regular borders that extended itself in the upper mediastinum from the retrosternal region to the anterior wall of the esophagus, interposing to epiaortic vessels; no significant intra-lesional enhancement was observed. References: Romeo V, Maurea S, Guarino S. A case of lower neck cystic lymphangioma: correlative US, CT and MR imaging findings. Quant Imaging Med Surg. 2013 Aug;3(4): 224-7.

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**Fig. 3:** T2 TSE axial and coronal MR images: the mass is clearly hyperintense compared to the muscular tissues and showed internal septations, suggesting cystic structure; epiaortic vessels show a low signal intensity and are well differentiated by the mass. References: Romeo V, Maurea S, Guarino S. A case of lower neck cystic lymphangioma: correlative US, CT and MR imaging findings. Quant Imaging Med Surg. 2013 Aug;3(4): 224-7.

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Conclusion

In conclusion, we suggest that an appropriate diagnostic imaging protocol in patients with cystic lymphangioma should initially include an US study and a subsequent MRI scan with contrast administration. CT should be avoided because of radiation exposure. US and MRI may also be useful in the follow-up of patients who refuse surgical resection or in whom surgery is contraindicated or post-poned as well as to early detect a possible disease relapse.
References


