Case 1720

Combined Thoracic and Abdominal Lymphangioma in an Adult
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Section: Chest imaging
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Case Type: Clinical Cases
Authors: A. M. Fauquenot-Nollen, M. L. Plaisier, R. T. Tjon A Tham
Patient: 23 years, male

Clinical History:
Persistent cold without fever, no significant past medical history.

Imaging Findings:
A patient without significant medical history presented to our hospital with a persistent cold without fever. He had no abdominal complaints.
Physical examination revealed some dullness to percussion and reduced breath sounds at the base of the right lung. Routine laboratory tests were normal and HIV serology was negative. The chest radiograph showed an elevation of the right hemidiaphragm and scalloping of the left one. Subsequently abdominal ultrasound and CT scan of the thorax and abdomen were performed.
Radiograph of the chest, PA-view (fig. 1) shows marked elevation of the right and scalloping of the left hemidiaphragm. Ultrasonography of the abdomen (fig. 2) demonstrates a large multilocular cystic mass at the base of the right hemithorax. The cysts are separated by multiple thick septa.
Contrast-enhanced CT scan of the thorax (fig. 3) shows well defined fluid collections in both pleural spaces (A), and extension into the anterior, upper mediastinum (B).
On contrast-enhanced CT scan of the abdomen (fig. 4), the intrathoracic cystic lesions communicate through the hiatus of the anterior diaphragm with similar lesions in the left upper abdomen (A) and extension is seen through the mesentery with presence of multiple small and large cystic lesions (B). There is a ring-calcification at the right side. Thin needle aspiration of the lesion at the right pleural space yielded milky yellow-white fluid. Cytological analysis showed chylomicrons. At thoracotomy, a large lobulated cystic tumor was found located between the diaphragm and the right lower lobe of the lung and resected. The left sided cystic mass was not resected. At laparatomy, multiple small cysts and dilated veins were found. The abdominal cystic lesions could not be removed radically, but biopsies were taken.
Pathological examination confirmed the diagnosis of lymphangioma, with lesions of the cystic hygroma type in the thorax and a mixed cystic and cavernous type in the abdomen.

Discussion:
Lymphangiomatosis is a rare condition probably related to a developmental malformation of the lymphatic system. Lymphangiomas most often present during childhood or adolescence but may occur at any age, mostly with minor symptoms. A potential aggressive behavior has been described but is uncommon. They present as (pseudo-) cystic lesions with fluid attenuation. They seldom show intralesional calcifications. They tend to surround and invade
normal structures but have no malignant potential. Most commonly they arise in the head and neck (95%). Abdominal lymphangiomas arise in the mesentery and omentum but are extremely rare. Combination of an abdominal and intra-thoracic lymphangioma is seldomly found. Four histologic subtypes of lymphangiomas have been described:
- cystic hygroma,
- cavernous lymphangioma,
- capillary lymphangioma and the 
- vasculolymphatic malformation.
These types are considered as a spectrum of the same disease. Combinations of these four types may be seen in the same lesion. The presence of endothelial-lined lymphatic channels separated by connective tissue, is the dominating histologic feature. Anatomic location of the lymphatic malformation and size of the lymphatic spaces play an important role in determining the histologic type of lymphangioma.
The lesions in our patient were predominantly of the cystic hygroma type in the thorax and of the mixed cystic-cavernous type in the abdomen. A vasculolymphatic component in the abdomen is suggested based on laparoscopic and CT scan findings.
Differential diagnosis includes other cystic masses such as enteric duplication cyst, pancreatic pseudocyst, enteric and mesothelial cyst, teratoma and cystic pancreatic tumors. Mediastinal lymphangioma has to be differentiated from cavernous hemangioma or pericardial cyst. The simultaneous presentation of thoracic and abdominal lesions are in favor of the systemic lymphatic origin of the tumor. Treatment of lymphangiomas should be aimed at complete surgical excision whenever possible. Radiotherapy and sclerotherapy have also been described as treatment modalities for non resectable lesions.
**Differential Diagnosis List:** Lymphangioma

**Final Diagnosis:** Lymphangioma

**References:**

Figure 1

Description: Radiograph of the chest, PA-view shows marked elevation of the right and scalloping of the left hemi-diaphragm. Origin:
Description: Ultrasonography of the abdomen demonstrates a large multilocular cystic mass at the base of the right hemi-thorax. The cysts are separated by multiple thick septa. Origin:
Figure 3

a

**Description:** Contrast-enhanced CT scan of the thorax shows well defined fluid collections in both pleural spaces **Origin:**

b

**Description:** Contrast-enhanced CT scan of the thorax (fig. 3) shows well defined fluid collections and extension into the anterior, upper mediastinum (B). **Origin:**
**Figure 4**

*Description:* On contrast-enhanced CT scan of the abdomen, the intrathoracic cystic lesions communicate through the hiatus of the anterior diaphragm with similar lesions in the left upper abdomen.

*Origin:* 

*Description:* Extension is seen through the mesentery with presence of multiple small and large cystic lesions. There is a ring-calcification on the right side.

*Origin:*