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Case Report

# Lymphangiomatosis in a 14-year-old female presenting with chylothorax and multiple cystic lesions

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ABSTRACT

Lymphangiomatosis is a rare congenital disease; diagnosis is made in the first 2 decades and affects almost all body parts. Imaging findings play an important role in the diagnosis. We present a case of a patient with lymphangiomatosis whose diagnosis was made solely with imaging findings; we also include a small review of the topic.



## Presentation

Our case is of a 14-year old female without significant past medical history who presented to the emergency department with symptoms of dry cough, fever, and malaise. Physical examination revealed absent right lower breath sounds. The initial chest x-ray showed a right pleural effusion (Fig. 1). A right thoracostomy returned thick, white fluid consistent with a chylous pleural effusion. A chest computed tomography (CT) was subsequently obtained and demonstrated a nonenhancing fluid density and an anterior mediastinal mass without mass effect to adjacent structures (Fig. 2). CT of the abdomen and pelvis revealed nonenhancing hypodense splenic lesions as well as multiple lytic lesions in the lumbar spine (Fig. 2). Based on the CT findings, the treating team ordered a whole body magnetic resonance imaging (MRI) to seek lymphoproliferative pathology or infectious causes. In the MRI there were multiple T2 hyperintense lesions in the neck, anterior mediastinum, retroperitoneum, and in multiple bones such as the ribs, sacrum, vertebral bodies, and pelvis. The spleen had multiple nodular lesions, which were T1 hypointense and T2 hyperintense. In the pelvis there were also multiple cystic lesions. In the T1 post gadolinium the majority of the lesions did not enhance, except for some bony lesions that had peripheral ring-like enhancement and none of the lesions had a solid component (Fig. 3). In order to rule out malignancy, clinicians performed bone marrow aspirate and lumbar puncture, which were negative for malignancy. The patient was discharged with propranolol, alendronate, calcium, vitamin D1, and a low-fat diet. The patient returned 6 months later after the first visit, with similar symptoms and a whole body MRI was performed. This exam revealed hyper intense lesions in the spleen and bones and cystic lesions in the neck, mediastinum, and retroperitoneum, without a significant change

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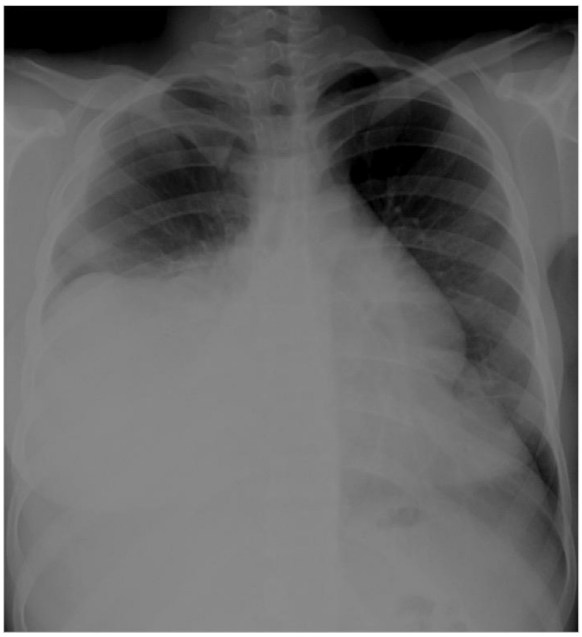

**Fig. 1 – Posterioranterior chest x-ray where right pleural effusion is seen.**

since the first magnetic resonance (Fig. 4). After this, the treating team ordered a lymphography (Fig. 5) to assess the patency of the lymphatic system. Interventional Radiology tried to access the lymphatic system, but was only able to demonstrate some lymphatics in the right extremity without pathologic changes.

As a palliative treatment, thoracoscopy, mechanical, and chemical (talc) pleurodesis was performed. Based on the clinical symptoms, laboratory findings and the imaging features the diagnosis of generalized lymphangiomatosis was made.

In the past, this diagnosis was made based on pathologic findings; today however, there are several imaging features that help us make the diagnosis without the need of a biopsy. These features are: the presence of cystic lesions with sharply defined margins and without contrast enhancement. Bone lesions with fluid signal in all sequences without periosteal reaction and osseous destruction [1,2] and of course, involving multiple organ systems [3].

## Discussion

Lymphangiomatosis is a congenital benign malformation of the lymphatic system that can be classified in capillary, cavernous, and cystic [3], it was described in 1828 by Dr. Rodenber for the first time [4], but still up to our time its etiology is unknown. The most accepted theory is that the perilymphatic vessels do not connect with the main lymphatic vessels.

Lymphangiomatosis can affect any part of the organism, but the brain (which is devoid of lymphatic channels), being more common in mediastinum, lungs, pleura, and bone. It can be solitary o multiple.

It is known that with the concomitant involvement of the osseous and thoracic system, the prognosis is usually poor [5]. This pathology has multiple synonyms like cystic lymphangiomatosis, diffuse lymphangiomatosis, and generalized systemic lymphangiomatosis.

Lymphangiomatosis is often diagnosed during the first 2 decades of life with no gender or race predilection and it is not an inherited condition. It knows that the visceral involvement is associated with a high mortality rate [6].

Usual clinical manifestations include nonspecific symptoms like dyspnea, chest pain, chylous pleural effusion, bone pain, pathologic fractures, joint deformity [5], abdominal pain, nausea, vomit, anemia, thrombocytopenia, and among others.

### *Embriology*

Lymphangiomatosis is a rare congenital lymphatic malformation originating from the persistence of dilated lymphatics at 14–20th week of life [7].

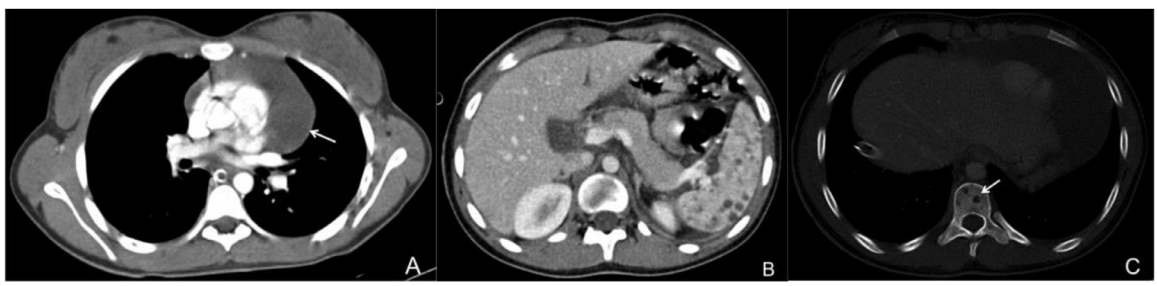


**Fig. 2 – Contrast computed tomography in soft tissue window (A and B) demonstrating a cystic lesion in the anterior mediastinum (white arrow) without mass effect and multiple hypodense lesions in the spleen, none of the lesions showed enhancement with the contrast. computed tomography in bone window (C) where multiple lytic lesions (white arrow) in the vertebral bodies without associated soft tissue mass are shown.**