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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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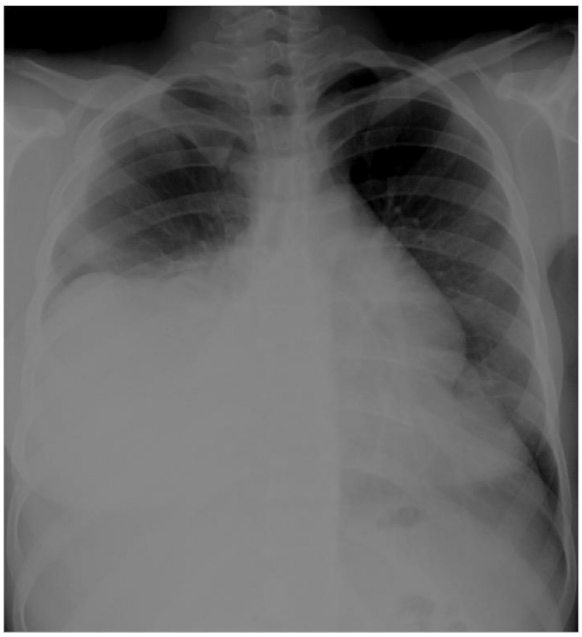

Fig. 1 – Posterioranterior chest x-ray where right pleural effusion is seen.

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].

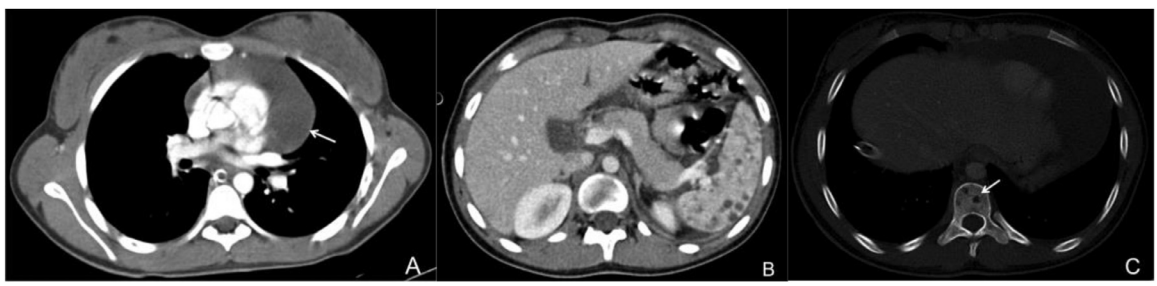

Fig. 2 – Contrast computed tomography in soft tissue window (A and B) demonstrating a cystic lesion in the anterior mediastinum (white arrow) without mass effect and multiple hypodense lesions in the spleen, none of the lesions showed enhancement with the contrast. computed tomography in bone window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

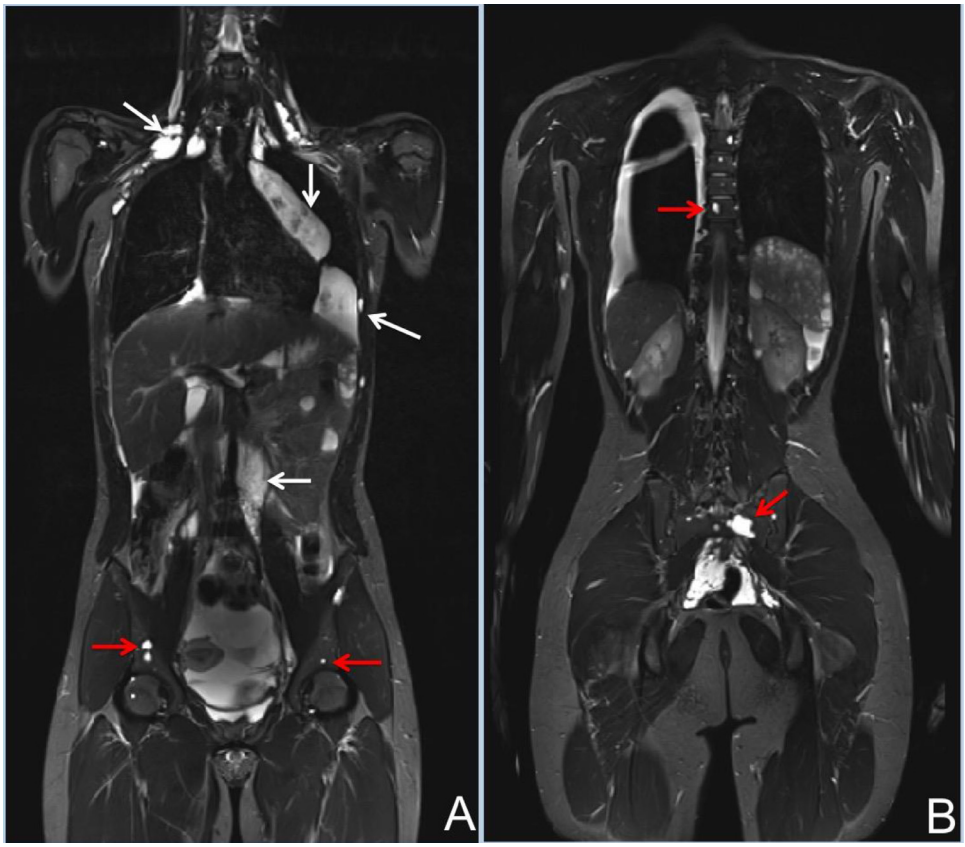

Fig. 3 – Coronal T2WI (A and B) showing multiple T2 hyperintense lesions in the neck, anterior mediastinum, retroperitoneum (white arrows), and osseous structures such as: ribs, sacrum, vertebral bodies, and pelvis (red arrow). The spleen had multiple T2 hyperintense nodular lesions. Coronal (C and D) and axial (E) T1 post contrast: the majority of the lesions did not enhance in exception for some bony lesions that had peripheral enhancement (red arrow). None of the lesions had a solid part. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

Lymphangiomas are benign lesions secondary to an abnormal lymphatic system development. Pathology findings include: endothelium supported by lymphoid tissue, smooth muscle, and round cells [3].

Imaging

The lymphatic malformation can be diagnosed with different imaging modalities (ultrasound (US), CT, and magnetic resonance) playing an important role in the diagnosis. If the findings are characteristic enough (cystic lesions) and multiorgan involvement is seen, biopsy can be excluded [3,7,8].

By US, cystic lesions are anechoic with no solid component, and with Doppler evaluation, no vascular structures are seen in the interior of the lesions. When the lesions are studied by CT, hypodense homogeneous lesions without enhancement are usually seen.

In MRI the cystic lesions are hypointense on T1WI and hyperintense on T2WI and there is no enhancement after IV gadolinium is injected. If uptake is present only mild peripheral enhancement may be seen [8]. In *diffusion-weighted* imaging (DWI), these lesions may show facilitated diffusion.

Associated infection may occur, under this scenario the lesions may show a peripheral ring-like enhancement with variable T1WI and T2WI signal [9].

Bone involvement has been described in almost 75% of the patients, being more common lytic type lesions of the skull, ribs, pelvis, femur, tibia, humerus, and vertebrae [4]. These lesions grow slowly and patients with this manifestation usually have a good prognosis [6]. With bone x-ray, well-defined, round, and radiolucent lesions are seen with multiple septae resembling a moth-eaten pattern [10].

Differential diagnosis include: Langerhans cell histiocytosis, hemangiomatosis, infection, multiple myeloma, and bone metastasis [10].

Thoracic presentation may involve the lungs, mediastinum, and the pleura. Findings may include soft tissue infil-

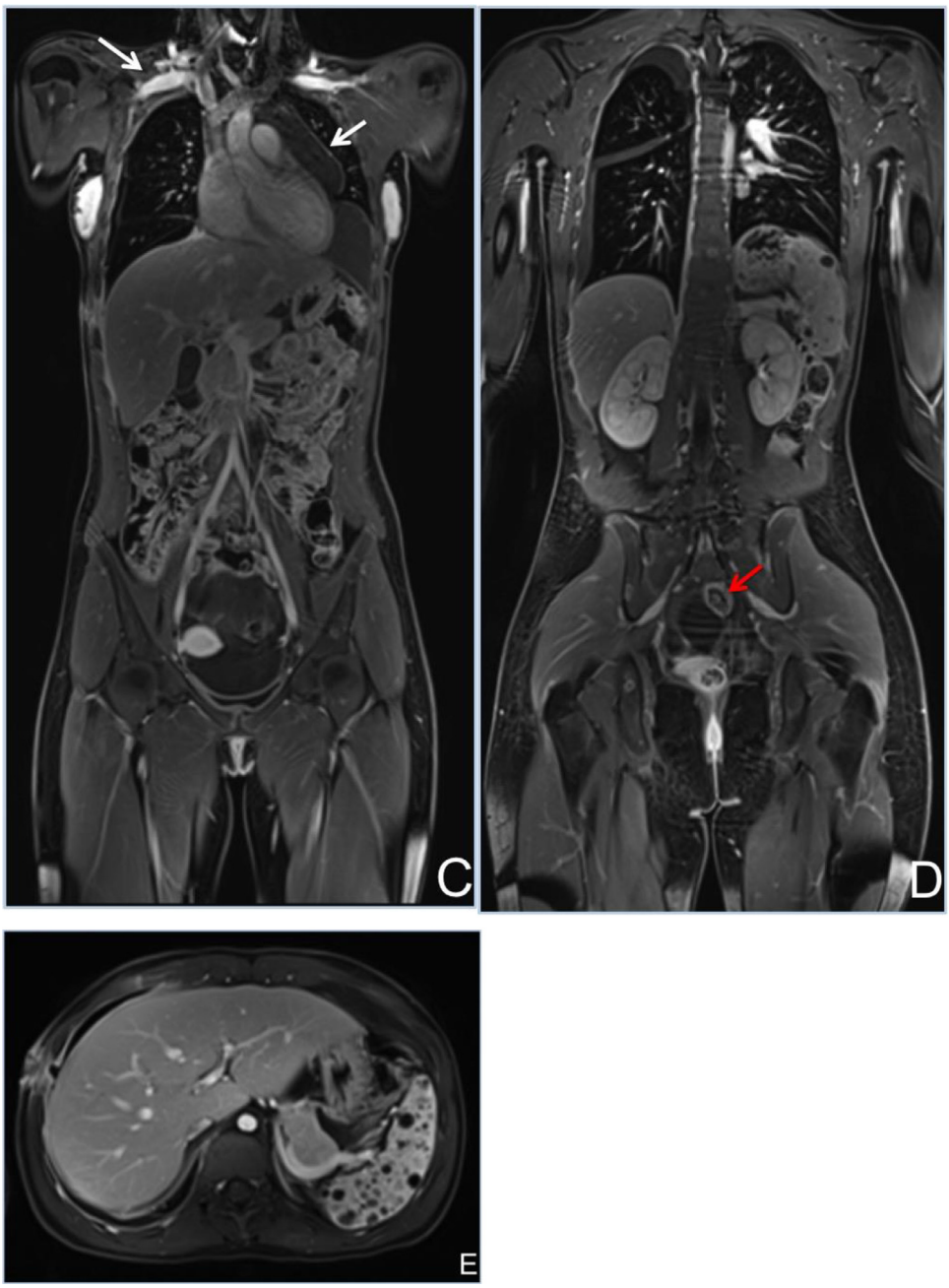

Fig. 3 – Continued

tration to the mediastinum, thickening of the peribroncovascular interstitium and ground glass opacities [11]. There may be chylous effusion with fat attenuation in CT and it is considered a sign of bad prognosis.

Mediastianal noncalcified masses with low attenuation may be seen with encasement of normal mediastinal structures, but without infiltrative pattern or displacement of adjacent structures [12]. The main differential diagnosis is lymphoma.

In the abdomen, there may be involvement of the peritoneum, retroperitoneum, solid organs, and in very rare cases, the mesentery. The thin-walled cystic lesions may complicate with spontaneous rupture, obstruction, and hemorrhage. The diagnosis of this pathology can be made with radiological and histologic findings, although under the microscope the disease may be misinterpreted with generalized fibromatosis and diffused hemangiomatosis.

Usually palliative treatment is preferred for pericardic and pleural effusions, but selected cases can be referred for embolization, sclerotherapy, or surgical removal.

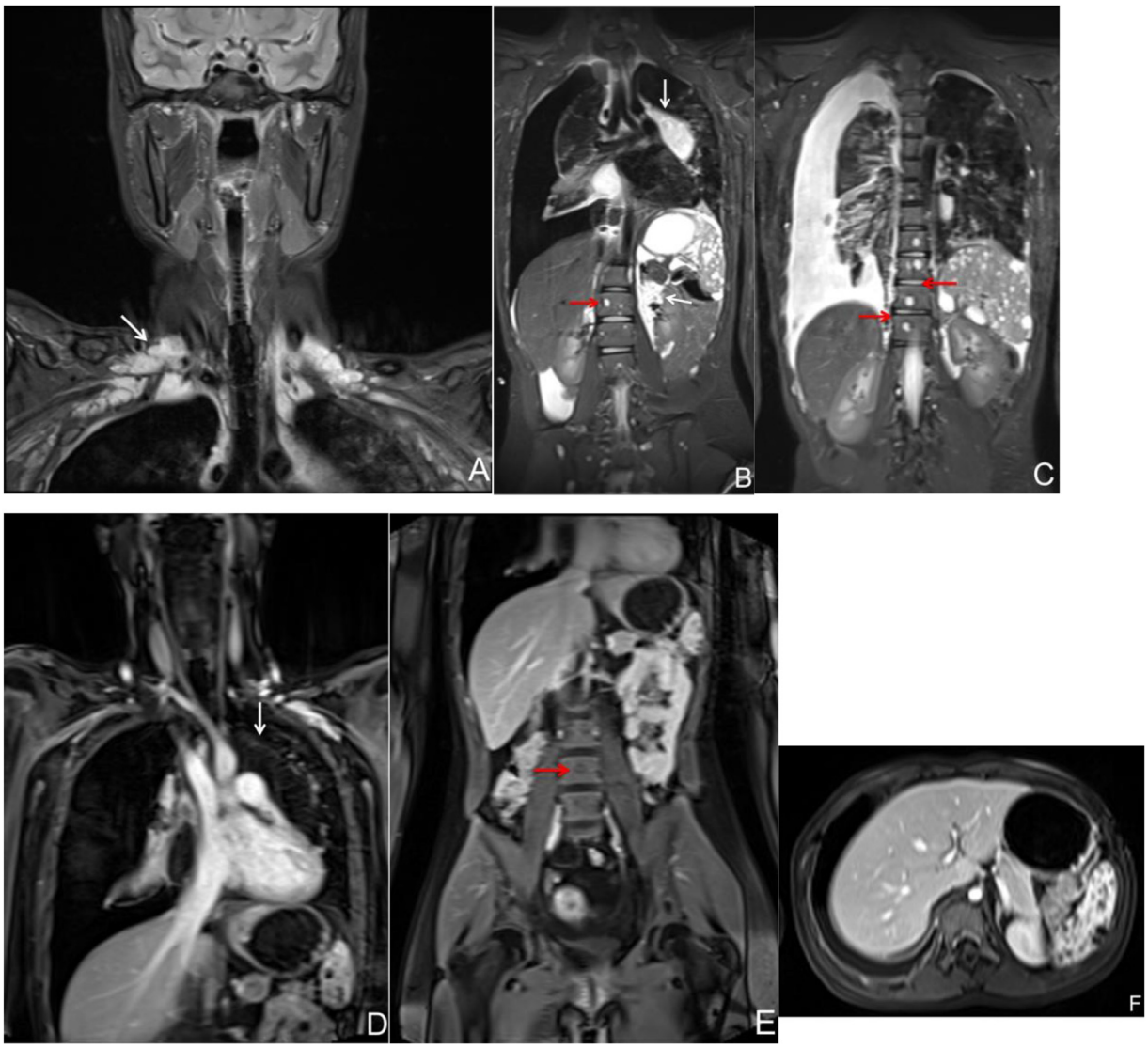

Fig. 4 – Coronal Short-TI Inversion Recovery (STIR) of the upper thorax (A) obtained 6 months apart. Lesions (white arrow) of the neck described in the past exam are stable in time. Coronal STIR of the thorax and abdomen (B and C) showing mediastinal and retroperitoneal lesions (white arrow) and multiple hyperintense lesions in the vertebral bodies (red arrows) and the spleen with no change. There is pleural effusion in the right hemithorax. Coronal T1WI post gadolinium of the thorax and abdomen (D and E) and axial T1WI of the abdomen (F) showing absence of enhancement of the mediastinal lesion (white arrow), bone lesion (red arrow), and spleen lesion. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

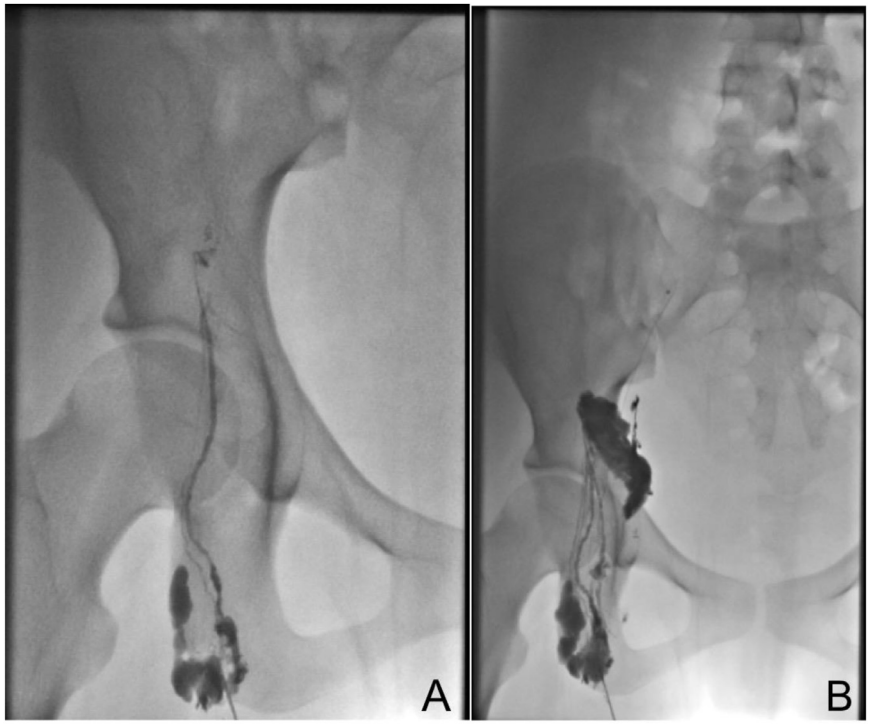

Fig. 5 – Right femoral lymphography (A and B) showing normal lymphatic structures.

There is usually good prognosis associated to this condition, but the course and symptoms of the disease are related to the organ and/or systems involvement [3].

Conclusion

Generalized lymphangiomatosis is a rare condition with variable prognosis and symptoms depending on the organ and/or system is involved.

Multimodality imaging techniques (US, CT, and MRI) play an important role on the assesment and diagnosis of generalized lymphangiomatosis as the diagnosis of this condition can be made based solely on imaging findings.

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