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Case Report

Abdominal Distension Revealing Retro-Peritoneal Cystic Lymphangioma

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Abstract: Cystic lymphangiomas are benign tumors of the lymphatic vessels. It is characterized by its clinical polymorphism. Its retroperitoneal localization is rare. Its diagnosis relies mainly on imaging but needs histological confirmation. Its treatment is surgical. It can have an adverse evolution, especially when its volume causes compressive complications. We report the case of a 65-year-old male patient who presented abdominal distension and pain, whose abdominal scan showed a voluminous retroperitoneal mass. The interest of our case resides in the impressive volume of the tumor, its expressive scanographic semiology and its favorable evolution.

Keywords: Lymphangioma, retroperitoneal, CT-scan.

INTRODUCTION

Cystic lymphangioma (CL) is a rare benign tumor of the lymphatic tract.

Its retroperitoneal location is rare. It is characterized by its clinical polymorphism. The diagnosis is made by imaging and requires histological confirmation. The treatment of choice is surgical.

Serious complications can be found, possibly compressive.

We report the observation of a 65-year-old man who consulted for abdominal distension.

PATIENT(S) AND OBSERVATION(S)

This is a 65-year-old man, with no particular pathological history, who consulted for a progressive abdominal distension for 5 years, complicated 2 months ago by abdominal pain and constipation.

Clinical examination revealed significant abdominal distension with eversion of the umbilicus and collateral venous circulation (Figure 1). Palpation revealed a large soft mass occupying the entire abdomen, mobile in relation to the superficial plane. The rest of the clinical examination was normal.

The biological workup was normal.

Abdominal ultrasound and CT scan showed a large right retroperitoneal cystic formation, multilocular, with exophytic development, measuring 35x28 cm and extending over 41 cm, with a thin and regular wall and fluid content, with multiple fine partitions.

It exerted a mass effect with scalloping on the liver above, the abdominal aorta and its visceral branches, the portal system, the right kidney below and its vascular pedicle, the digestive system all around, and the abdominal wall in front (Figure 2).

Surgical exploration revealed a large lobulated retroperitoneal cystic mass containing citrine fluid; a complete resection was performed.

Pathological examination of the surgical specimen concluded that it was a retroperitoneal cystic lymphangioma.

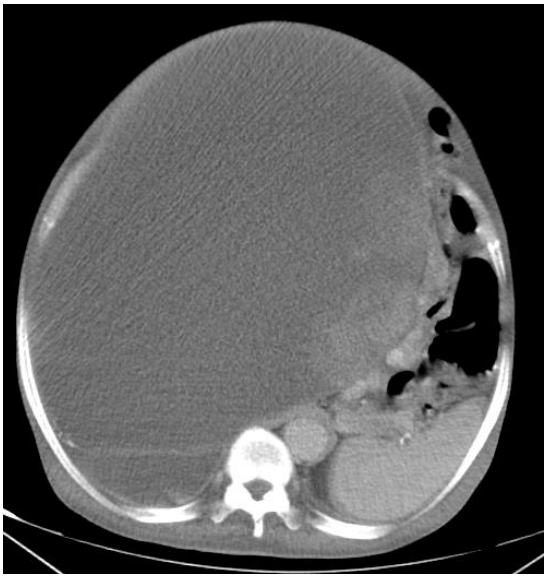

Figure 1: Axial section: Voluminous right retroperitoneal cystic formation, multilocular, with multiple fine partitions, exerting a mass effect on the abdominal aorta and the left kidney

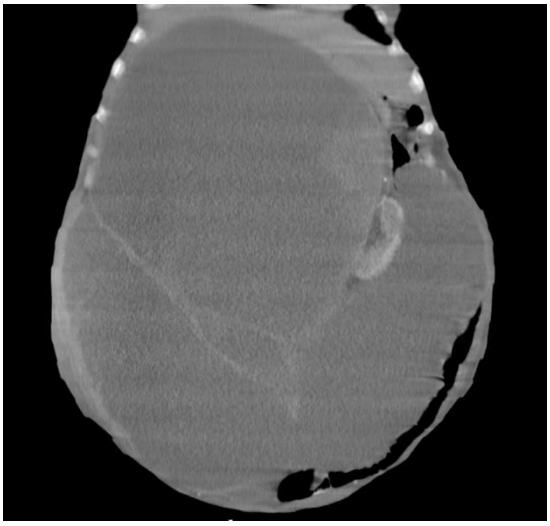

Figure 2: Coronal section: Voluminous retroperitoneal cystic formation, multilocular, with multiple fine partitions, exerting a mass effect with scalloping on the liver above, the left kidney below and behind and the digestive structures

DISCUSSION

Cystic lymphangiomas are rare benign tumors [1, 2] and mainly affect children [3]. In adults, they represent only 7% of abdominal cysts [4] with a sex ratio equal to 1.

Physiologically, two theories have been described to understand the origin of these cystic lymphangiomas.

The malformative theory is the most frequently found, given its frequency of discovery in childhood and during the antenatal period.

The second theory suggests an acquired origin resulting from obstruction of lymphatic vessels due to inflammation, trauma or degeneration [5].

The clinical picture of LK varies from patient to patient.

Its most frequent localization is the cervico-facial and axillary region [6], but it can also involve the mediastinum, the abdomen and the limbs. Retroperitoneal localization is very rare, as was the case in our patient.

In its retroperitoneal location, several symptoms have been described: asthenia, abdominal pain, vomiting, ascites, intestinal obstruction and edema of the lower limbs [6].

Imaging plays a key role in making the diagnosis [7].

Ultrasound is the first-line examination. It allows the identification of a well-limited fluid formation, with regular contours, uni- or multilocular, possibly with fine septa, with a transonoral content, which may however become echogenic in case of intracystic hemorrhage, or contain some calcifications [8, 9].

Computed tomography (CT) allows a better study of the tumor by objectifying a liquid formation, unilocular or multilocular, hypodense, with a thin wall, which can be the seat of fine partitions, not enhanced after injection of contrast medium.

Magnetic resonance imaging (MRI) is a second-line examination; it allows a precise diagnosis and a better analysis of the nature of the cyst contents, in hyposignal T1 and hypersignal T2, with partitions in hyposignal T1 and T2, without parietal or septal enhancement [12], and also to appreciate the perivascular extension of the lesion [6].

The diagnosis of certainty is provided by pathological examination. LK lesions are often whitish or translucent. They may be unilocular (25%) or multilocular (oligomacrocystic, micropolycystic or mixed) [6].

Different therapeutic means can be used, ranging from simple surveillance to open or laparoscopic surgical removal.

Surgery should be as complete as possible to avoid recurrence [13], while remaining conservative [8].

CONCLUSION

Cystic lymphangioma, although benign in nature, can have a noisy picture the larger its volume

becomes, as was the case in our patient. Its retroperitoneal localization being rare, imaging, particularly in cross-sections, allows to determine it with certainty. Its diagnosis of certainty is histological, and its treatment is surgical.

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