CASE REPORT

Retropancreatic cystic lymphangioma – considerations upon a case

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Abstract
Cystic lymphangiomas are benign tumors developing as an anomaly of the lymphatic channels. Most of them are discovered in children and are located mostly in the head and neck with no differences between sexes. Retroperitoneal locations are rare, clinical symptoms not specific and are discovered incidentally. We report the case of a 32-year-old female sex patient with a cystic tumor situated in the retroperitoneum, behind the pancreas, discovered by abdominal ultrasound during a diagnostic work-out for urinary infection and hypertension. Anamnesis, physical examination and CT-scan could not indicate the nature of the cyst. The cystic tumor had a shape of a butterfly, crossing the midline in front of the aorta and vena cava. A laparotomy was indicated and established the diagnosis of a cystic lymphangioma based on its macroscopic appearance. The tumor was approached from its right side by a generous Kocher maneuver and then from the left by disinsertion of the Treitz ligament and elevating the pancreatic body and splenic vessels. It was completely removed. Postoperative course was uneventful. Histopathologic examination confirmed the diagnosis of benign cystic lymphangioma. At 2 years of follow-up the patient was free of recidive.

Conclusions: Retroperitoneal cystic lymphangioma are rarely seen in adults. Precise diagnosis is made at laparotomy, imagistic test give details about walls, content, relationship with the surrounding structures and major blood vessels. Complete surgical resection represents the treatment of choice avoiding recurrence.

Keywords: cystic lymphangioma, retroperitoneal cyst.

Introduction
Cystic lymphangiomas are benign tumors developing as an anomaly of lymphatic channels [1]. Retroperitoneal location is rare, clinical symptoms are not specific, its discovery is incidental or when complications as internal bleeding, infection or compression of adjacent structures occur. The preoperative diagnosis is very difficult, a laparotomy being mandatory in most cases. We report a retroperitoneal retropancreatic cystic lymphangioma and its diagnostic and therapeutic management.

Patient, Methods and Results
A 32-year-old woman was admitted in our surgical department complaining of abdominal, non-specific pain located in the epigastrium combined with fullness and arterial hypertension. The history of her symptoms revealed an obscure onset, progressive and non-remissive for a few months. Clinical examination showed a moderate obesity, no other peculiarity being noticed. Vaginal and rectal examinations were normal. Laboratory usual tests did not reveal any abnormality.

Abdominal ultrasonography identified a cystic tumor behind the pancreas and in front of aorta and vena cava but it could not offer more precise information.

CT-scan revealed a collection, located behind the pancreas, well defined, with thin walls, extending from right to left across the midline, anterior to the kidneys and renal pedicles, with the shape of a butterfly, containing clear fluid with no deposits. This cystic mass was compressing the right renal vein and vena cava without invasion (Figure 1).

Figure 1 – Small arrows indicating the lymphangioma situated behind the pancreatic head anterior to the kidneys and renal pedicles, extending across the midline.
The patient had no previous history of pancreatitis and no biliary lithiasis was identified in abdominal ultrasonographic examination.

The diagnosis was set as retroperitoneal retro-pancreatic cystic mass. A laparotomy was proposed to the patient in order to establish the true nature of the cyst and achieve its excision.

At laparotomy, a cystic mass was identified behind the second part of the duodenum and the pancreas, prolonging to the left side and bulging behind the 4th part of the duodenum and in the root of the transverse mesocolon (Figures 2 and 3).

![Figure 2 – Right part of the cystic lymphangioma visible behind the pancreatic head and the duodenum (black arrows).](image)

Excision was carried out from the right side after a large Kocher maneuver and from the left side after mobilizing the angle of Treitz and opening the root of the mesocolon. The walls were thin, white and the content serous.

![Figure 3 – Left part of the cystic lymphangioma (arrow) visible behind the angle of Treitz, root of the mesocolon and inferior mesenteric vein.](image)

The diagnosis of cystic lymphangioma, probably benign, was set upon its macroscopic appearance. The tumor was excised completely. No intraoperative accidents or incidents were noted and a perfect hemostasis was achieved. Postoperative course was uneventful.

Pathological examination showed cystic areas of variable size walled with single layer endothelia in a fibrovascular stroma with lymphocytes (Figure 4), disorganized smooth muscle cells (Figure 5) and lymphoid aggregates (Figure 6).

![Figure 4 – Cystic areas of variable size, walled with a flattened endothelia in a fibrovascular stroma with lymphocytes (HE stain, ×100).](image)

![Figure 5 – Detail: cystic areas walled by a single layer endothelia with disorganized strips of smooth muscle (HE stain, ×200).](image)

![Figure 6 – Cystic areas with intralumenal endothelial papillary proliferations in a fibrovascular stroma with lymphoid aggregates (HE stain, ×40).](image)

The patient was discharged at the 6th postoperative day. CT scan control at 1-year follow up showed a small incisional hernia in the upper part of the cicatrice and no recurrence of the cystic mass.

**Discussion**

The etiology of the lymphangiomas is not completely understood. It is believed that they develop from lymphatic vessels that dilate progressively because of insufficient drainage due to atresia or inadequate efferent channels [1]. The vast majority of them – 90%
are discovered before the age of two, and half of them at birth. There is no difference in incidence between sexes and most of them are located in the upper extremity of the body – head and neck [2]. Less than 1% is intra-abdominal [3]. The location in the retroperitoneum is even rarer. Less than 200 cases were reported in the international literature comprising all ages [4]. They are usually asymptomatic; their discovery is incidental, during abdominal ultrasound or computed tomography performed for an unrelated reason or at surgery. The cysts vary in size and shape from a few centimeters to a mass that fills the entire abdomen [5]. They are usually round and can be single or multiple, multiloculated, or uniloculated. They rarely produce symptoms by sudden enlargement as probably due to internal bleeding, infections or by compression to adjacent structures [6, 7]. Symptoms are usually related to its size. When they become large enough, as in our patient, they can displace or compress the bowel or urinary tract and present as chronic abdominal or back pain, or as a sensation of fullness or distension. Approximately 40% of cases present with symptoms of intestinal obstruction [8]. Other symptoms include ascites and intermittent fever. Rarely, bleeding, infection or rupture will cause an acute abdomen [9]. In our case, the presence of the cyst was revealed by pain and a sensation of fullness in the epigastrium. Morbid associations are also described. Papadopoulos VN et al. [10] found a retroperitoneal lymphangioma situated behind the left kidney associated with urinary bladder litiasis, renal cyst and sigmoid adenocarcinoma. In our case, the cyst was associated with moderate obesity and arterial hypertension. The first imagistic test usually performed is abdominal ultrasonography (US). In our case, it did not reveal any abnormality in the liver nor the gallbladder, but a cystic tumor behind the pancreatic head in relationship with vena cava and aorta. Usually, abdominal US is signaling the presence of the cystic tumor but CT-scan or MRI are necessary to better characterize the formation and its relationship with other structures [11].

CT-scan is considered the ideal diagnostic tool for assessing retroperitoneal tumors because it provides important information: lesion location, size and shape; the presence and the thickness its wall; the presence of septa, calcifications or fat; involvement of adjacent structures [2]. At CT, cystic lymphangioma typically appears as a large, thin-walled, multiseptate cystic mass with fluid content with attenuation varying from that of fluid to that of fat. An elongated shape and a crossing from one retroperitoneal compartment to an adjacent one are characteristic of the mass. Rarely, cystic lymphangiomas may have wall calcification [12]. In our case, there was a strange form of a butterfly or of the letter “H”. It also offered more information about the relationship with the pancreas, duodenum, retroperitoneal major blood vessels – aorta and vena cava and the upper mesenteric vessels.

The most difficult aspect of the diagnosis is to establish the true nature of the cyst. Randin R et al. [13] published a case of retroperitoneal cystic lymphangioma situated behind the left kidney in which the ultrasonography and CT-scan only suggested a cystic mass with low density fluid (-15 Hounsfie1d units) but the precise diagnostic was established after a guided puncture removing chylous fluid and biperped lymphography showing some lymphatic channels extending to the tumor (the abdominal X-ray immediately after injection) and several globules of contrast material inside the cystic tumor at 24 hours.

Differential diagnoses that should be considered generally for cystic lesions in the retroperitoneum include a large array of entities: duplication cysts, pancreatic pseudocysts, ovarian cysts, hematomas, sarcomas, teratomas, abscesses, retroperitoneal nodal metastases, retroperitoneal sarcomas, and also other benign conditions, including hematoma, lymphocele secondary to prior surgery or abscess, that may be found in this area [2].

An important significant differential diagnosis was about a cystic tumor pertaining to the pancreas. An endoscopic ultrasound could theoretically offer more information about the tumor and its relationship with the pancreas and the pancreatic ducts. It could guide a puncture with fine needle aspiration for biopsy, cytology examination, dosage of amylases, tumor markers and mucin in order to help distinguish between a benign or a malignant cystic tumor of the pancreas. This or laparotomy were discussed in staff. Finally, the decision was made in favor of the last for two reasons: first, the fine needle aspiration carries a risk for false negative or incomplete information, and second, the lesion should anyway be removed. In our case, the anamnesis of the patient excluded abdominal trauma, acute pancreatitis or previous surgery and the tumor clearly delineated from the pancreas in CT-scan.

No other abdominal or genital lesion was found. Laparotomy with biopsy and pathological examination is the attitude supported by the majority of the studies in the literature. The intraoperative macroscopic aspect suggested the diagnosis of cystic lymphangioma. The tumor was completely removed and the diagnosis was confirmed by histopathologic examination extemporaneous and after paraffin inclusion. Surgical resection is the treatment of choice. Complete resection should be performed whenever possible. Some authors state that if there has been invasion of other tissues then complete excision including the affected area of the involved organ is necessary. Others consider that if major structures are at risk during resection a partial resection should be performed to avoid high morbidity and mortality [14] and because of the benign nature of the cyst [15]. After complete excision recurrences can also occur but the rate is much lower at 7% [16]. Hauser H et al. [17] considers that the stem of the cyst and the feeding lymphatic vessels must be ligated to avoid chylous ascites postoperatively.

Laparoscopic excision was also proven feasible [5]. Partial resection is associated with a high recurrence rate, 50% in one series after a mean follow-up period of two years [4].

Aspiration, cystenterostomy and peritoneal cavity
marsupialization as conservative alternative methods to partial resection are usually followed by early recurrence. Drainage and subsequent partial excision of the cystic lymphangioma are soon followed by recurrence of the mass [16].

Other therapeutic measures that have been tried are diathermy, radiotherapy and percutaneous sclerotherapy. Sclerotherapy was accomplished with success [18, 19]. There is still much controversy about the agent that is rarely seen in adults. Precise diagnosis is made at non-surgical candidates [18, 23]. Complications as induration of the cyst and infection can occur after these procedures [4].

Conclusions

Retroperitoneal retropancreatic cystic lymphangioma is rarely seen in adults. Precise diagnosis is made at laparotomy, imagistic test give details about walls, content, relationship with the surrounding structures and major blood vessels. Surgery with complete resection represents the treatment of choice avoiding recurrence.

References


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