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#### **Case Report**

Visceral Surgery

# Gastro-Intestinal Bleeding Indicative of Retro-Duodenopancreatic Cystic Lymphangioma: A Case Report

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#### Abstract

Cystic lymphangioma is a rare malformative congenital tumor of the lymphatic vessels. It is commonly seen in children and mainly occurs in the head and cervicothoracic region. Abdominal and especially retroperitoneal involvement is rare in adults. This tumor is often asymptomatic but it can be manifested by mass effect with polymorphic symptomatology depending on the location and size. Digestive haemorrhage due to retroperitoneal cystic lymphangioma with posterior duodenopancreatic infiltration in an elderly patient is an exceptional manifestation of a benign tumor. We report a case of retro-duodenopancreatic cystic lymphangioma revealed by gastrointestinal bleeding in a 65 year-old man who was treated by tumor resection with organ preservation. The recovery was complicated by a duodenal fistula with spontaneous dry-up within 17 days, the patient left the hospital in 3 weeks.

Keywords: Cystic lymphangioma, Retroperitoneum, Duodenopancreas, Gastrointestinal haemorrhage.

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## **INTRODUCTION**

Cystic lymphangioma (CL) is a benign vascular tumor resulting from an abnormality embryological development of the lymphatic system. It is rare in adults and uncommonly occurs in the retroperitoneal regions [1, 2]. Only a few cases of simultaneous lymphangiomatous infiltration of the 2nd duodenum and pancreas in adults has been described in the literature. We report herein a new case of this exceptional clinical situation with a brief review of the relevant literature.

## CASE REPORT

A 69-year-old patient, treated for pulmonary tuberculosis and urothelial carcinoma in 2020, a chronic smoker recently stopped and a chronic alcoholic weaned 6 years ago. He had been followed in the gastroenterology department for moelena complicated by an anemic syndrome without other digestive symptoms for 9 months. The upper endoscopy and total colonoscopy were initially normal. The clinical course was marked two months ago by a rectal bleeding complicated by a severe anemia with a hemoglobin level of 3.6 g/dl for which the patient was transfused with 6 red blood cells volumes in the emergency room. At the moment, esogastroduodenal fibroscopy revealed a friable white polypoid formation at the posterior wall of the 2<sup>nd</sup> duodenum, but the histological examination of biopsy fragments was non-specific. The patient finally joined our department for surgical treatment. He was in a good general condition, apyretic, hemodynamically and respiratory normal. No tenderness or palpable mass on abdominal examination. Rectal touching revealed the moelena. Laboratory investigation showed hemoglobin level decreased to 9 g/dl, all other biological analyzes were unremarkable. Abdominal CT scan showed an irregular and lobulated retro-duodenopancreatic mass, measuring 99/66 mm and extended over 107 mm. It circumscribes the head and isthmus of the pancreas and maintains a thin line of separation with the superior mesenteric artery. Complementary MRI showed a thinly septated duodeno-pancreatic cystic mass (T1- hypointense and T2-hyperintense), measuring 97/90 mm and extending over 107 mm. The septa are discreetly enhanced after gadolinium injection. This formation molds the adjacent structures in particular; the inferior vena cava, the liver and the lesser omentum. It is associated with infiltration of the posterior duodenal wall and the posterior surface of the pancreas (fig.1).

The patient underwent open surgery, the exploration revealed a retroperitoneal polycystic mass infiltrating the entire posterior surface of the pancreas, the posterior duodenal wall as well as the root of the mesentery (fig.2). Complete resection was impossible without mutilating surgery with total duodenopancreatectomy. Considering the heaviness of

radical surgery (R0) and its high morbidity and mortality paradoxically to the benignity of the tumor, we performed an incomplete tumor resection (R2) with preservation of the pancreas (infiltrated on its entire posterior surface). The area of duodenal invasion that caused the bleeding was resected with macroscopically healthy margins. A direct duodenal split-stitch suture with 3/0 PDS was performed and followed by total duodenal exclusion. Histological examination of the surgical specimen confirmed the lymphangiomatous nature of the tumor (fig.3). The postoperative course was complicated by a duodenal fistula with spontaneous dry-up within 17 days, and the patient was discharged in 3 weeks.



Fig-1: MRI view of the abdomen shows large duodenopancreatic multicystic mass measuring 97/90/107 mm.



Fig-2: Intraoperative view showing a large retroperitoneal mass with extensive duodenopancreatic invasion. 1-Tumor, 2- Second part of duodenum.



Fig-3: Benign cystic tumour proliferation consisting of dilated vascular structures lined by flattened endothelial cells (HE, G x 400).

# **DISCUSSION**

Cystic lymphangioma account about 5% of benign tumors in children and adolescents without gender predominance. It is very rare in adults [3, 4]. The craniofacial, cervical and axillary regions represent the most frequent locations. Intra-abdominal forms are rare (less than 10%) and mainly affect the mesentery, however, hepatic, splenic, pancreatic, renal, adrenal, colic and duodenal involvment has been reported. Retroperitoneal location is even rarer than intraabdominal 5]. Pancreatic forms [4, cystic lymphangioma was first recognized in 1913 [6], it currently accounts for less than 1% of all lymphangiomas and less than 5% of pancreatic neoplasms [7]. Gastrointestinal involvement is now more frequently discovered due to the easy and frequent use of endoscopy to explore the digestive tract [8], but the number of published cases remains very low.

The etiopathogenesis of cystic lymphangioma is controversial with several hypotheses suggested; acquired such as the trauma, inflammation and infection, hormonal such as the impact of lymphangiogenic growth factors and neoplastic such as lymphatic ducts degeneration, but the congenital cause is the most likely [3, 6]. The latter is related to an abnormal connection between a group of abdominal lymphatic chains and the venous system with isolation of a lymphatic bud which would progress to cystization [9, 10].

CL is often asymptomatic, discovered incidentally during imaging for other indications. It may be revealed by non-specific gastrointestinal symptoms such as abdominal pain and nausea /

vomiting or exceptionally by complications such as rupture, torsion, infection, intracystic hemorrhage, fistulization, infiltration of adjacent structures and malignant degeneration. A palpable abdominal mass with or without signs of compression has also been described [4, 11]. Gastrointestinal hemorrhage is an uncommon event in the CL [4, 12], however it was the main symptom in our case. Imaging exams help to guide the diagnosis, in fact the abdominal ultrasound shows a well-limited, unilocular or multilocular fluid mass with thin septa which is transonorous but may become echogenic in the case of intracystic hemorrhage. CT scan and MRI allow to study the characteristics of the tumor: density, MRI signal intensity, topography, size and relationships with adjacent organs. Endoscopically, cystic lymphangioma of the digestive tract presents as small polyps or a mass that may cause bowel obstruction [8].

All these modalities can not specify the diagnosis in front of the very wide range of differential diagnoses [6, 13, 5]. The CL can be confused with many cystic pathology of the pancreas such as serious and mucinous cystadenoma, intraductal papillary mucinous neoplasm, pancreatic pseudocyst, congenital cyst and hydatic cyst [14]. The differential diagnosis may include lymphoma, digestive duplication and postoperative lymphocele [12]. Lymphangioma which is a primary lymphatic malformation should also be distinguished from lymphangiectasia or secondary obstructive dilation of the lymphatic vessels mainly in gastrointestinal involvement. This distinction is made more difficult by the histological over-lap [8]. Diagnostic certainty can only be obtained by histology. Macroscopically, lymphangioma lesions are white or translucent with serous or chylous content, unicystic (25%) or multicystic (75%). Microscopically, the cyst wall is bordered by endothelial- type cells (confirming the vascular origin) resting on fibrous tissue containing lymphocyte islands and sometimes smooths muscle fibers [15].

Complete surgical excision is the standard treatment for symptomatic tumors. Its extension to adjacent organs requires one-piece resection surgery to avoid recurrence [3]. This attitude is not always easy in front of extensive infiltration of several organs which the removal is burdened with a high morbidity and mortality. The risk of recurrence after total resection is very low, but in the event of incomplete resection, recurrence may affect one in two patients but the overall prognosis is good [16].

## CONCLUSION

Although it is a benign tumor, retroduodenopancreatic lymphangioma when it infiltrates the digestive wall can cause severe gastrointestinal bleeding. Therefore, it should be included in the list of etiological diagnosis of digestive hemorrhage. In addition, simultaneous and extensive infiltration of the pancreas and the duodenal wall complicates therapeutic management and may require simple reduction surgery with a high recurrence rate. Indeed, weighing the advantages (avoiding recurrence) and the disadvantages (high morbidity and mortality) of complete and major surgery of a benign tumor makes the therapeutic decision difficult.

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