Large Cystic Mass of the Pancreatico-Duodenal Region Revealing Duodenal Lymphatic Malformation: A Case Report

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ABSTRACT: Lymphatic malformations are benign tumors rarely located in the gastro-intestinal tract. They are usually asymptomatic; however, they can cause clinical discomfort when enlarged.

Imaging leads to diagnosis, but the confirmation can only be done through histology findings.

We report the case of a 59-year-old male patient, admitted in our ward for melena, to which esophago-gastroduodenoscopy showed budding formations in the second portion of the duodenum wall, with Magnetic Resonance Imaging features and histological examination confirming diagnosis of a duodenal lymphatic malformation.

KEYWORDS: Duodenal, pancreatic, lymphatic, malformation, imaging

Introduction

Lymphatic malformations are benign tumors rarely located in the gastro-intestinal tract. They can be caused either by a congenital malformation or inflammation or trauma. They may remain asymptomatic for a long time, however, when enlarged, they can cause compression of adjacent organs. Clinical symptoms are vague, and imaging is the examination of choice to lead diagnosis though ultrasound, Computed Tomography, and Magnetic Resonance Imaging (MRI). Diagnosis confirmation can only be done through histological findings.

We report the case of a 59-year-old male patient, admitted in our ward for melena, to which esophago-gastroduodenoscopy showed budding formations in the second portion of the duodenum wall, with MRI features and histological examination confirming diagnosis of a duodenal lymphatic malformation.

Case Description

A 59-year-old man, with a medical history of chronic smoking, pulmonary tuberculosis, and a high-grade urothelial carcinoma treated surgically, was admitted to the emergency room of the Military Teaching Hospital Mohamed V of Rabat for melena evolving for 4 months.

Clinical examination revealed a conscious pale and asthenic patient, with normal vital signs, no fever, and a normal abdominal examination, without any noted pain or palpable mass.

Laboratory tests revealed an isolated anemia at 4.9 g/dl, and the patient received 3 units of blood bags.

We performed an esophago-gastroduodenoscopy that revealed several budding non-ulcerative formations, including one located in the second portion of the duodenum (D2) measuring 2 cm that was bleeding spontaneously.

An abdominal MRI, performed, showed a large cystic, lobulated mass in the duodeno-pancreatic region, with low signal intensity on T1 weighted images and high signal intensity on T2 weighted images, containing thin septations, enhanced with contrast injection, measuring 97 × 90 mm.

This mass was budding in the duodenal wall causing a small defect, revealing duodenal invasion, and presenting a tight contact with the head of the pancreas with irregularity of its surface (Figure 1).

Histological examination revealed dilated lymphatic vessels in the duodenal mucosa in favor of a duodenal lymphatic malformation (Figure 2).

The patient was transferred to the surgery department and underwent a duodenectomy and mass resection with conservation of the pancreas, due to heavy follow up and risks after cephalic duodeno-pancreatectomy.

The patient was discharged 1 week later, with a good follow up.
Lymphatic malformations are benign tumors that form from the lymphatic system, usually found in the neck, head (75%), or axillary region (25%), discovered mostly among children, but can be diagnosed at any age. Localization in the gastrointestinal tract is very rare (<1%) and can be localized in the esophagus, stomach, pancreas, small bowel, and colon.

These lesions are either congenital because of a malformation in embryological development of the dorsal mesoduodenal lymphatic system in the third month of gestation such as sequestration and absence of communication with the lymphatic tissue that causes it to dilate and form a cyst, or it can be caused by trauma, inflammation, radiation therapy, degeneration of lymphatic ducts, or a lymphatic system obstruction. However, its etiopathogenesis remains unclear.

There are 3 types described: Cystic, capillary, or cavernous:

- Cystic lymphatic malformations are formed by cystic formations separated by fibrous septations, containing either a serous, chylous, or hemorrhagic fluid.
- Cavernous lymphatic malformations are more of a large cystic compressible mass.
- Capillary lymphatic malformations are small, dilated lymphatic vessels with a rich cellular stroma.

Cystic and cavernous lymphatic malformations can be caused by sequestration of the lymphatic system and differentiating between both of them can be hard.

Cystic lymphatic malformations occur in the abdominal cavity or in the retroperitoneum.

They are generally asymptomatic, but may cause chronic discomfort and symptoms in adults when enlarged, such as: a palpable abdominal mass, abdominal distension and discomfort, constipation, vomiting and as these symptoms vary and are non-specific, diagnosis remains difficult and imaging is required to lead it.
Imaging modalities include ultrasonography, Computed Tomography (CT), and Magnetic Resonance Imaging (MRI). Ultrasound shows a cystic lobular mass, that’s anechoic with some internal septations.

CT and MRI show a well circumscribed, multilocular, cystic mass, hypodense in CT scan, with low signal in T1 weighted images and high signal in T2 weighted images in MRI, containing septations enhanced by contrast.3

CT and MRI allow better evaluation of the lesion: its borders, origin, extension, and invasion of adjacent organs. It may also indicate if its content is cystic, chylous, or hemorrhagic.1

When this lesion is localized in the pancreas, diagnosis is hard to establish, and differential diagnosis is the mucinous cystadenoma of the pancreas.

When the lesion is very large, its origin is hard to recognize through imaging alone.3

With its atypical localization, histological examination is required to confirm diagnosis.

Its macroscopic aspect is a cystic multilocular lesion separated by septations, and microscopic examination shows dilated lymphatic formations with a single layer of flattened endothelium made of fibrocollagenous and lymphoid tissue.1

Treatment of lymphatic malformations of the pancreaticoduodenal region consists of radical excision of the mass, to avoid further invasion of adjacent organs and recurrence. When the mass is large, small bowel resection or cephalic duodenopancreatectomy may be necessary.3

Another alternative treatment that may be used is an ultrasonography fine needle aspiration.3

In case of incomplete resection, or large abdominal masses that can’t be operated, a sclerotherapeutic product: “OK-432” (containing a lyophilized mixture of a low virulence strain of group A streptococcus Pyogenes incubated with benzylpenicillin) may be injected in the mass.5,6

If the mass is asymptomatic, a simple follow up is required.3

Conclusion
Lymphatic malformations are benign tumors, rarely localized in the duodenum or pancreas therefore its diagnosis is hard to suggest at first but shouldn't be neglected with patients admitted for a pancreatico-duodenal asymptomatic or enlarged lobulated cystic mass. Imaging is the examination of choice to lead to diagnosis, and histological examination is necessary to confirm the diagnosis.

Author Contributions
LB collected imaging of the patient and wrote the discussion with all the radiology features; RS corrected the article as a radiology professor; KB and BA collected clinical informations, did the clinical examination of the patient and wrote the case report; TA corrected the article being a gastroenterology professor; MAE and AK collected histological examination images and corrected the histological information stated in the article.

Consent
Informed consent of the patient was obtained for publication of this case report

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