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A very rare case of duodenal hemolymphangioma presenting with iron deficiency anemia

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ABSTRACT

INTRODUCTION: Intraabdominal lymphangiomas account for less than 5% of all lymphangiomas and small intestinal hemolymphangioma is a very rare benign tumor.
PRESENTATION OF CASE: Here we describe the first case of primary ulcerated duodenal hemolymphangioma in a 24-year-old woman, causing occult bleeding from gastrointestinal tract. She presented with an unexplained refractory iron-deficiency anemia and gastroduodenoscopy revealed an ulcerated and polypoid lesion of the second portion of the duodenum. Partial resection of the duodenum was thus performed and the final pathological diagnosis was hemolymphangioma.
DISCUSSION: There were only two reports, one of a hemolymphangioma of the pancreas invading to the duodenum and another of a small intestinal hemolymphangioma, presenting with gastrointestinal bleeding until May 2012.
CONCLUSION: The aim of this case report is to highlight the difficulty in making an accurate preoperative diagnosis and describe the surgical management of an unusual location for a very rare tumor. To arrive at a definitive diagnosis and exclude malignancy, partial resection of the duodenum was considered to be the required treatment.

1. Introduction

Hemolymphangiomas are rare benign tumors, characterized by the presence of dilated lymphatic spaces, extravasation of red blood cells, hemosiderin deposition and fibrosis. These malformations are either congenital or acquired and may be explained as a consequence of an obstruction of venolymphatic communication or dysembrioplastic vascular tissue.[1]

Intraabdominal lymphangiomas account for less than 5% of all lymphangiomas.[2] The most common location is the mesentery, followed by the omentum, mesocolon and retroperitoneum.[2–4] Although approximately 60% of these patients are younger than 5 years, a significant number of abdominal lymphangiomas do not manifest until adulthood.[3] Regardless of the diagnostic modalities used, the preoperative diagnosis is very difficult and for most of them a definite diagnosis is made from pathological examination of the resected specimens.

Herein, we report an unusual and unexpected case of ulcerated hemolymphangioma along the medial wall of the second portion of the duodenum, at the papilla of Vater region, in a young woman presenting with severe anemia.

2. Presentation of case

A 24-year-old South American woman was admitted to our hospital because of severe and undetermined anemia (hemoglobin, 6.8 g/dl) in August 2012. No abnormalities were revealed except for iron deficiency anemia in laboratory data. Tumor markers (Alpha fetoprotein, CEA, CA 19.9, CA 125, CA 15.3) were also negative. Gastroduodenoscopy revealed a bleeding, ulcerated, polypoid mass located at the papilla of Vater region, suspected for ampulloma (Fig. 1A and B). Histology from endoscopic biopsies did not provide a conclusive diagnosis but revealed only fragments of duodenal mucosa showing a chronic inflammation pattern with marked lymphangiectasia and mild dysplasia of glandular component. Patient underwent repeat endoscopic biopsies but without additional information.

Magnetic resonance demonstrated a solid, polypoid mass (40 mm × 15 mm) at the lateral wall of the second/third portion of the duodenum with mild contrast enhancement, with no evidence of ampullary obstruction or periduodenal tissue infiltration (Fig. 2A and B).

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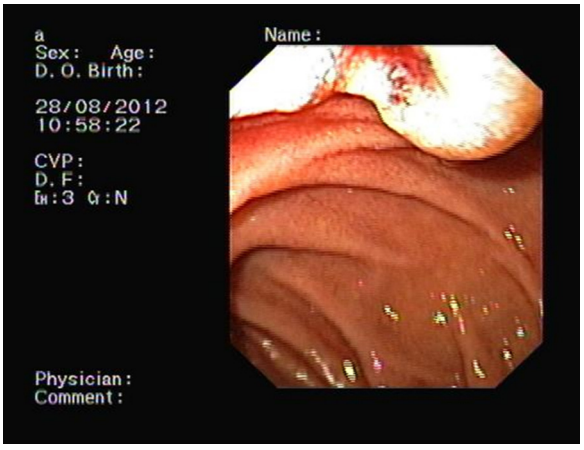

Fig. 1. Gastroduodenoscopy revealing a bleeding, ulcerated polypoid mass in the second portion of the duodenum and precisely at the papilla of Vater region. (Kindly granted by Dr. Raffaele Bennato, Gastroenterology Unit, "Antonio Cardarelli" Hospital, Naples, Italy.)

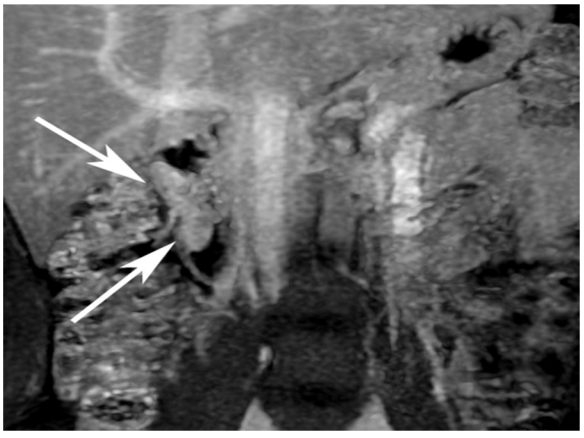

Fig. 2. Cholangio-MRI in coronal plane on T2-weighted images showing a solid, polypoid mass (40 mm × 15 mm) at the lateral wall of the second/third portion of the duodenum with no evidence of ampullary obstruction or periduodenal tissue infiltration. (Kindly granted by Dr. Francesco Di Pietto, Department of Radiology, "Antonio Cardarelli" Hospital, Naples, Italy).

She was then transferred to our General Surgery Unit in September 2012. The patient underwent a wide local resection of the tumor, after complete mobilization of the duodenum by Kocher's manouevre and through a longitudinal incision of the anterior wall of the descending duodenum, about 4–5 cm long, with preservation of the ampulla (Fig. 3A and B). Macroscopically, the lesion was about 5 cm diameter soft, polypoid mass with irregular lobulated margin and ulcerated surface, extending from lateral to medial wall of the second portion of the duodenum, around the medial and inferior border of the ampulla of Vater, but without involving it. A local wide excision of the tumor was considered as a good alternative to duodenocefalopancreasectomy as the location and size of the lesion and the intraoperative extemporaneous examination excluded malignancy. A plastic biliary stent above the ampulla was placed to avoid postoperative ampullary obstruction. The duodenotomy was closed transversely using a two-layer, hand-sewn technique. Final pathological analysis revealed a benign soft tissue mass consisting of lymphatic and blood vessels and diagnosis of this tumor was a hemolymphangioma of the duodenum (Fig. 4A and B). Her postoperative course was characterized by a transient and moderate elevation of cholestatic liver enzymes (GGT, ALP) and she was discharged 17 days after surgery. On January 2013, after about four months, an endoscopic retrograde cholangiopancreatography was performed, and the stent was removed. At the last follow-up visit, no recurrence was observed, and the patient is currently enjoying normal life.

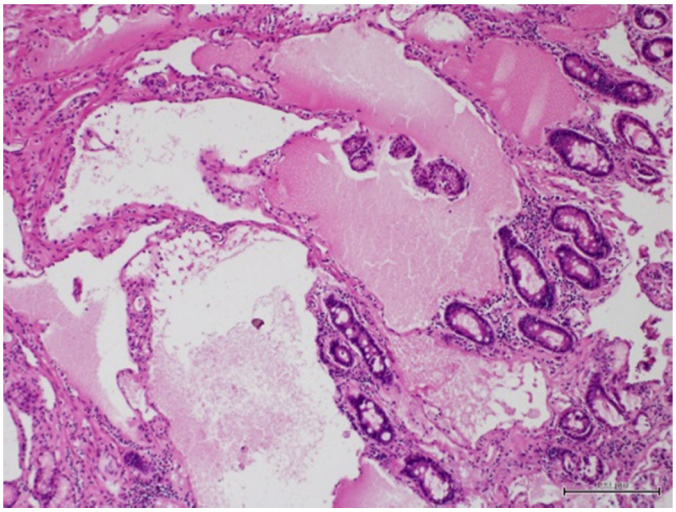

Fig. 4. Histopathology showing thin walled vascular channels lined by flattened endothelium containing acellular eosinophilic amorphous material. Hematoxylin and eosin stain, 250×. (Kindly granted by Dr. Amedeo Boscaino and Dr. Giovanna Carrillo, Department of Anatomic Pathology, "Antonio Cardarelli" Hospital, Naples, Italy.)

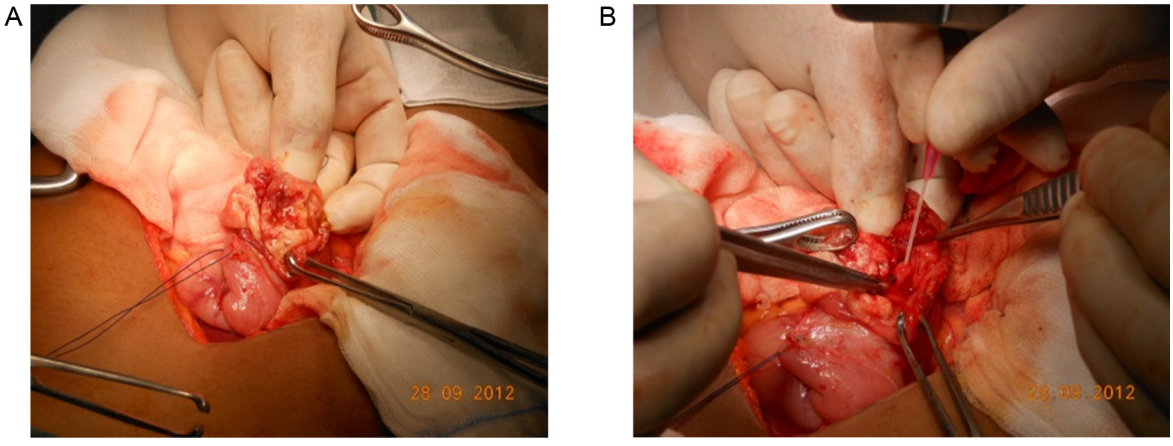

Fig. 3. Wide local resection of the tumor via duodenotomy (A), with preservation of the ampulla (B).