A Rare Presentation of Ménétrier's Disease as Gastroduodenal Intussusception

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ABSTRACT

Ménétrier's disease is a rare cause of hypertrophic gastropathy that is usually confined to the gastric body and fundus. It is characterized by giant rugae, hypoalbuminemia, and foveolar hyperplasia. Here we report the case of a 26-year-old woman who presented with epigastric pain, postparandial nausea-vomiting, and weight loss. Paraclinic evaluation revealed hypoalbuminemia and hypochromic microcytic anemia. Gastroscopy and barium meal study showed diffuse polypoid, nodular lesions that affected the entire stomach, invaginating into the duodenum, leading to partial duodenal obstruction. The histologic, radiologic and endoscopic findings fulfilled the diagnosis of Ménétrier's disease. To the best of our knowledge, gastroduodenal intussusception by Ménétrier's disease has been rarely described in the literature.

KEYWORDS
Menetrier's disease; Gastroduodenal; Intussusception

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INTRODUCTION

Ménétrier's disease is characterized by hypertrophic gastric rugae,[1] gastrointestinal upset,[2] gastropathy resulting in protein loss,[3] normal or hyposecretion of gastric acid,[4] and occasional severe upper gastrointestinal hemorrhage.[5-7] Duodenal obstruction is not a common presentation of this rare disorder. Here, we report a case of Ménétrier's disease complicated by gastroduodenal intussusception.

CASE REPORT

The patient was a 26-year-old female with a chief complaint of epigastric pain since one year previous. Anorexia, heartburn, sensation of postparandial fullness, nausea-vomiting, early satiety and a 10 kg weight loss were among the other signs and symptoms experienced over the previous year. She was prescribed omeprazol and clidinium C with no significant improvement. The patient was admitted to the hospital for further evaluation of worsening symptoms. On physical examination, the vital signs were stable, with no abnormal findings observed in the lungs, heart and extremities. The epigastric area was tender without guarding or detection of a mass. During her hospital

course, the patient had two episodes of melena. Laboratory data were remarkable for the following results (Table 1).

Table 1: Laboratory findings in a case with Ménétrier's disease.

Hematology Tests	Patient	Normal Range
WBC	7700/µL	4000-11000
RBC	4710000/µL	4.5-5.1x10^6
Hb	9.9 g/dL	12.3-15.3
Hct	35.1%	35.9-44.6
MCV	74.5 fL	80-96
MCH	21 pg	27.5-33.2
MCHC	28.2 g/dL	33.4-35.5
Plt	221000/µL	150000-450000
Biochemistry Tests		
BS	125 mg/dL	<140
Cr	1 mg/dL	0.5-1.6
Beta-hCG	0 mIU/mL	
Ferritin	23 ng/mL	9-135
Iron	46 micg/dL	35-160
TIBC	287 micg/dL	0.5-2.1
T4	9.1 micg/dL	4.4-11
T3	1.1 ng/mL	0.5-2.1
TSH	1.4 Mu/L	0.39-5.95
AST	30 IU/L	5-40
ALT	25 IU/L	5-40
Alk-p	220 IU/L	100-290
Total bilirubin	0.5 mg/dL	1-1.2
Albumin	2.9 gr/dL	3.8-5
Total protein	5.8 gr/dL	6-8
PT	13 second	10-13
INR	1	
PTT	35 second	25-35
LDH	504 IU/L	225-450
Amylase	100 U/L	35-260
Stool exam		
Stool OB	Positive	Negative

Alk-p,Alkaline Phosphatase; ALT, Alanine Aminoteransferase; AST, Asparate Aminoteransferase; BS, Blood Sugar; Cr, Creatinine; Hb, Hemoglobin; Hct, Hematocrit; INR, International Normalized Ratio; LDH, Lactate Dehydrogenase; MCH, Mean Corpuscular Hemoglobin; MCHC, Mean Corpuscular Hemoglobin Concentration; MCV, Mean Corpuscular Volume; Plt, Platelet; PT, Prothrombin Time; PTT, Partial Thromboplastin Time; RBC, Red Blood Cell; Stool OB, Stool Occult Blood; TIBC, Total Iron Binding Capacity.

The patient underwent an endoscopic evaluation. The results indicated a large friable mass with tumoral features in the body, antrum, and prepyloric areas of the stomach which was invaginated into the bulb and the second portion of the duodenum (Figure 1). The endoscopist recommended a barium meal and/or abdominal contrast CT scan. The barium study was remarkable for enlarged gastric folds and varying sizes of multiple filling defects in the distal stomach that continued into the whole parts of the duodenum (Figure 2). In order to relieve the obstruction, surgical interventions were planned. During the surgery, the surgeon observed large polypoid lesions in the stomach that lead to gastroduodenal intussusception. A distal partial gastrectomy and intussusception release was performed.

The gross appearance of stomach wall resembled cerebral convolutions and there was diffuse involvement of the stomach (Figure 3). The major histopathologic findings in the gastric specimen included foveolar cell hyperplasia, cystic gland changes, and glands that penetrated into the muscularis mucosa. There were significantly diminished parietal cells noted on histologic examination (Figures 4 A, B). No evidence of malignancy was found in multiple histologic sections. These in addition to other clinical and paraclinical findings were all suggestive for diffuse type Ménétrier's disease.

DISCUSSION

Ménétrier's disease, first described by Pierre Ménétrier (1888), is characterized by gastric mucosal hypertrophy that resembles the brain's convolutions.[1,8] Thickening of the rugal folds are mainly due to proliferation and expansion of epithelial components of gastric mucosa.[9] Ménétrier's is a rare disorder affecting 1 in 200000 of the general population.[8] In most patients it presents with epigastric pain and hypoalbuminemia secondary to the loss of albumin into the gastric lumen, in addition to increased loss of enteric protein.[10] Other signs and symptoms of Ménétrier disease include anorexia, asthenia, weight loss, nausea, gastrointestinal bleeding, diarrhea, edema, and vomiting. In the adulthood type, the disease tends to progress over time. The average age at diagnosis is 55 years, and men are affected more

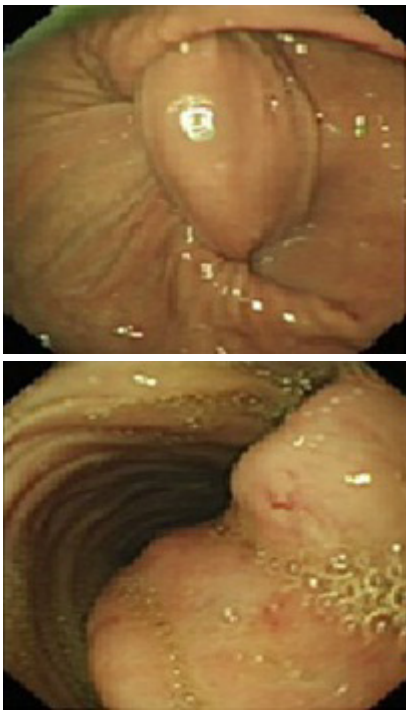

Fig. 1: Upper gastrointestinal endoscopy. A. Gastric area revealed mucosal twisting and invagination at the site of the intussusception. B. Duodenum exhibited a large polypoid mass invaginated in its lumen.

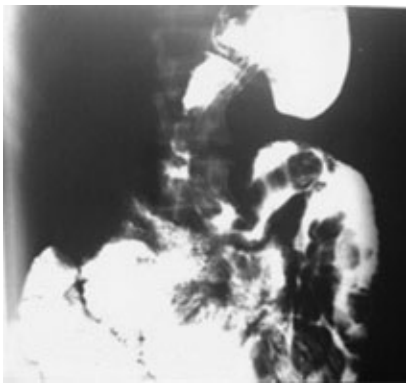

Fig. 2: Upper gastrointestinal barium meal showed gastric thickened mucosal folds along with multiple filling defects in the duodenum.

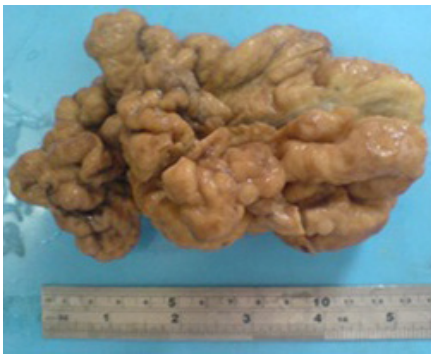

Fig. 3: Gross appearance of antrum that resembled cerebral gyri.

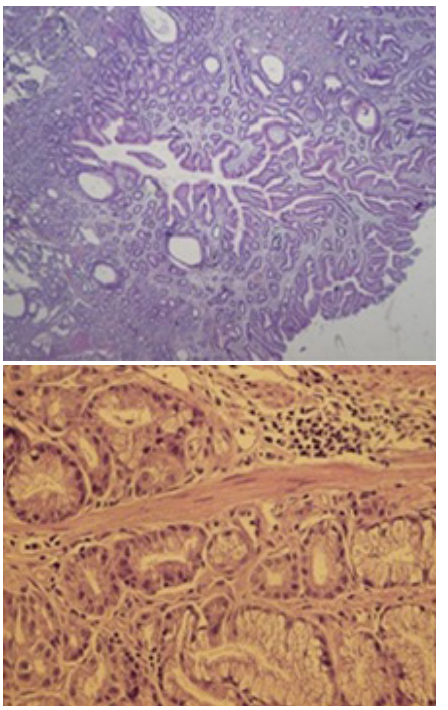

Fig. 4: Microscopic findings of Ménétrier's diease. A_Hyperplastic mucosa with characteristic cystic dilatation of deeper crypts (H&E 40x). B_Glandular penetration to the muscularis mucosa (H&E: 400x).

often than women.[9] In the typical form of the disease there is diffuse involvement of the fundic portion, with sparing of the antrum. In the current case involvement of the antrum has distinguished this disease from conventional Ménétrier's disease that originates from the upper part of the gastric mucosa, body and fundus.[11]

On the other hand, gastroduodenal intussusceptions are a rare clinical entity and comprise the least frequent type of gastrointestinal tract invagination.[12] There have been only two cases of gastroduodenal intussusceptions in the context of Ménétrier's disease so far[13,14] and the current case is the third. Thus, Ménétrier's disease should be considered in the differential diagnosis of upper gastrointestinal obstruction and a possible underlying cause of gastroduodenal intussusceptions.

CONFLICT OF INTEREST

The authors declare no conflict of interest related to this work.

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