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(CASE REPORT)

Menetrier's disease, an uncommon disease and a diagnostic challenge: Case Report

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Abstract

Acquired hypoproteinemic, hypertrophic gastropathy also known as Menetrier's disease is one of the infrequently encountered disorders in clinical practice. Understanding of the pathogenesis, diagnostic features and course of this disease is important as the symptoms may closely resemble a variety of other conditions like malignancies. one of the case control studies, the disease is found to be associated with increased mortality and can predispose to adenocarcinoma

Keywords: Menetrier's disease; Computed tomography; Endoscopy; Multi-disciplinary; Case report

1. Introduction

Menetrier's disease (MD) is a rare disease, which has a prevalence of about 1 in 200,000 people. This was first described by a French pathologist named Pierre Menetrier in 1888. The disease is more commonly observed in adults, particularly in the age range of 30 to 60 years. [1]. The classical symptoms include abdominal pain, vomiting, nausea and peripheral oedema which is secondary to the protein loss seen in this condition. Hyperchlorhydria and normal serum gastrin levels are typically observed in this condition. [2]. The unique feature of this disease is hypertrophy of the gastric mucosa. The gastric mucosa exhibits a convoluted appearance resembling brain convolutions due to thickening of the gastric rugae. [3]. To make an accurate diagnosis, a systematic approach utilizing clinical findings, laboratory tests, imaging, and endoscopic findings is required. The primary challenge for a clinical team is that this disease closely mimics various other conditions, including gastric malignancies, polyposis syndromes and Zollinger Ellison syndrome. Therefore, unnecessary surgical interventions may be performed to establish the diagnosis. Despite MD being a rare disease, a comprehensive understanding of imaging would potentially aid in early diagnosis and treatment.

2. Case Presentation

2.1. History

A 23-year-old female presented to the hospital with severe abdominal pain, persistent vomiting and weight loss. She was found to have bilateral ankle oedema with metabolic acidosis and hypoalbuminemia. She had experienced recurrent episodes in the past two years. On examination, the abdomen was soft with mild tenderness in the upper abdomen. She underwent upper gastrointestinal endoscopy, which demonstrated diffuse thickening of the rugal folds throughout the gastric wall. The mucosa appeared erythematous, edematous with few linear ulcerations and showed abundant mucus. Punch biopsies were taken from the fundus, body and antrum of the stomach. The duodenum appeared normal.

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2.2. Imaging findings

Abdominal MDCT was performed following intravenous infusion of 100 mL of Omni Paque with the following scanning parameters: 5-mm axial collimation, 120 kVp, 92 mAs, and a pitch of 1.5:1. It showed thickened broad rugal folds which were projecting into the lumen of the stomach. This was seen in the fundus, body (involving both lesser and greater curvature) and antrum of the stomach. The maximum thickness of the fold measured 2.4 cm. Furthermore, there was an avid enhancement of the mucosa with homogenous hypo enhancement involving the rest of the layers of the stomach wall. The serosal surface appeared smooth with avid enhancement. The esophagus, duodenum as well as the rest of the small bowel and large bowel appeared normal.

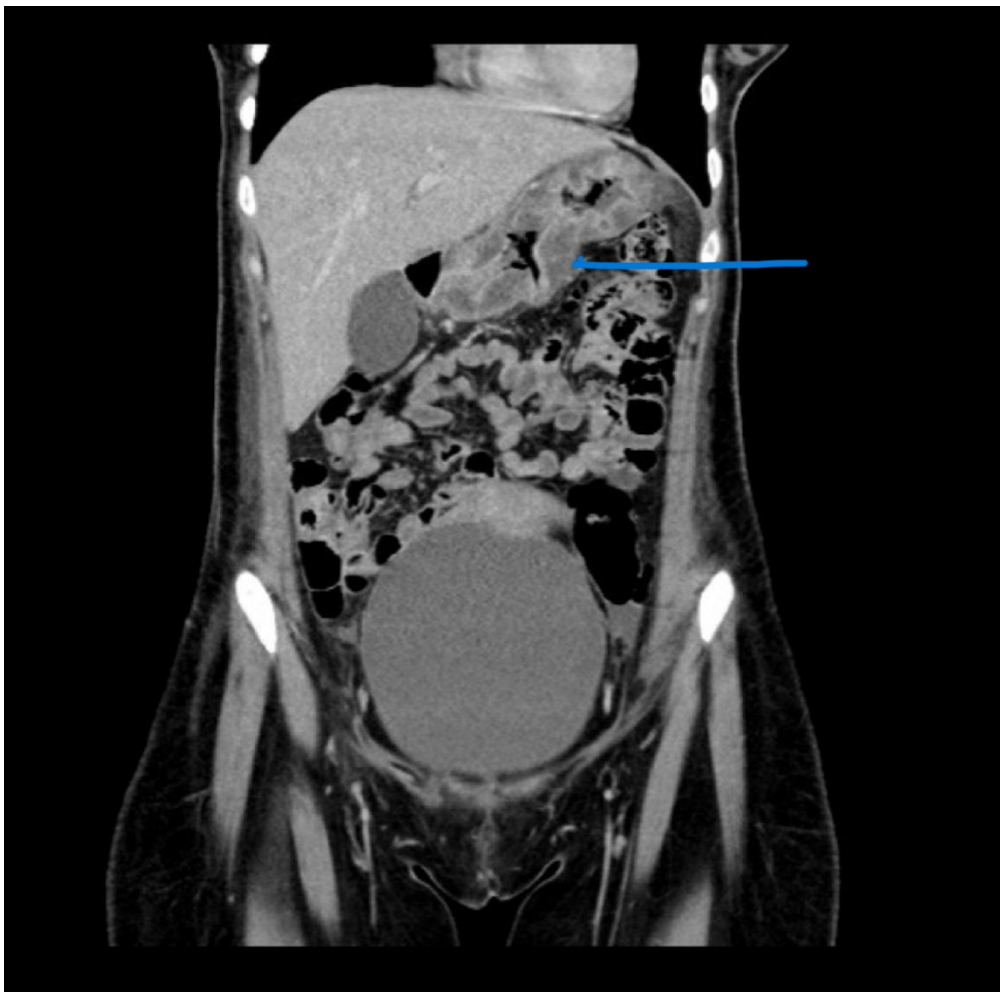

Figure 1 Coronal CT image demonstrates thickened, broad rugal folds projecting into the lumen of the stomach with a convoluted pattern, deep gastric pits, and a three-layered enhancement pattern of the stomach

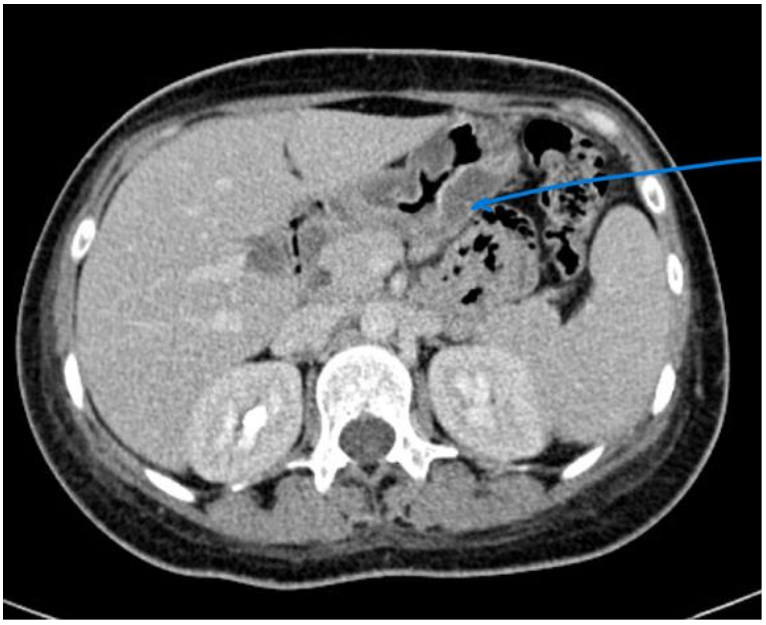

Figure 2 Axial CT image demonstrates thickened, broad rugal folds projecting into the lumen of the stomach with a convoluted pattern, deep gastric pits, and a three-layered enhancement pattern of the stomach

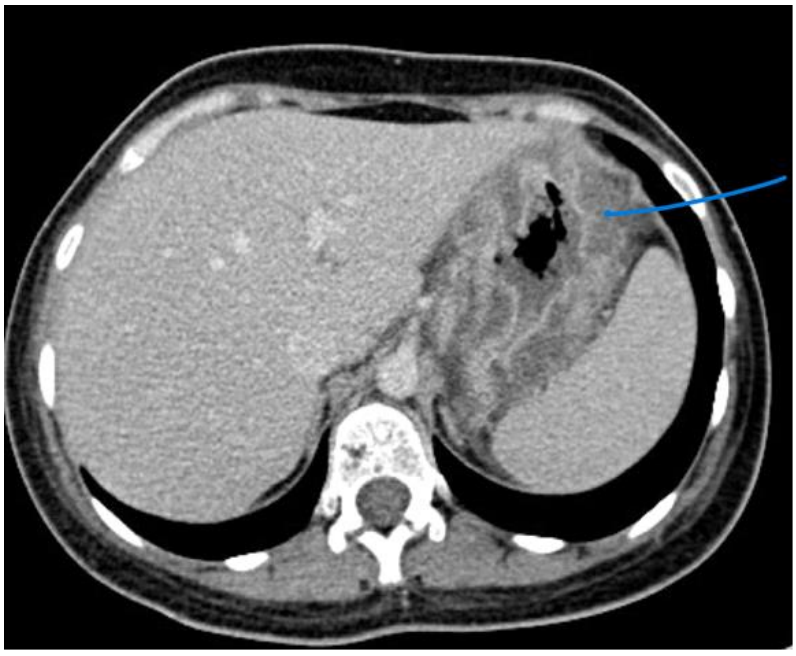

Figure 3 Axial CT image demonstrates thickened, broad rugal folds projecting into the lumen of the stomach with a convoluted pattern, deep gastric pits, and a three-layered enhancement pattern of the stomach

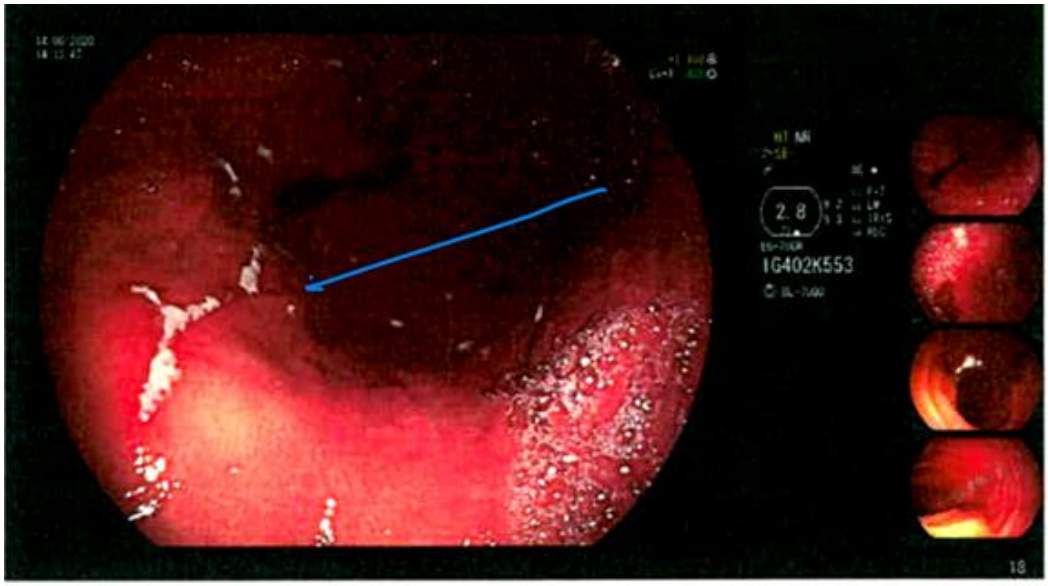

Figure 4 Upper gastrointestinal endoscopy demonstrates severe gastritis like appearance with involvement of most of the greater curvature of the gastric body with multiple linear ulcerations and abnormal vascular pit pattern

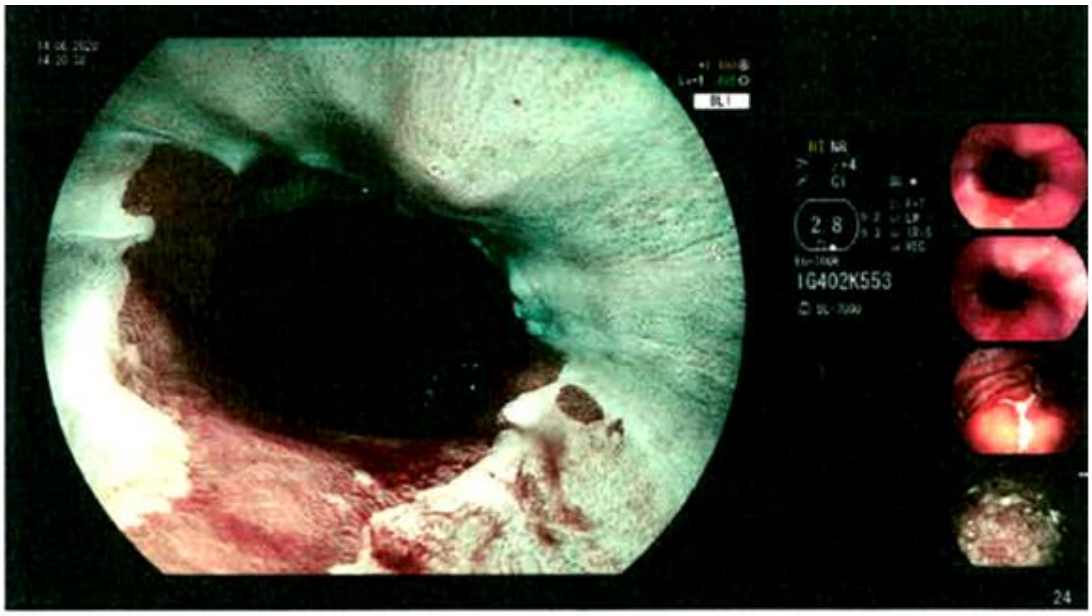

Figure 5 Upper gastrointestinal endoscopy demonstrates severe gastritis like appearance with involvement of most of the greater curvature of the gastric body with multiple linear ulcerations and abnormal vascular pit pattern

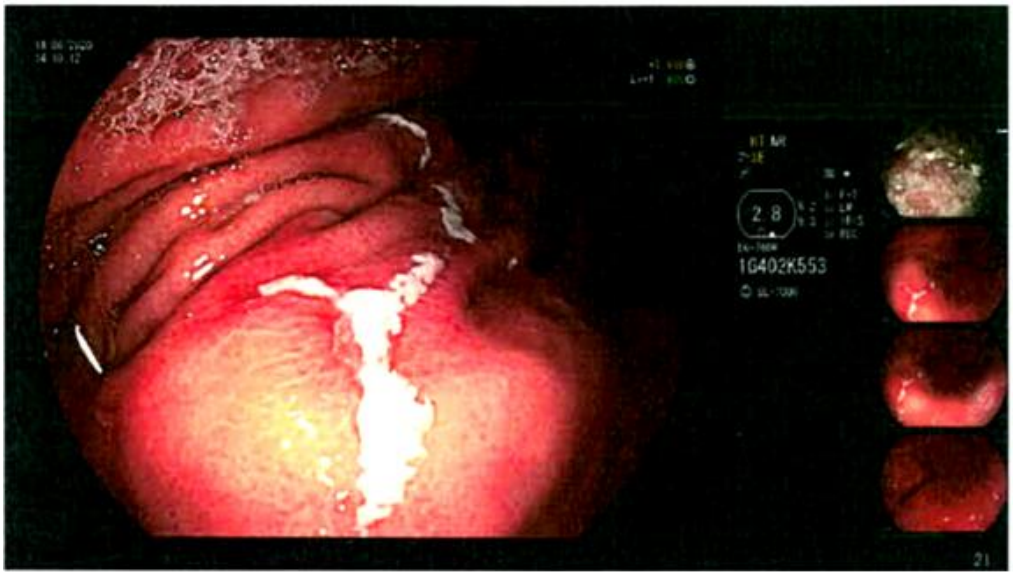

Figure 6 Upper gastrointestinal endoscopy demonstrates severe gastritis like appearance with involvement of most of the greater curvature of the gastric body with multiple linear ulcerations and abnormal vascular pit pattern

2.3. Pathologic evaluation

The punch biopsy specimens obtained from the stomach showed expanded hyperplastic polypoidal mucosal pattern, mild oedema, mild to moderate chronic lymphoplasmacytic and eosinophilic inflammation with elongated cystic dilated glands and decreased oxyntic glands. No Helicobacter pylori or Cytomegalovirus organism, intestinal metaplasia, dysplasia, or malignancy identified.

2.4. Therapeutic intervention and outcome

Patient was started with anticholinergic drugs and proton pump inhibitors. This was followed by histamine blockers. Patient was put on a high protein diet. Rapid improvement of the patient's condition was noticed, and a satisfactory long-term outcome is expected.

3. Discussion

The diagnosis of Menetrier's disease is made through a combination of clinical findings, upper gastrointestinal endoscopic imaging, CT imaging and histological analysis. The presence of bilateral ankle oedema with hypoalbuminemia supports a protein-losing entity. No renal impairment or liver impairment noted in the patient. The other symptoms such as vomiting, abdominal pain and weight loss are quite non-specific and can be present in various other conditions.

The upper gastrointestinal endoscopic imaging is an excellent diagnostic tool in the diagnosis of Menetrier's disease. The presence of gastric fold thickening is a hallmark for this condition. The degree of thickening sets this condition apart from the other differentials [4]. The other differentials for this appearance include infectious causes such as cytomegalovirus and H. pylori infection, infiltrative diseases like sarcoidosis and amyloidosis, and malignancy including lymphoma and gastric carcinoma. Apart from the degree of thickening, the presence of pliability and associated ulcers may help in the endoscopic diagnosis. Gastric neoplasm usually shows folds, which are rigid while MD folds are more pliable. Thin linear ulcers may be seen in Menetrier's disease, but they are not seen in post bulbar region as seen in Zollinger-Ellison syndrome [5]. Lymphoma usually affects the distal stomach and the lesser curvature while Menetrier's disease is more pronounced along the greater curvature. However, in our case, it was seen in both lesser and greater curvatures. Amyloidosis demonstrates a nodular thickening while MD shows a tortuous [6], solid and spongy fold thickening resembling cerebral convolutions [7].

Histologically, increased mucosal thickening caused by hyperplasia of the epithelial cell is an important pathological finding. Besides, deep gastric pits, cystic dilated glands secreting mucin are also seen. There is an overall reduction in the number of parietal and chief cells thereby decreasing the gastric acid secretion [8]. Surface mucous cells stain positive with PAS, MUC5AC, gastrokine and TFF1. Reduction in the number of parietal cells and chief cells can be

visualised with reduced expression of H+/K+ATPase and pepsinogen, respectively, by immunohistochemical staining. Ki67 may show an increase in the number of proliferating cells in the expanded, downwardly displaced progenitor zone, which is the main feature of MD [9].

As mentioned earlier, imaging itself may not help in clinching a diagnosis of Menetrier's disease but it too has its unique features. An endoscopic ultrasound shows a thickened echogenic mucosal layer, while the submucosal and the muscularis propria appear normal [9]. A barium meal may show enlarged and convoluted rugal folds. Poor coating of the folds may be seen due to hypersecretion, and this may decrease the effectiveness of the study.

The CT scan with intravenous contrast shows multiple folds which are more than 1 cm in thickness. This also shows convoluted thickened folds with the normal-appearing wall in between the folds [10]. However, the serosal contour is smooth throughout the stomach. One of the important CT features seen is the presence of at least three layers of differential enhancement of the thickened fold. The more superficial fold which is close to the lumen shows hyper-enhancement and this may be related to foveolar hyperplasia with intraepithelial lymphocytes and other inflammatory cells [11]. However, the second layer which is the thickest layer on CT shows diffuse homogenous hypo-enhancement. Edematous lamina propria with hyperplasia is a feature of MD and this may correspond to the second layer seen in CT. The third layer, which is the serosa, demonstrates avid enhancement in the contrast study. This three-layered pattern within the convoluted thickened folds is not commonly observed in other conditions such as lymphoma or gastric adenocarcinoma.

A close mimicker of MD when it comes to CT imaging is the various forms of polyps and polyposis syndromes. Clinically, these entities can be ruled out to some extent as peripheral oedema, absence of anemia and presence of vomiting are more common in the MD. Likewise, low serum albumin, high gastric pH and normal serum gastrin values also favor patients with MD.

One of the main challenges seen while performing a CT scan of a patient with MD is the absence of a well-distended stomach. A poorly distended stomach lumen may mimic thickened fold, and this can be prevented by giving oral contrast to distend the stomach. A neutral oral contrast has an added advantage as it will highlight the three-layered pattern of the stomach wall.

Treatment for this condition can be challenging. It includes anticholinergic drugs, proton pump inhibitors, prednisone, and histamine blockers and a high-protein diet. However, all these showed variable results [11]. In severe cases, gastrectomy may be performed. In addition, monoclonal antibodies are found to be effective in some case studies [12]. Considering the increased rates of mortality and association with adenocarcinoma, it is advisable to follow up patient with surveillance endoscopy [13]

4. Conclusion

Although MD can be accurately diagnosed by histopathology and upper gastroscopy, accurate clinical evaluation, laboratory findings and a well performed CT scan are essential for an early diagnosis of a patient with MD. Thickened stomach folds of more than 1cm with a convoluted pattern in a distended stomach, deep gastric pits and a three-layered enhancement pattern of the stomach in CT scan should raise a high suspicion of MD. This would further guide a gastroenterologist in planning for an upper gastroscopy and a full-thickness mucosal snare/punch biopsy. A multidisciplinary approach with a radiologist, gastroenterologist, anesthesiologist, and pathologist is important.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no conflicts of interest.

Statement of informed consent

Written informed consent obtained from the patient for publication.

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