

A Case Report of Menetrier's Disease

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ABSTRACT

Menetrier's disease (MD) is a rare premalignant disorder of unknown etiology, characterized by giant hypertrophic folds involving fundus, with antral sparing. Microscopy shows hyperplasia and corkscrew appearance of gastric foveolae, often associated with prominent eosinophilic inflammatory infiltrate. Common presentation is pain, vomiting, diarrhea and hypoproteinemia. Differential diagnoses include polyposis and infiltrating disorders, and gastrectomy recommended if debilitating disease or risk of carcinoma. A 50-year-old male presented with abdominal pain, vomiting and hypoproteinemia. Radiology and endoscopy showed thickened gastric folds and biopsy suggested a possibility of eosinophilic gastritis or MD. Total gastrectomy specimen and microscopy showed a classical appearance of MD. Exact incidence of MD in India is not known. Diagnosis can be missed on biopsy and adequate sampling with follow-up is essential due to associated risk of malignancy.

Keywords: Menetrier Disease, Corkscrew, Foveolar Hyperplasia, Hypertrophied Gastric Folds

Introduction

Menetrier's disease (MD) was first described in 1888 by French pathologist Pierre Menetrier.^[1] It is an acquired premalignant disorder of unknown etiology, characterized by giant hypertrophic rugal folds that involve the fundus but often spare the antrum.^[2] Most characteristic features of MD on histology are foveolar hyperplasia, elongation and corkscrew appearance. It is listed by the Office of rare disease, National institutes of Health (USA), indicating prevalence of less than 1 in 200000 people.^[3] We report a case of MD which was initially mistaken for eosinophilic gastritis on biopsy, but was later diagnosed correctly on gastrectomy.

Case Report

A 50-year-old male presented in the out-patient department of our hospital with a history of dull aching, persistent, non-radiating pain in the epigastric region for 2 years which was associated with intermittent nausea/vomiting since 1 ½ months. On investigations, he was found to have hypoproteinemia and anaemia. USG abdomen showed diffuse gastric wall thickening. Upper GI endoscopy showed thickened gastric folds extending from gastroesophageal junction to the fundus and body with antral sparing (Fig1a). A possibility of MD or infiltrative disorders was suggested clinically, for which gastric biopsy was done. Histopathology showed elongated tortuous gastric foveolae and a prominent eosinophilic inflammatory infiltrate in lamina propria, thus a differential diagnosis of eosinophilic gastritis or Menetrier disease was considered. Meanwhile, contrast enhancing CT scan was

done which showed hypertrophied rugal folds, diffusely involving the stomach without any muscle layer thickening (Fig1b), suggesting a possibility of chronic inflammatory process or low grade neoplastic etiology. Reviewed CT suggested a possibility of Menetrier's disease.

A total gastrectomy was performed which showed markedly enlarged and dilated stomach, measuring 30 cm in length. Serosa was normal, antrum was unremarkable and mucosa at rest of the places showed marked hypertrophy of mucosal folds (Fig1c), associated with gastric wall thickening (inset). Histology showed elongated, enlarged gastric foveolae (Fig: 2a) with a classical corkscrew appearance (Fig: 2b), hypoplastic glands, few gland abscesses (Fig: 2c) and a mixed inflammatory infiltrate in lamina propria, rich in eosinophils (Fig: 2d). Thus a diagnosis of MD was made. H. Pylori organisms were absent both on H&E and special stains.

Discussion

MD is an idiopathic, hypertrophic gastropathy characterised by hyperplasia of superficial mucosal epithelium. Clinically patient can have epigastric pain, bleeding due to erosions, diarrhea due to excess mucus secretion with hypoalbuminemia and non-dependent edema along with signs of chronic obstruction notably nausea, vomiting, anorexia and weight loss. This can be associated with significant morbidity, and mortality as well due to an increased risk of adenocarcinoma.^[1-3] Anaemia is unusual in MD unless there is hematemesis.^[2,4] Our patient did not have any bleeding; hence anaemia in him was likely to be nutritional which is quite common in our set up, being a public hospital which primarily caters to the poor patients.

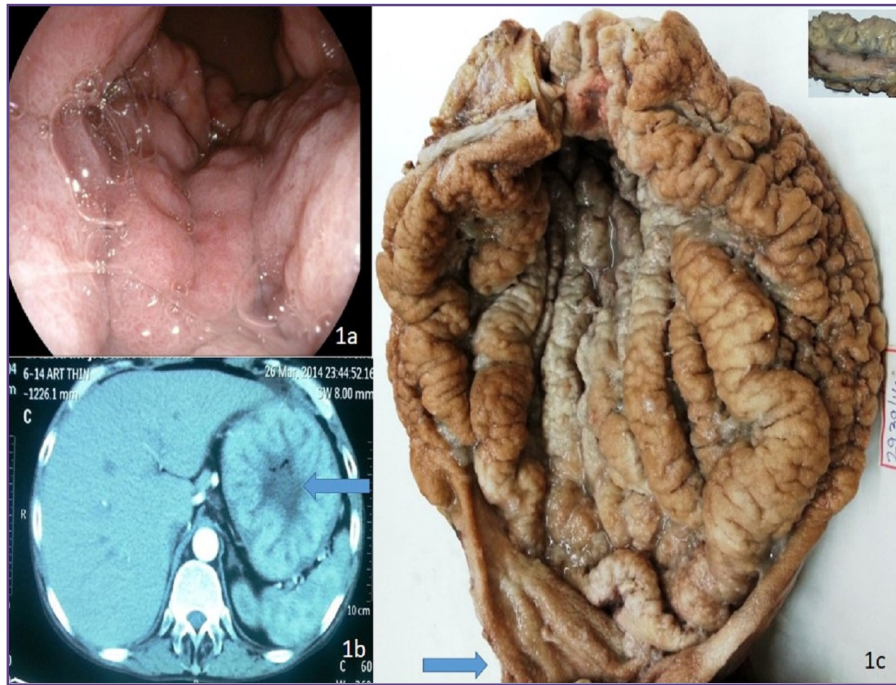


Fig. 1: a) Endoscopy showing thickened rugal folds from GE junction to fundus and body, sparing the antrum; b) Contrast enhancing CT scan showing Stomach with hypertrophied rugal folds resembling sulci and gyri of cerebrum (arrow); c) Enlarged stomach showing markedly hypertrophied mucosal folds with thickened wall (5-6 cm) (inset) and antral sparing (arrow).

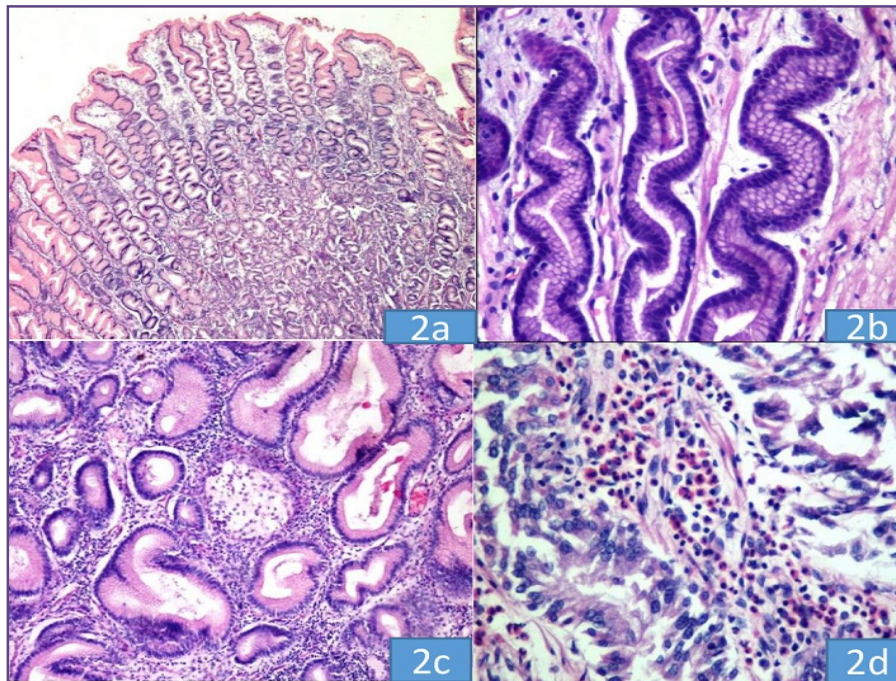


Fig. 2: a) Thickened gastric mucosa showing foveolar hyperplasia (H&E, 40 X) b) Elongated foveolae with characteristic “corkscrew appearance” (H&E, 400 X) c) Cystically dilated glands (H&E, 100X) d) prominent eosinophils in the lamina propria. (H&E, 400X) .

The aetiology of Menetrier’s disease is unknown. Various possible causes leading to mucosal hypertrophy in stomach can be bacterial infections, toxins; neurogenic, emotional,

congenital, endocrine and mechanical causes. The mucosal changes in Menetrier disease are hypothesized to be secondary to an overproduction of transforming growth factor alpha.

MD occurs in two forms, a childhood form and the adult form. In paediatric form of the disease, association of the allergy is common along with the CMV infection which resolves spontaneously, but should be distinguished from CMV gastritis. Adult form occurs commonly between 4th-6th decade with a male predilection and is commonly associated with over expression of TGF-alpha and Helicobacter Pylori infection.^[3] Our patient was an adult male; however, his stomach mucosa was negative for H. Pylori.

Gross of MD is quite classical, mostly shows antral sparing similar to our case, which however may not be always present.^[5,6] Two distinct histological forms of MD have been reported in the literature, one is hypertrophic lymphocytic gastritis (HLG) which is characterised by a large number of intraepithelial lymphocytes associated with marked inflammation in the lamina propria, which was not seen in our case. Another form i.e. massive foveolar hyperplasia (MFH) which can show prominent eosinophils similar to our case and sometime prominent smooth muscle fibres in lamina propria, which were not seen in our case.^[6]

The differential diagnosis of Menetrier disease is broad and includes hyperplastic polyps/ polyposis or infiltrative diseases such as sarcoidosis, amyloidosis; Zollinger Ellison syndrome (ZES); and malignancies notably lymphoma and gastric carcinoma. However, features like localized rather than diffuse involvement, associated with loss of architecture and parallelism of the glandular units in polyps, decrease fold pliability on scopy in infiltrative diseases, parietal cell hyperplasia rather than gland hypoplasia or atrophy in ZES and specific histologic features like characteristic eosinophilic material in amyloid may help in making the correct diagnosis.^[3] Often a prominent eosinophilic inflammation in lamina propria can be seen in many patient, which can mimic eosinophilic gastritis especially on biopsy as happened in our case.^[6]

Treatment for MD includes high protein diet, albumins, plasma, diuretics, anticholinergics, proton pump inhibitors and monoclonal antibodies to EGFR. Gastrectomy is reserved for the patients in whom pharmacologic therapy fails or who have bleeding, malignancy, protein loss or crippling clinical features as were present in our case and also to make correct diagnosis too as needed in our case.^[1, 3, 7]

Conclusion

MD is a well-known entity but rare. Exact incidence in India is not known but few case reports are available in association with trichobezoar and primary pachydermoperiostosis.^[8, 9] An adequate sampling with proper follow up is essential due to associated increased risk of malignancy, however there are contradictory reports available about the same.^[3] Diagnosis can be missed on biopsy but characteristic gross and microscopy, in association with classical endoscopic findings and radiology helps arriving at correct diagnosis.

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Reference

1. Azer M, Sultan A, Zalata K, Abd El-Haleem I, Hassan A, El-Ebeidy G. A case of Menetrier's disease without Helicobacter pylori or hypoalbuminemia. *Int J Surg Case Rep.* 2015;17:58-60.
2. Coffey RJ, Washington MK, Corless CL, Heinrich MC. Ménétrier disease and gastrointestinal stromal tumors: hyperproliferative disorders of the stomach. *J Clin Invest.* 2007;117(1):70-80.
3. Friedman J, Platnick J, Farruggia S, Khilko N, Mody K, Tyshkov M. Menetrier Disease. *Radiographics.* 2009;29:297-301.
4. Stamp GW, Palmer K, Misiewicz JJ. Antral hypertrophic gastritis: a rare cause of iron deficiency. *J Clin Pathol.* 1985;38(4):390-392.
5. Burdick J.S, Chung E, Tanner G, Sun M, Paciga JE, Cheng JQ. Treatment of Menetrier's Disease with a Monoclonal Antibody against the Epidermal Growth Factor Receptor. *N Engl J Med.* 2000; 343:1697-1701.
6. Sweeney AR, Lynch MK. A case of Ménétrier's disease localized to the gastric antrum without helicobacter infection or hypoalbuminemia. *Int J Surg Case Rep.* 2013;4(10):839-841.
7. Rich A, Toro TZ, Tanksley J, Fiske WH, Lind CD, Ayers GD et al. Distinguishing Ménétrier's disease from its mimics. *Gut.* 2010;59:1617-1624.
8. Lakshmi TS, Rao PN, Nagaria M. Primary pachydermoperiostosis associated with Menetrier's disease. *Indian J Dermatol Venereol Leprol.* 2001;67(5):256-8.
9. Chakrabarti AK, Singh TD, Joshi K, Malik AK. Ménétrier's disease and trichobezoar of stomach-an unusual association. *Postgrad Med J.* 1983;59(693):464-466.

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