Ménétrier's Disease (MD) was first described in 1888 by Pierre Ménétrier.1 It is an uncommon acquired disorder of unknown etiology characterized by giant hypertrophy of the rugal folds. Patients present with a unique constellation of signs and symptoms that include abdominal pain, nausea and vomiting, hypochlorhydria (due to markedly reduced, if not absent, number of parietal cells), and edema of peripheral tissues (due to leakage of protein across the gastric mucosa). They may also develop anemia.2 It is more common in males (male to female ratio=3:1), between fourth and sixth decades of life.

Herein, we report a case, for the first time in our country, of a 30-year-old woman presenting with the main complaint of peripheral edema and diagnosed as MD on endoscopic biopsy.

CASE REPORT

A 30-year-old woman with no known co-morbid condition presented to the out patient department (OPD) with the main complaint of gradual swelling of both legs extending to the knees. On further questioning, she gave history of dyspepsia and vague abdominal pain. She described no abdominal distention, heartburn, dysphagia, melena, hematemesis, diarrhea, fever, night sweats, or other cardiopulmonary or urinary symptoms. Her past medical history was unremarkable. She was not using any medications and denied drug addictions. She smoked Huqqah and had non-significant family history. Family history was negative for gastrointestinal disease. Physical examination was unremarkable except for bilateral pitting edema upto the knees. It was neither warm nor red and was non-tender.

Her laboratory investigations showed complete blood count with hemoglobin (Hb) of 10.7 g/dl, Mean Corpuscle Volume (MCV) of 84.6 fl and platelets at 464 x 10³ per microliter. Markedly decreased serum albumin of 1.4 g/dl and increased serum globulin of 6.8 g/dl was found. The liver function tests and serum chemistry were within normal limits. Urine analysis showed no proteinuria. Ultrasound abdomen was reported normal.

Esophagoduodenoscopy (EGD) demonstrated mosaic pattern of gastric mucosa with prominent rugal folds. Biopsy of gastric body was obtained which showed marked foveolar hyperplasia with elongated, dilated and tortuous gastric pits. No evidence of Helicobacter (H.) pylori or malignancy was observed. Histopathological findings were consistent with MD.

Patient was started on Proton Pump Inhibitors (PPIs) and advised for follow-up. However, the patient was lost to follow-up.

DISCUSSION

MD was first described by Ménétrier in 1888.1 It represents the diffuse giant mucosal growth of stomach lining in the shape of wide, long, twisted cerebral-like folds. This morphological change is attended by the loss of proteins from the stomach and consequential hypoproteinemia (protein losing gastroenteropathy). In adults, the disease has a progressive course with...
Cytomegalovirus (CMV) infection in children.1,8,9 In this patient, gastric biopsy did not reveal concomitant H. pylori or CMV infection. Pathogenesis of the disorder is also not completely clear. There is an evidence that Epidermal Growth Factor Receptor (EGFR) signaling is involved in the pathogenesis of the disease.1

EGD shows massively thickened gastric rugal folds, resembling cerebral convolutions, along the greater curvature in body and fundus, usually sparing the antrum with copious amounts of thick mucus.3 Markedly hypertrophic and enlarged folds are subject to erosions. Most often gastric pH is alkaline.

Histologically, MD shows striking foveolar hyperplasia with massive expansion of surface mucous cells associated frequently with reduced number of parietal cells and chief cells. Gastric pits are often tortuous and undergo cystic dilatation. Modest inflammation may be noted in the lamina propria. The muscularis mucosa is usually thickened, with strands of smooth muscle extending into the lamina propria. Deep snare biopsies or large capacity biopsies are essential for histological evaluation of suspected MD.5

Gastric body biopsy in this patient revealed foveolar hyperplasia with elongated, dilated and tortuous gastric pits and atrophy of the oxyntic glands. The morphological findings were consistent with MD. No H. pylori or other infective organism was found.

Gastric mucosal thickening with prolonged PPIs use can clinically imitate MD. But it can be excluded on the basis of abundance of parietal cells in the biopsy. This patient denied any use of PPIs and excess of parietal cells was not documented on her gastric biopsy.

MD is also considered a premalignant disorder. However, the magnitude of risk of malignancy in MD is uncertain.

There are no evidence-based guidelines available for treatment strategy of MD. It usually has abrupt onset and spontaneous resolution in children while an insidious onset and progressive course characterizes the disease in adults.8 Spontaneous remissions are rare except in CMV-associated disease in children.1 Empirical treatment with anticholinergic therapy, octreotide, H2-receptor blockers, glucocorticoids, antifibrinolytic agents, or monoclonal antibody against EGFR, as well as eradication of H. pylori, has not provided constant benefit. Surgical intervention has been reserved for patients with biopsy proven malignancy or dysplasia, high amount of protein loss and uncontrolled and/or recurrent bleeding. We were not able to document the response of this patient to PPI use, as she was lost to follow-up. This is one of the major drawbacks of our health care system and the society.

In summary, this case highlights the variation in the presentation of the disease. A high index of suspicion along with scrupulous and diligent endoscopic and histological evaluation will help in an accurate diagnosis of the condition.
Ménétrier's disease presenting with edema

REFERENCES


