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A case of Menetrier's disease without *Helicobacter pylori* or hypoalbuminemia

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ABSTRACT

INTRODUCTION: Menetrier's disease is a rare premalignant hypertrophic gastropathy. It is characterized by huge gastric mucosal folds, peripheral edema due to protein loss and upper GIT symptoms such as epigastric pain, discomfort, nausea and vomiting.
PRESENTATION OF CASE: Female patient 35 years old complaining of severe epigastric pain, dyspepsia, nausea and vomiting for 1 year. Upper GIT endoscopy and CT scan revealed thickening of gastric mucosa. Endoscopic biopsy was non-specific but showed moderate grade dysplasia and no *Helicobacter pylori* infection. All laboratory investigations were within normal including serum albumin. She underwent total gastrectomy with marked postoperative improvement of symptoms after recovery. Postoperative pathology revealed gastric fovular hyperplasia and glandular atrophy which are diagnostic for Menetrier's disease.
DISCUSSION: the preoperative diagnosis of Menetrier's disease in this case was challenged by its unusual features. There were neither *H. pylori* nor hypoalbuminemia. Literature review showed similarcases which can raise the suspicion of the presence of an undescribed subtype of the disease.
CONCLUSION: Menetrier's disease should be suspected in cases of upper GIT symptoms and hypertrophied gastric mucosa with or without *H. pylori* or hypoalbuminemia. The preoperative diagnosis could not be confirmed unless a whole mucosal thickness biopsy is performed. Surgical management is a good option when medical treatment fails to relieve the symptoms and erase the risk of malignancy.

1. Introduction

Menetrier's disease (MD) is a rare disease that was first described by the French pathologist Pierre Menetrier in 1888 [1]. It is a rare acquired hypertrophic gastropathy leading to dilatation of the mucus-secreting gastric pits (fovula) on the expense of the gastric glands containing the acid-producing parietal cells and pepsinogen-producing chief cells which undergoes atrophy. These changes gives the disease it characteristic features of the huge expansion of gastric mucosa, thick mucus secretion, protein loss and hypochlorhydria [2].

The disease is more dominant in middle-aged males [3] but several authors reported it in pediatric population [4]. The etiology is not well established. Some theories tried to link it to *Helicobacter pylori* (*H. pylori*) infection [5] and enhanced gastric epidermal growth factor receptor signaling by transforming growth factor-alpha [1]. The clinical picture includes epigastric pain and discomfort, nausea, vomiting and peripheral edema due to low serum albumin levels. MD has a recognized premalignant potential [6–8]. Also it can lead to severe uncontrollable protein loss or upper GIT bleeding [8].

Medical treatment of MD includes proton pump inhibitors, high-protein diet, eradication of *H. pylori* [5], cetuximab (monoclonal antibody) [6] and octreotide long-acting release [4]. Also total and partial gastrectomy are attempted in managing resistant cases. We describe a case of MD with an unusual presentation that was recently managed in our hospital.

2. Presentation of case

Female patient 35 years old, married with 2 children was presented to our outpatient clinic complaining of severe epigastric pain, dyspepsia, nausea and vomiting for 1 year. Her condition started with gradual onset and showed progressive course. She went to her primary care giver at the onset of her condition and was diagnosed to have chronic gastritis. She underwent multiple courses of proton pump inhibitors and Prokinetic medications with

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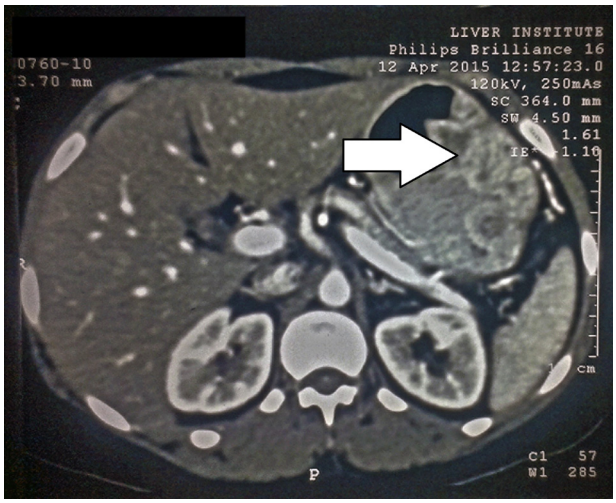

Fig. 1. CT scan of the abdomen with IV contrast revealing marked mucosal thickening (arrow) of the fundus, body and antrum of the stomach.

no significant improvement. She has no relevant family, medical or surgical history.

Upper GIT endoscopy was done. It revealed unhealthy mucosal growth occupying all the fundus and the body of the stomach. Histopathological examination revealed tubulovillous adenoma with moderate grade dysplasia with no evidence of *H. pylori*. Abdominal CT scan (Fig. 1) revealed marked mucosal thickening of the fundus, body and antrum of the stomach with few spared areas in-between. Uniform enhancement after contrast administration and the preserved fat planes suggested benign nature of the mucosal pathology. Laboratory investigations including CBC, liver and kidney functions in addition to tumor markers (CEA–CA 19–9) and Cytomegalovirus (CMV) were all within normal range. It is worthy to state that the serum albumin was 4 g/dL.

On 18th April 2015, the patient underwent total gastrectomy and esophago-jejunal anastomosis after laparoscopic assessment that did not reveal any other intra-abdominal pathology. The patient underwent an uneventful postoperative period except for transient drop in serum albumin level on the third and fourth postoperative days (2.7 and 3.1 g/dl respectively). She was discharged on the 8th postoperative day. 3 months follow up showed marked improvement of her health status with no signs of recurrence of her complaint.

Histopathological examination of the resected specimen revealed diffuse huge rugea of the whole gastric body showing cerebriform appearance (Fig 2). Antral region appeared grossly free. Microscopic examination revealed fovular hyperplasia reaching the muscularis mucosa with evident glandular atrophy confirming the diagnosis of MD (Fig 3). Both proximal and distal cut margins were free.

3. Discussion

The diagnosis of MD remains an everlasting challenge. This is due to the rare nature of the disease and the lack of clear diagnostic criteria [2,8].

We could not find an exact estimate of MD incidence in literature except that is “rare”. In our experience, it is so rare that each new case is usually encountered at first by an endoscopist who did not see a similar case before. This explains why the diagnosis of the huge gastric folds can be mixed with other types of unhealthy growths like gastric polyps or polyposis syndromes [2], Hypertrophic hypersecretory gastropathy with protein loss [9], Zollinger–Ellison syndrome and gastric malignancies [8].

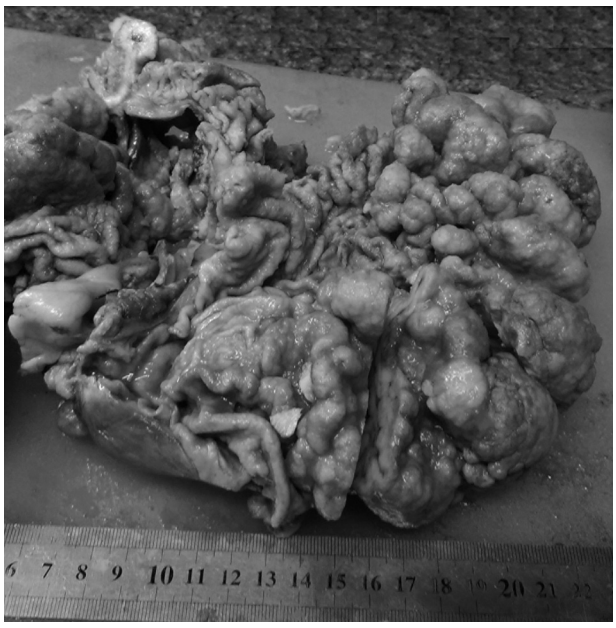

Fig. 2. Gross appearance of the resected stomach after the total gastrectomy revealing huge gastric folds.

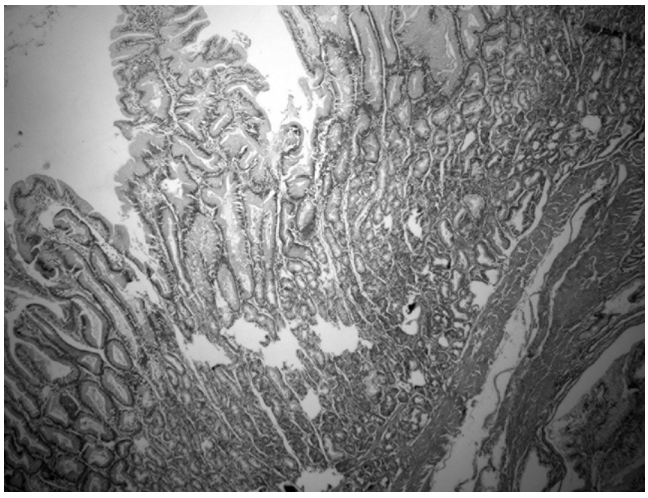

Fig. 3. Microscopic examination of the resected specimen showing the characteristic features of Menetrier’s disease: fovular hyperplasia reaching the muscularis mucosa with evident glandular atrophy.

Rich et al. [2], proposed an algorithm for the diagnosis of MD that rely on clinicohistopathological analysis of the following criteria: upper GIT endoscopy with gastric pH, appropriate laboratory tests (CBC, serum albumin, serum gastrin, *H. pylori* and CMV) and full thickness mucosal biopsy. This is to be correlated with classic clinical features of epigastric pain, nausea and vomiting due to the local effect of the huge gastric folds, in addition to peripheral oedema due to the protein losing nature of the disease that leads to Hypoalbuminemia.

The previous criteria is only useful in dealing with typical cases of MD. In our case, as well as in several cases in literature, the diagnosis was obscure due to the presence of variant clinical presentations.

Although hypoalbuminemia is considered as a corner stone in the diagnosis, our case did not show it on neither symptoms (peripheral edema) nor laboratory values. Her albumin levels remained within normal ranges except for a transient drop in the postoperative period, which we believe to be due to dilution effect

of postoperative fluid therapy. The same feature of MD with normal albumin level could be found in literature in both adult [3] and pediatric [4] population suggesting the presence of a subtype of the disease that should be further studied.

Pinch biopsy in our case could not establish the diagnosis. The histopathological diagnoses of MD needs a deeper snare biopsy (with or without electrocautery) to examine the full thickness of the hypertrophied mucosa for fovular hyperplasia and glandular atrophy [2]. Sanchez et al. [10] reported a case of MD in which diagnosis was made by an invasive laparoscopic-assisted full thickness biopsy after failure of multiple endoscopic pinch trials.

Another distinctive feature of our case is that it was not associated with *H. pylori* infection. Although many authors proposed that *H. pylori* plays an essential role in the pathogenesis process of MD [5], many cases were reported without infection rendering this causal relation unsupported [3,4]

Gastrectomy is a well-established treatment of MD due to the high rate of failure of medical treatment and the risk of malignant transformation. Surgical treatment in our case was justified by the long duration of the disease (1 year), severe symptoms resistant to conservative medical treatment and the presence of dysplasia in the endoscopic biopsy that represented an alarming sign of impending malignancy. Also the extensive nature of the pathology affecting the majority of the stomach encouraged the operators to opt for total gastrectomy. While the antrum appeared grossly free, its thick wall in CT scan and during surgical exploration pushed towards the decision of total gastrectomy. The added benefit of proximal gastrectomy could not balance the risk of performing the esophageal anastomosis with a suspiciously unhealthy antrum [8]. This was supported by reports of MD that affected the antrum as well as the fundus and the body of the stomach [3].

4. Conclusion

MD should be suspected in cases of upper GIT symptoms and hypertrophied gastric mucosa with or without *H. pylori* or hypoalbuminemia. The preoperative diagnosis could not be confirmed unless a whole mucosal thickness biopsy is performed. Surgical management is a good option when medical treatment fails to relieve the symptoms and erase the risk of malignancy.

Conflict of interest

None.

Funding

None.

Ethical approval

The Ethical committee of the Egyptian Liver Research Institute and Hospital revised and approved the ethical aspects and the patient consent of this work.

Authors' contributions

MA, GE and KZ mainly designed the study and drafted the manuscript. KZ participated in providing the images and its analysis. GE, AS, MA, IA and AH participated in the operation. MA, IA and AH carried out literature review. GE and AS revised the manuscript. All authors read and approved the final manuscript.

Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images.

Guarantor

Mina Azer, Ahmad Sultana, Khaled Zalata, Ibrahim Abdel Haleem, Adel Hassan, Gamal El-Ebeidy.

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