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Case report

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MÉNÉETRIER DISEASE - CASE REPORT

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SUMMARY

Ménétrier disease (hyperplastic hypersecretory gastropathy) is uncommon disorder of the stomach characterized by enlarged folds. It is often accompanied by a severe loss of plasma proteins (including albumin) from the altered gastric mucosa. The disease occurs in two forms, a childhood form due to cytomegalovirus infection and an adult form attributed to overexpression of transforming growth factor-alpha (TGF-). Our patient was a 44-year-old man, subbmited to hospital because of hemorrhagic shock. Due to the suspected malignancy, a subtotal gastrectomy was done. Currently, the patient is well and in good condition. In differential diagnosis, an infiltrating cancer, and Zollinger-Ellison syndrome were being considered.

Key words: **Menetrier disease, gastric body, hemorrhagic shock**

INTRODUCTION

The upper edge of normal stomach body mucous membrane from the surface to the muscularis mucosae is 1 - 1.4 mm (1). Foveolar layer occupies 1/5 - 1/4 of mucous membrane thickness and the rest is glandular layer. Foveolar part is a bit thicker in antrum, less sharply bounded from the glandular layer, and the whole mucous membrane is thinner than in the body. Each growth of mucous membrane over this limit is marked as hyperplasia. It seems that macroscopic hypertrophic creases can be made of normal mucous membrane but relevant macroscopic growth of mucous membrane and its persistence on energetic insufflation still confirms the existence of the real hyperplasia, which needed to be proved by macroparticular biopsy.

The real hyperplasia of the stomach mucous membrane is mostly restricted to the body segment and can be expanded to the antrum segment. Three basic conditions can be recognized in hypertrophic gastropathy: (a) Ménétrier's disease - hyperplasia of the foveolar layer, (b) Zollinger-Ellison syndrome - hyperplasia of the glandular layer, and (c) combined foveolar - glandular hyperplasia (2).

Ménétrier disease represents the diffuse giant growth of stomach mucous membrane, mainly of foveolar layer of gastric body mucosa in the shape of wide, long, twisted cerebral-like creases. This rare disease was first described by Ménétrier in 1888 (1). This morphological change is followed by the loss of proteins from the stomach and by the consequential low level of proteins in blood. It appears in men more often (3:1), between the fourth and the sixth decade of age. There are few family cases (only in twins) which suggest dominant heredity but of a little pathogenetic importance. The cause of this illness is not known. It is assumed that a stimulus of unknown genesis causes hyperplasia of the foveolar layer. It is related to the syndrome of multiple endo-

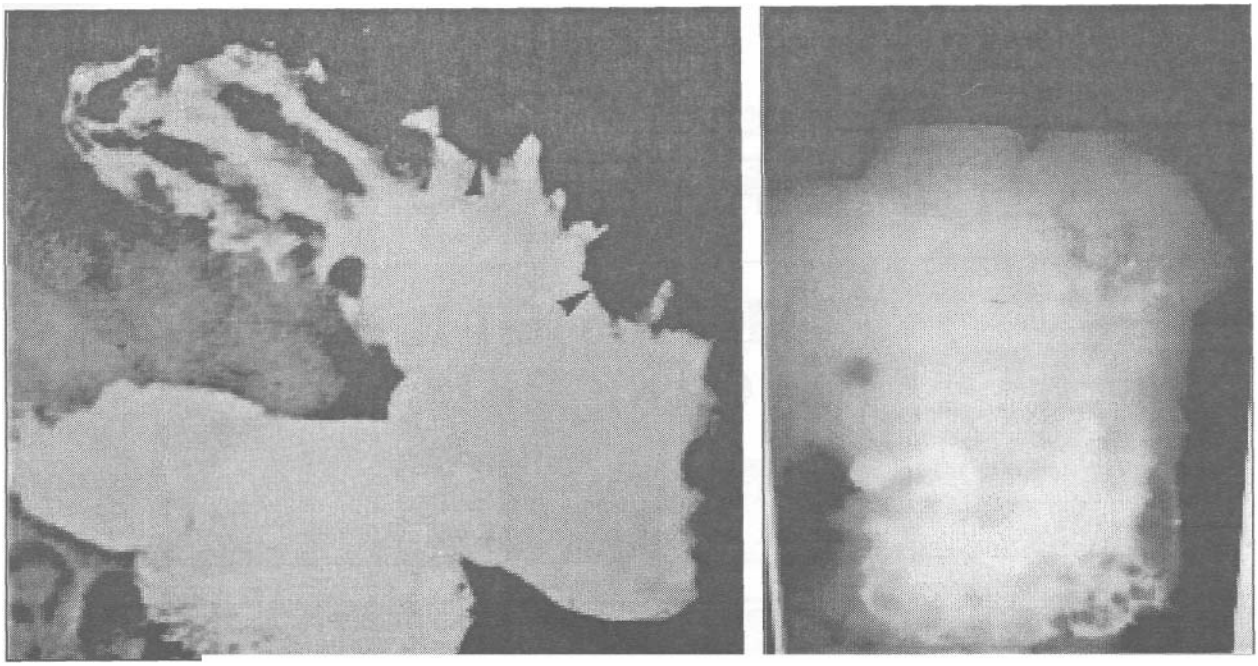

Figure 1. An x-ray of the gastro duodenum of different patients with Ménétrier disease

crine adenomatosis in some patients. The retrospective study of Hertz et al. (3), which deals with 138 patients with Ménétrier disease showed that this disease was related to *H. pylori* infection in 90% of the cases. Bayerdoffer et al. (4) showed the similar results. It is assumed that Ménétrier disease represents the special form of//, *pylori* gastritis. Previous studies showed the high correlation of Ménétrier disease and the stomach cancer. According to the new information about *H. pylori,* it is more likely to assume that the higher prevalence of cancer is related to *H. pylori* gastritis.

Histologically, giant hyperplasia of the foveolar layer, with polyp-like formations and large, bright, mucous cells, and unusually deep tortuous and often cystically dilated foveolas, at the expense of the reduced glands, can be found (5). Superficial mucous cells secrete large quantity of proteins, which leads to the low blood level of proteins. Patients have a low level of gastric acid because of the surface neutral to lightly alkali secretion and its buffer activity, as a consequence of atrophic glandular layer. The increased secretion of mucus is the cause of diarrhea.

The most often symptom is numb pain in upper abdomen, followed by atypical discomforts such as a feeling of disgust, nausea and vomiting. Subcutaneous edema may also appear as a consequence of hypoproteinemia; bleeding is also likely to appear because hyperplastic surface is vulnerable, which causes erosions (6).

Laboratory analyses show the presence of sideropenic anemia and hypoproteinemia. Hypochlorhydria to achlorhydria is present in gastric liquid. The x-ray shows a typical big and wide creases in the gastric body and eventually along its greater curvature which disappear at the antral edge (figure 1).

Endoscopically, the cobblestones creases of gastric body mucosa (figure 2), along the greater curvature are very voluminous and raised, without smoothing at maximal air insufflation. Another characteristic is the presence of large amount of transparent mucus which can not be easily absorbed. The diagnosis must be set by macro particular biopsy (7).

The most usual complications are hypoproteinemia, edema, stomach ulcer, and malignant alteration. It is recommended to have examinations once a year, and taking biopsy from different parts, especially those which show the most bizarre hyperplasia.

In differential diagnosis, an infiltrating cancer, and Zollinger-Ellison syndrome must be considered.

The treatment consists of more frequent, light snacks, high-protein diet, albumins, plasma, blood, diuretics, anticholinergics, etc. Surgical intervention is recommended in the following states: tumors proliferation, dysplasia in biopsies, high loss of proteins and recurrent bleeding which cannot be controlled.

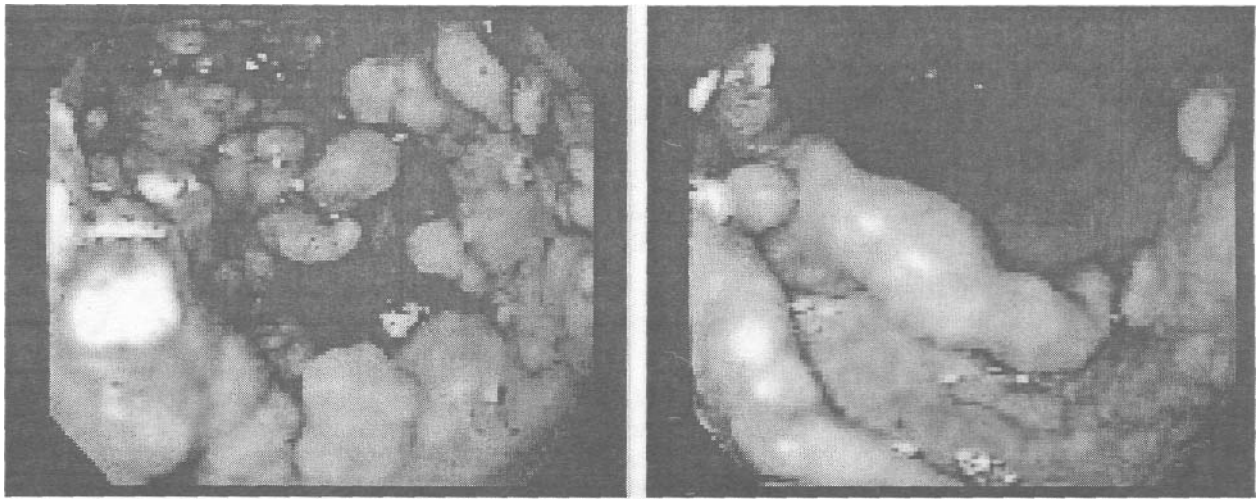

Figure 2. Endoscopic view of Ménétrier disease

CASE REPORT

A male patient, J.M., 44 years old, was sent to Surgical Clinic, Niš, under the diagnosis of hemorrhagic gastric ulcer, accompanied with shock. On the basis of anamnestic data, the patient had not been treated before and the first sign of the disease was vomiting of fresh blood more than once - that is the reason he came to the doctor's. During the surgery subtotal gastrectomy was done because of bizarre hypertrophical creases in the gastric body, with multiple eroded mucous membrane, and a risk of neoplasm. The material was analyzed at Institute of Pathology, Clinical Center of Niš (figure 3).

Pathohistological sections, 5 um thick, made from parafine blocks are stained by hemotoksylin and eozin (HE) (figure 4).

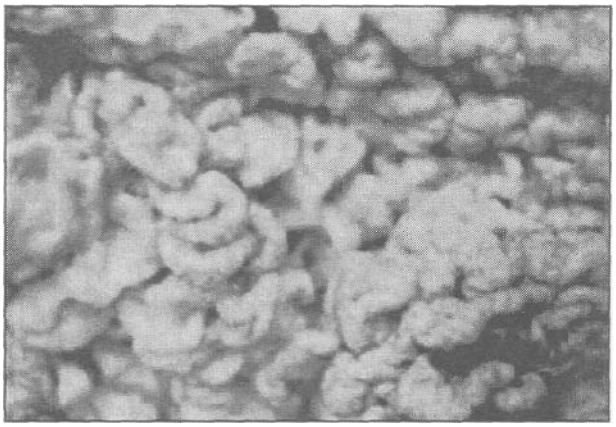

Figure 3. Macroscopic view of surgical material

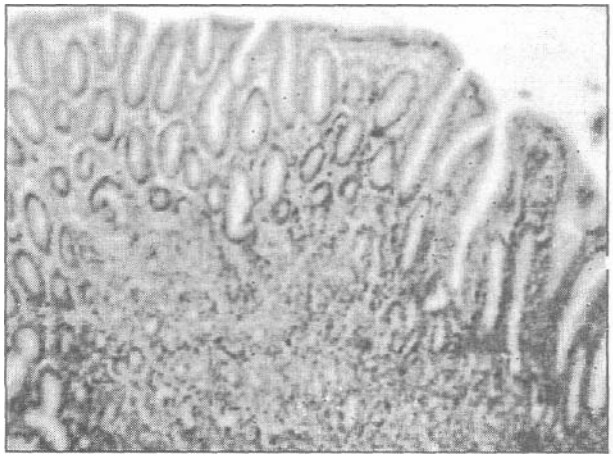

Figure 4. Gastric body mucous membrane is thicker, foveolas are longer and cystically changed, HE, obj. x 10

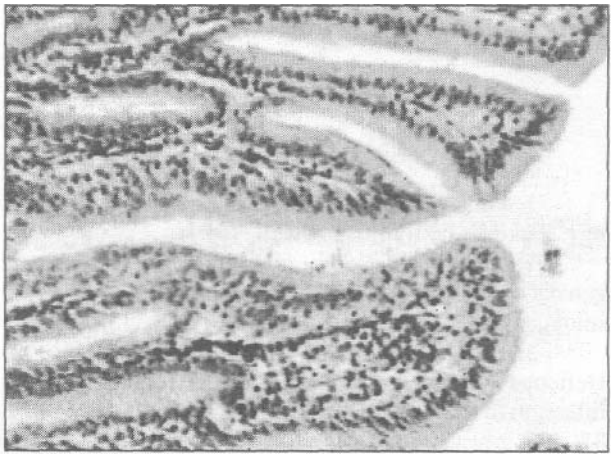

Figure 5. Foveolas are covered with high columnar cells producing mucus. HE, obj. x 25

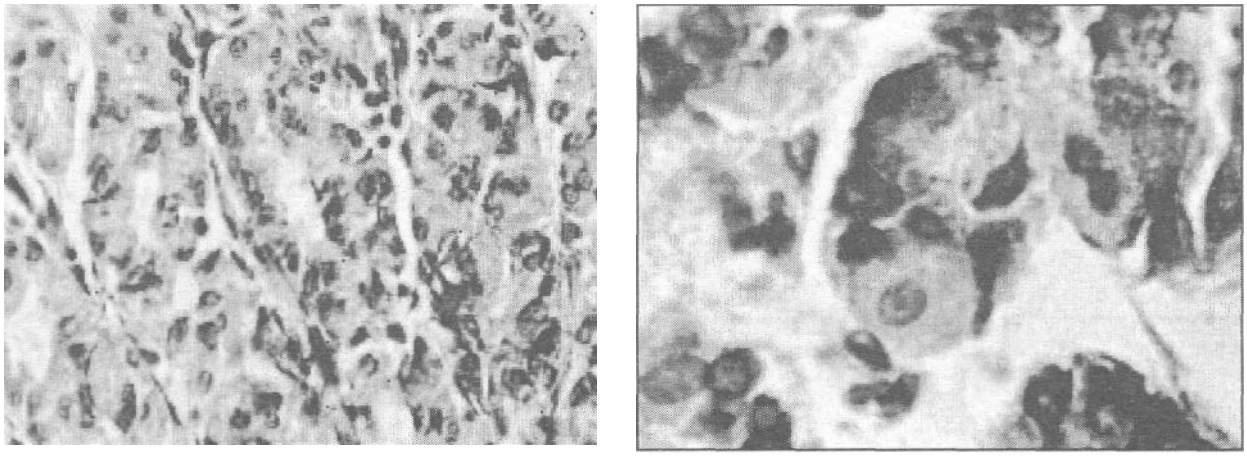

Figure 6. Parietal cells in gastric body mucosa. HE, (a) obj.x10, (b) obj. x 63

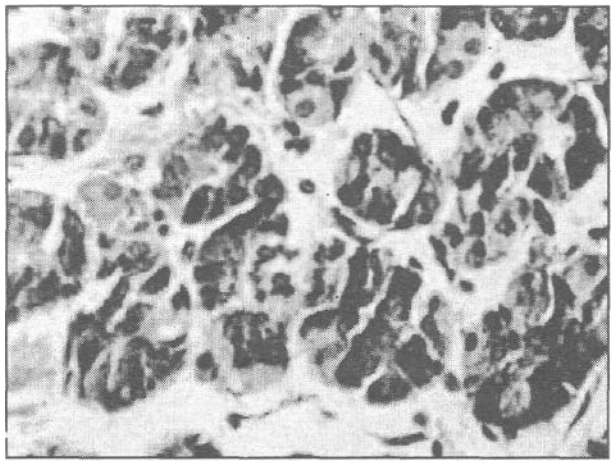

Figure 7. Reduced number of chief cell in body mucosa. HE, obj. x 25

DISCUSSION

Ménétrier disease (hyperplastic hypersecretory gastropathy) is uncommon disorder of the stomach characterized by enlarged folds. It is often accompied by a severe loss of plasma proteins (including albumin) from the altered gastric mucosa. The disease occurs in two forms, a childhood form due to cytomegalovirus infection and an adult form attributed to overexpression of transforming growth factor-alpha (TGF-a)(8).

Our patient was a 44-year-old man, subbmited to hospital because of hemorrhagic shock. Due to the suspected malignancy, a subtotal gastrectomy was done. Currently, the patient is well and in good condition.

CONCLUSION

Ménétrier disease represents the diffuse giant growth of stomach mucous membrane, mainly of foveolar layer of gastric body mucosa in the shape of wide, long, twisted cerebral-like creases. In differential diagnosis, an infiltrating cancer, and Zollinger-Ellison syndrome were taken into consideration.

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MENETRIEROVA BOLEST

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SAŽETAK

Menetrier-ova bolest (Hiperplastična hipersekretorna gastropatija) je retka bolest želuca koja se karakteriše uvećanim želudačnim naborima. Često je udružena sa velikim gubitkom plazmatskih proteina (uključujući i albumine) iz oštećene gastrične mukoze. Bolest obuhvata dve forme, kod dece u toku citomegalo virus infekcije i kod odraslih usled preterane izloženosti transformišućem faktoru rasta-a (TGF-a). Naš pacijent je 44 god. star muškarac primljen u bolnicu zbog hemoragičnog šoka. Zbog sumnje na malignitet urađena je subtotalna gastrektomija. Pacijent je u dobroj kondiciji i oseća se dobro. U diferencijalnoj dijagnozi razmatrani su infiltrativni karcinom i Zollinger-Elissonov sindrom.

Ključne reči: **Menetrierova bolest, želudac, hemoragični šok**