Ménétrier's disease

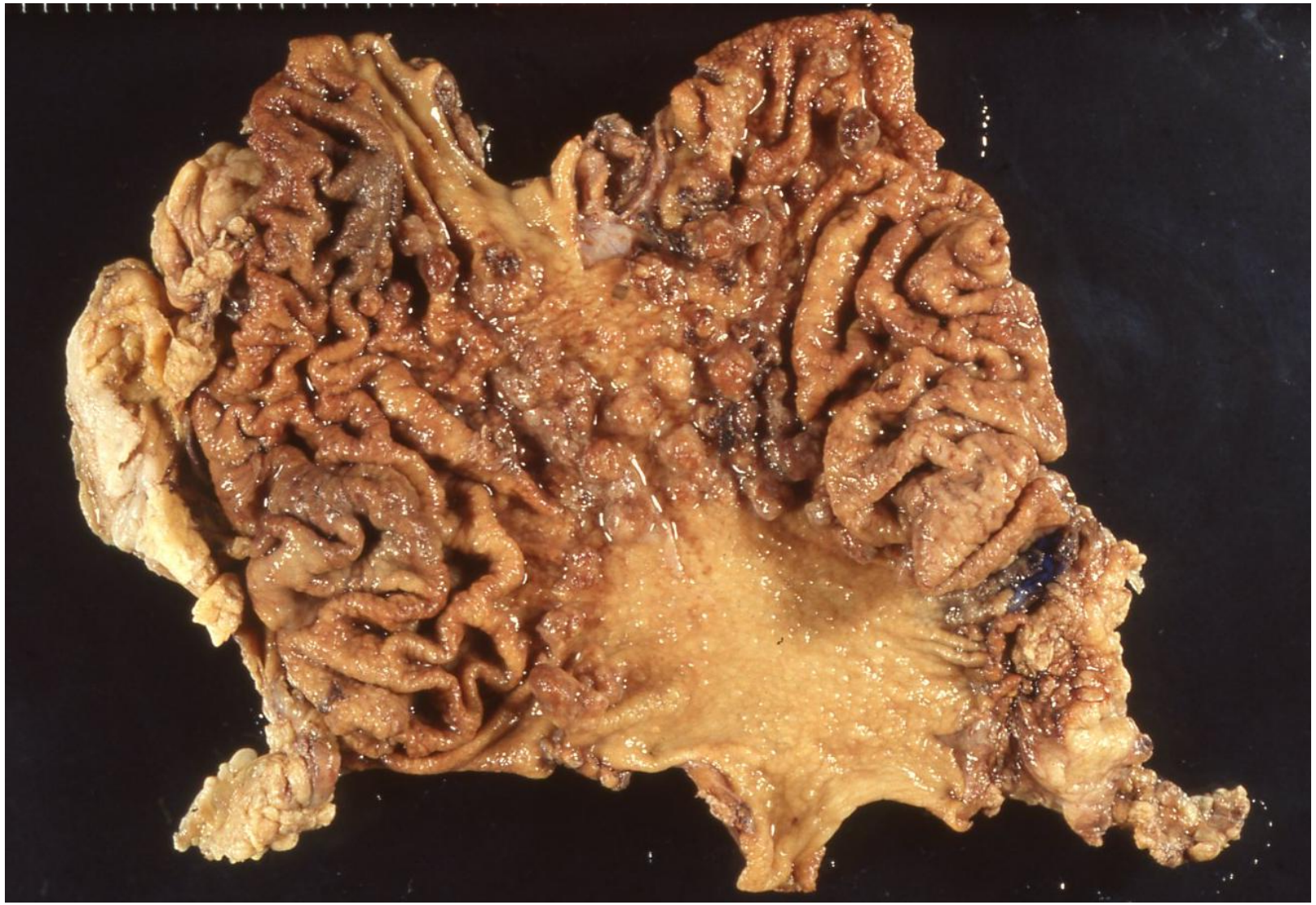
Ménétrier's disease, an acquired disease of the stomach mainly of adults aged 40 to 60 years, is featured by large, tortuous gastric folds, gastric epithelial hyperplasia and excessive mucus production in the gastric fundus. The fundic mucosa reveals a cobblestone or cerebriform appearance, The antral mucosa is spared. The association of protein-losing gastropathy and achlorhydria leads to low blood albumin and systemic edema. The disorder is closely associated with excessive secretion of transforming growth factor-alpha (TGF-α) in the gastric mucosa. The increase in epidermal growth factor receptor (EGFR) signaling is responsible for the inhibition of acid production and epithelial cell proliferation of the mucosa, Microscopically, massive foveolar cell hyperplasia with corkscrew-like appearance and cystic dilation is characteristic. Inflammation is not prominent, but in some cases marked intraepithelial lymphocytosis may be observed. The fundic glands show diffuse or patchy atrophy, with decrease of parietal and chief cells. Dysfunction of the tight junction of the foveolar cells may cause the protein loss. Glucagon cells may be found among the gland, a feature of fetal stomach (see GI-45-1-stomach), and can suppress gastric acid secretion. Hypersomatostatinemia may be associated. The cause is unknown. It has been hypothesized that CMV infection in children and *Helicobacter pylori* infection in adults may trigger the disease. Cetuximab, a monoclonal antibody against EGFR, is the first-line therapy for Ménétrier's disease. Severe disease with persistent and substantial protein loss may require total gastrectomy.

Ref.-1: Weisenberg E. Menetrier disease. PathologyOutlines.com website. 2025. https://www.pathologyoutlines.com/topic/stomachmenetriers.html

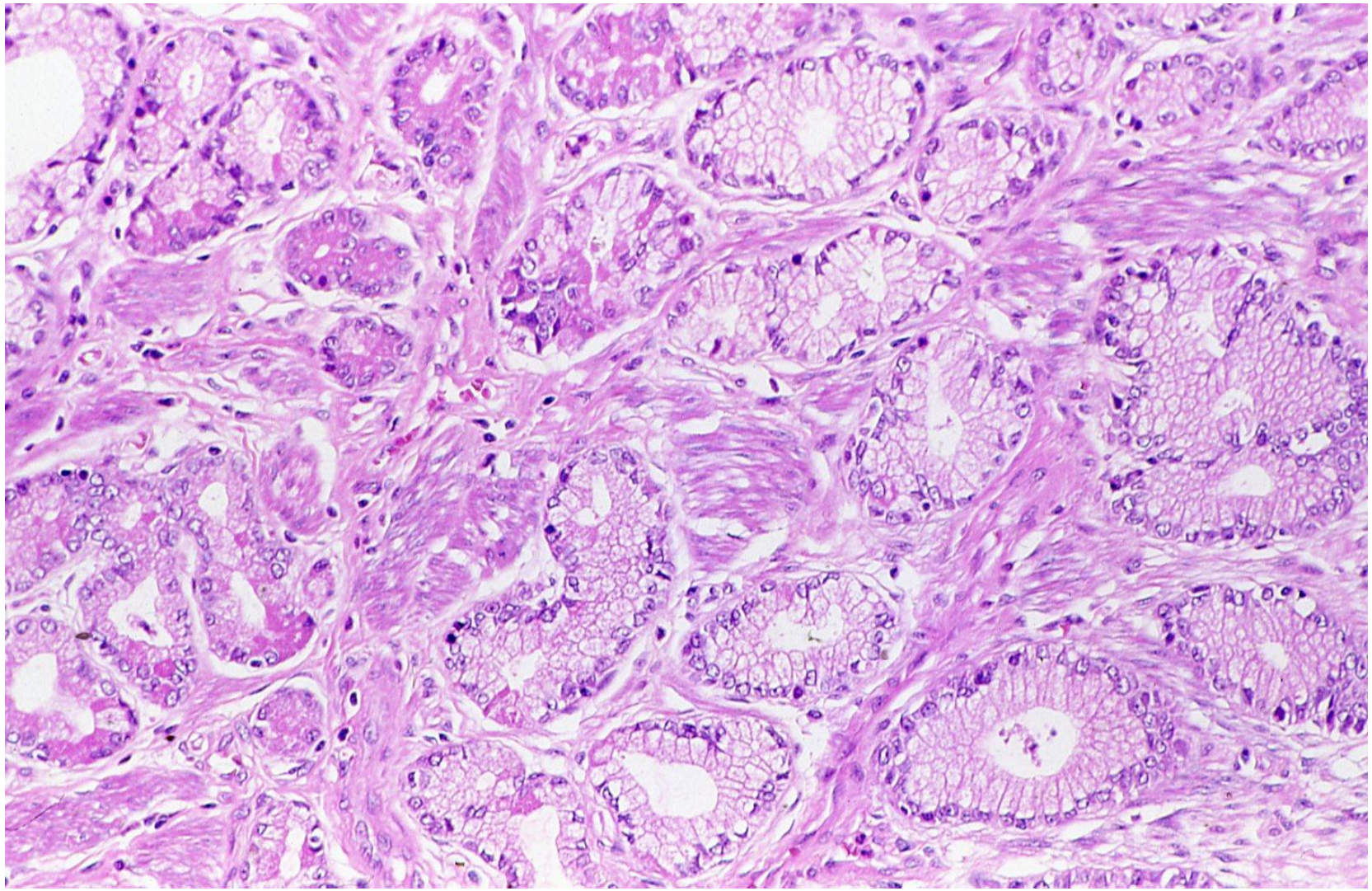
Ref.-2: Monés J, et al. Circulating hypersomatostatinaemia in Menetrier's disease. Horm Metab Res 1991; 23(3): 146-147. doi: 10.1055/s-2007-1003637

Case presentation

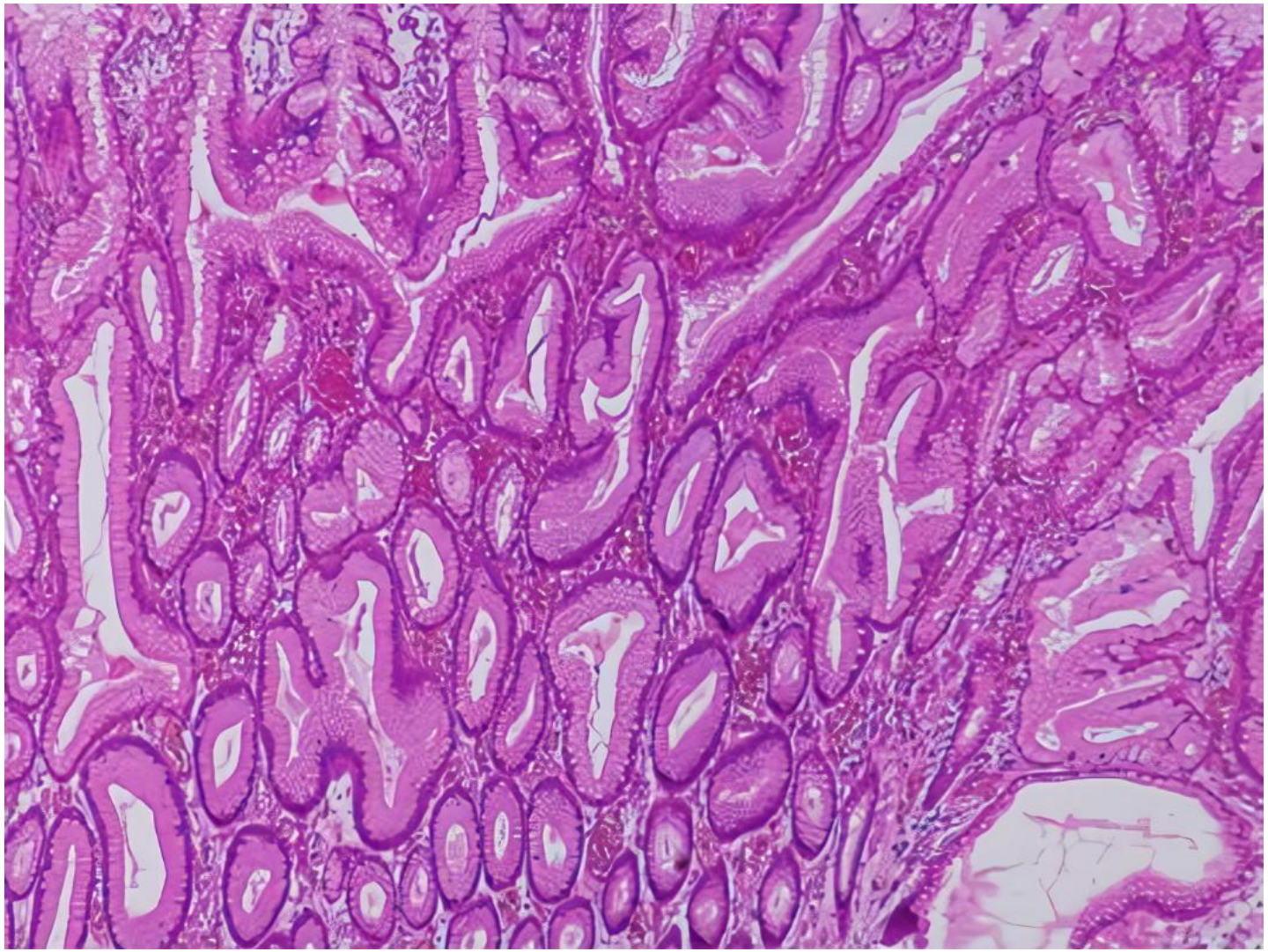
A 54 y-o male patient complained of epigastric dull pain and appetite loss. Weight loss (5 kg in half year) and edema on the lower legs were associated. Iron-deficiency anemia was pointed out by a local doctor. Laboratory data on admission included RBC 3,200,000/μL, Ht 28%, WBC 3,400/μL, platelets 340,000/μL, Fe 23 μg/mL, and total protein 5.2 g/dL. Protein-losing test using ^{51}Cr-albumin was positive. Gastric endoscopy revealed giant folds on the gastric body mucosa with mucin hypersecretion. Protein-losing was severe and resistant to therapy, and therefore total gastrectomy was performed.



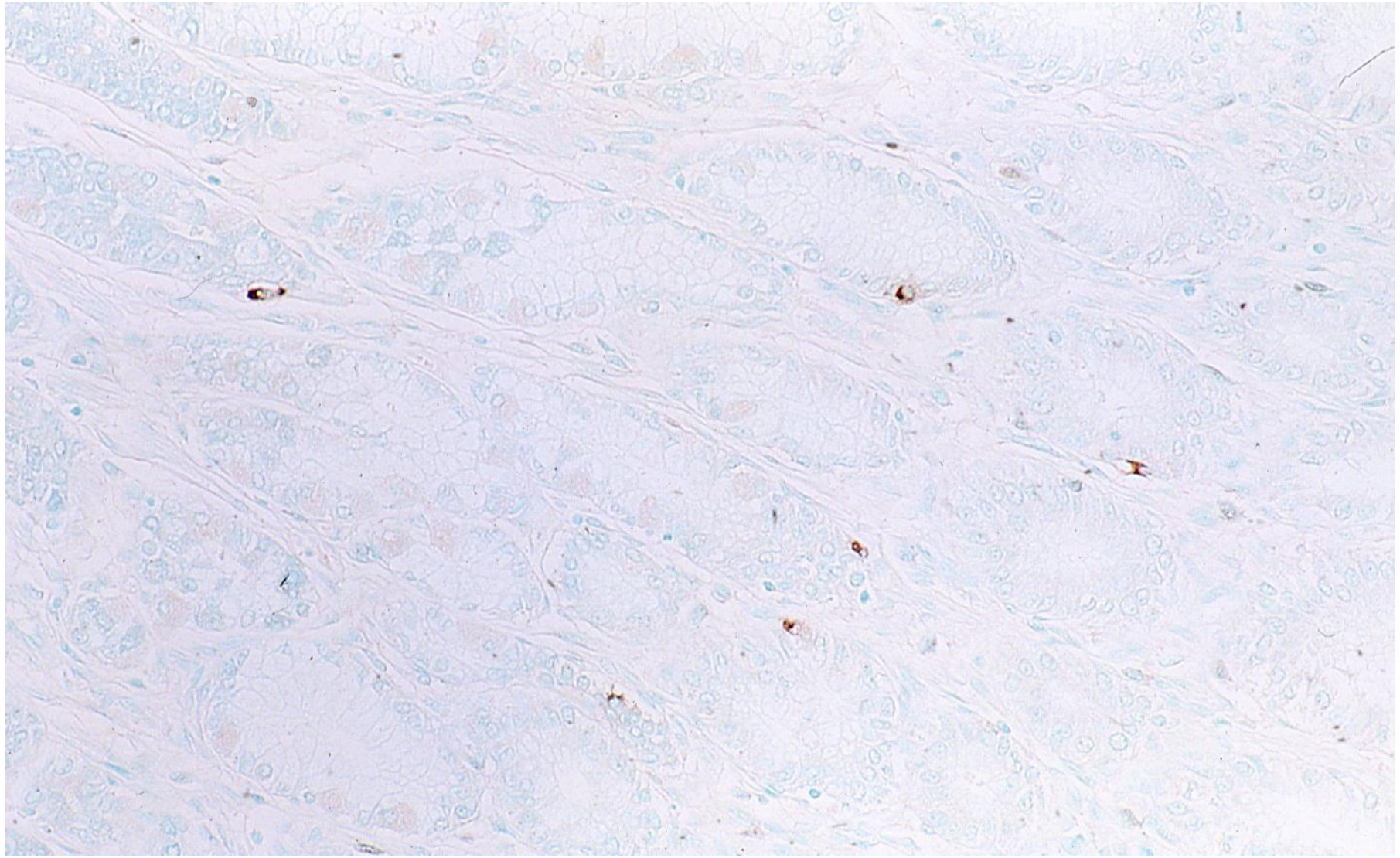
Ménétrier disease seen in a 54 y-o male patient. In the present case, intractable protein-losing gastropathy necessitated total gastrectomy (gross appearance after formalin fixation). Giant folds in the gastric body mucosa is characteristic. The antral mucosa is spared.



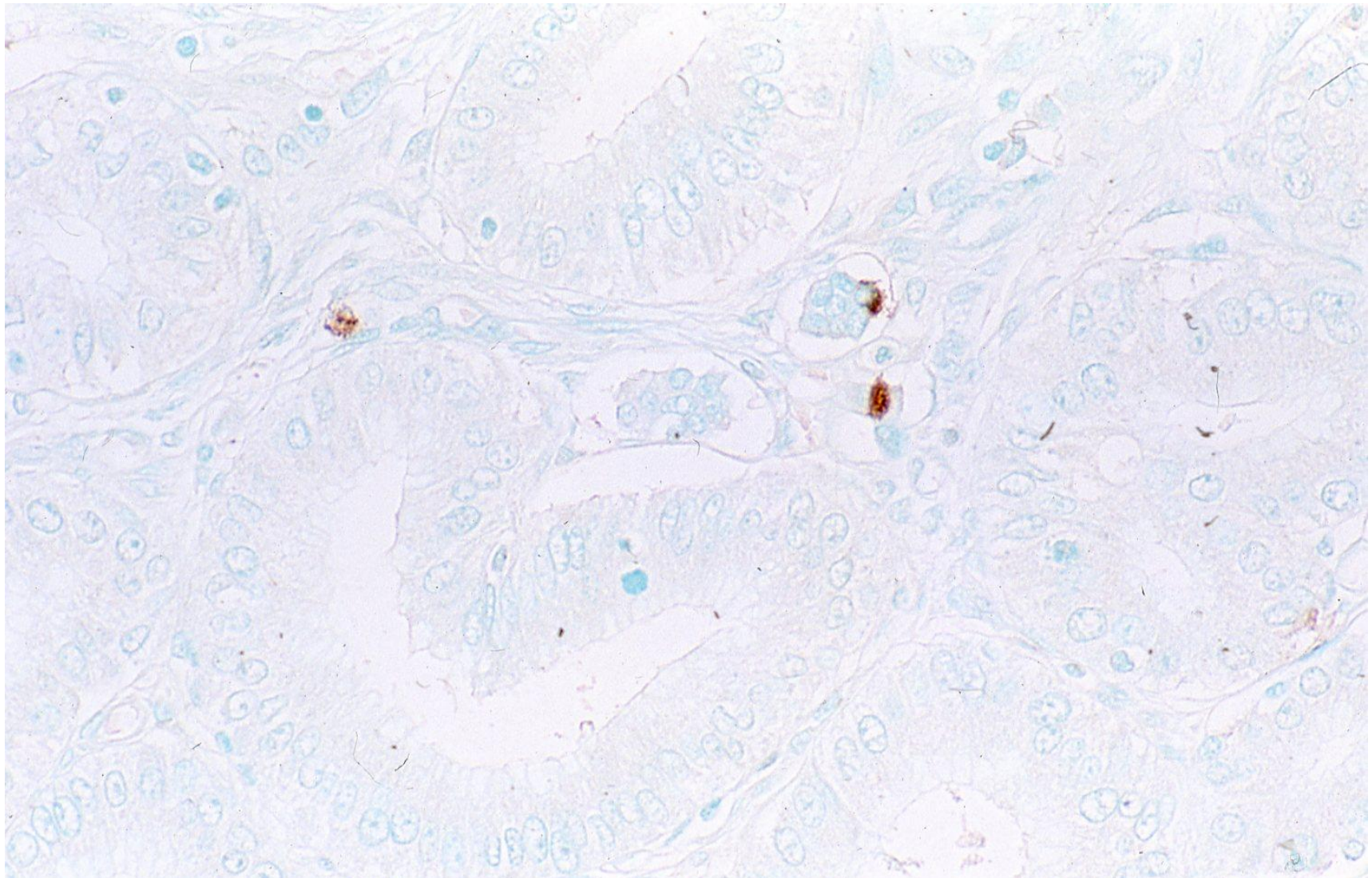
Ménétrier disease seen in a patient aged 50's. Intractable protein-losing gastropathy necessitated total gastrectomy. Microscopically, the giant fold fundic mucosa reveals foveolar cell hyperplasia with atrophic oxyntic glands. Inflammation is minimal (H&E-1).



Ménétrier disease seen in a patient aged 50's. Intractable protein-losing gastropathy necessitated total gastrectomy. Microscopically, the giant fold fundic mucosa reveals diffuse foveolar cell hyperplasia. Inflammation is minimal (H&E-2).



Ménétrier disease seen in a patient aged 50's. Intractable protein-losing gastropathy necessitated total gastrectomy. It should be noted that glicentin (proglucagon)-positive glucagon cells are distributed in the atrophic fundic gland, a phenomenon of fetalism (immunostaining for glicentin).



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