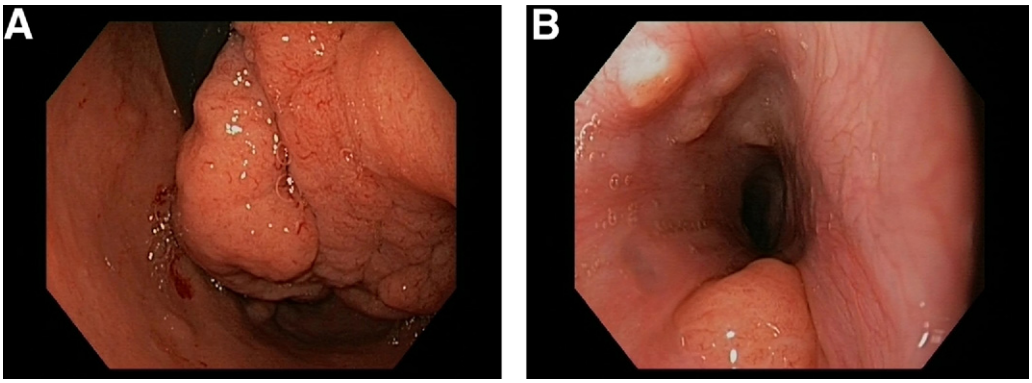
David A. Katzka and David L. Jaffe, Section Editors

Polypoid Lesions in the Stomach and Proximal Esophagus



Question: A 50-year-old man presented at our gastrointestinal clinic with a several-month history of dyspeptic symptoms without alarming features such as weight loss, vomiting, or gastrointestinal blood loss. He was not taking any medication. He had undergone an atypical resection of a part of the gastric fundus, 17 years ago, for a small carcinoid tumor with a subsequent unremarkable follow-up. Physical examination was normal. Laboratory tests showed a microcytic hypochromic anemia (hemoglobin, 9.6 g/dL), iron deficiency (serum iron, 22 μg/dL; serum ferritin, <5 μg/L), and serum albumin at the lower limit of normal (3.5 mg/dL). International normalized ratio coagulation tests, C-reactive protein, and white blood cell count, as well as liver and pancreatic enzyme values, were within normal ranges.

Upper gastrointestinal endoscopy showed a deformation of the upper part of the stomach owing to the partial gastrectomy, and in addition there were diffusely enlarged folds in the remaining proximal stomach (Figure *A*). Random deep biopsies and a snare resection of the top of a fold were performed. In the cervical esophagus a nodular polypoid lesion of about 20 mm in diameter, with a salmon-colored, smooth surface (Figure *B*) was identified and biopsies were taken.

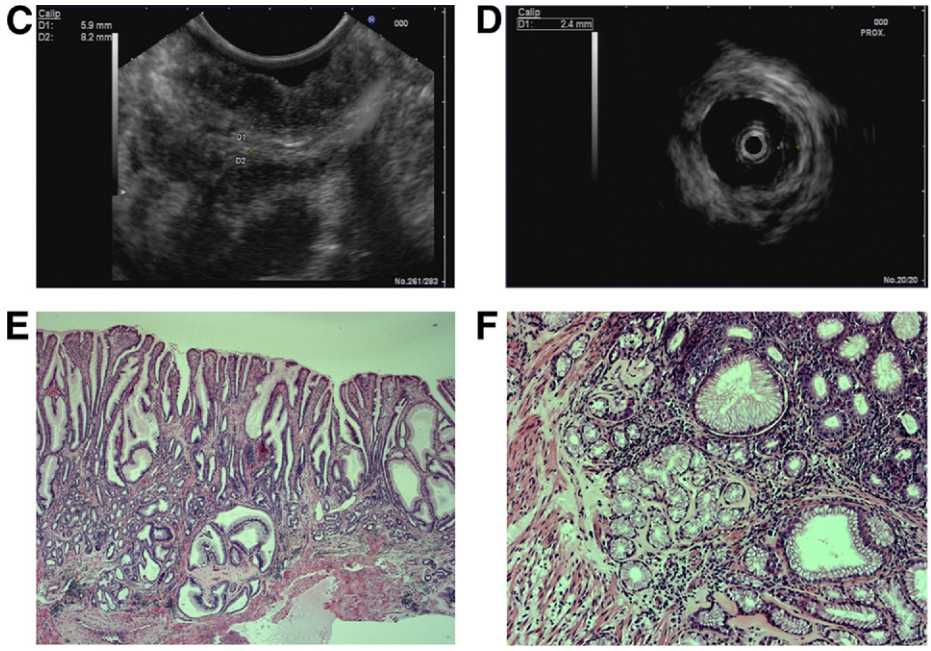
What further diagnostic imaging do you recommend for the 2 lesions? What are your diagnoses?

Look on page 369 for the answer and see the GASTROENTEROLOGY web site (www.gastrojournal.org) for more information on submitting your favorite image to Clinical Challenges and Images in GI.

MASSIMO MAFFEI
HUBERT PIESSEVAUX
Gastroenterology Departement
ANNE JOURET-MOURIN
Pathology Departement
Cliniques universitaires St-Luc
Brussels, Belgium

Conflicts of interest
The authors disclose no conflicts.

0016-5085/$36.00
doi:10.1053/j.gastro.2010.01.067



Answer to the Clinical Challenges and Images in GI Question: Image 1 (page 32): Ménétrier's Disease of the Stomach

Endoscopic ultrasonography disclosed a diffusely important thickening of the gastric mucosa with focal cystic dilatation (Figure *C*). Similar observations were made at the level of the esophageal lesion (Figure *D*). Gastric biopsies (Figure *E*) showed prominent expansion of the gastric pits with tortuous foveolar hyperplasia, hypertrophic muscularis mucosae with smooth muscle extension into the lamina propria, and focally cystic dilatation of the glands. No dysplasia was seen. Similar observations were made on the esophageal samples (Figure *F*). A diagnosis of Ménétrier's disease of the stomach and similar lesions in an esophageal heterotopic gastric mucosa (HGM) was made. The patient refused any therapeutic intervention.

This is the first reported case of development of Ménétrier's disease with comparable lesions in an HGM. This case is also peculiar because the disease occurred in a previously operated stomach. Revision of the initial surgical specimen showed no evidence of Ménétrier's at the time of the initial surgery. Ménétrier's disease was first described in 1888.[1] This disease is caused by dysregulated epidermal growth factor receptor signaling.[2] The over-expression of transforming growth factor-α, a ligand for epidermal growth factor receptor, results in selective expansion of surface mucous cells in the body and fundus of the stomach. The reason for this overexpression is still unknown. In addition, transforming growth factor-α is not over-expressed elsewhere in individuals with Ménétrier's disease.[2] Patients' signs and symptoms include abdominal pain, nausea and vomiting, anemia, hypochlorhydria, and peripheral edema. Diagnosis usually requires deep gastric macrobiopsies. Gastrectomy is often recommended. Alternatively, epidermal growth factor receptor inhibitors such as cetuximab have demonstrated favorable results.[2]

HGM is a congenital developmental anomaly that can be observed along the entire gastrointestinal tract.[3] In the cervical esophagus, it is called "inlet patch" and its prevalence is reported between 0.1% and 10%. Malignant transformation to adenocarcinoma is exceedingly rare; therefore, HGM is not considered a precancerous lesion. The present observation of Ménétrier's disease in an HGM suggests the presence of a gastric mucosa-specific trigger for transforming growth factor-α alpha over-expression.

References

1. Ménétrier P. Des polyadenomes gastriques et leur rapport avec le cancer de l'estomac. Arch Physiol Norm Pathol 1888;1:236–262.
2. Coffey RJ, Washington MK, Corless CL, et al. Ménétrier disease and gastrointestinal stromal tumors: hyperproliferative disorders of the stomach. J Clin Invest 2007;117:70–80.
3. Von Rahden HAB, Stein HJ, Becker K, et al. Heterotopic gastric mucosa of the esophagus: literature-review and proposal of a clinicopathologic classification. Am J Gastroenterol 2004;99:543–551.