Ménétrier’s Disease

Menetrier's disease is characterized by hypertrophied gastric rugae, protein loss, parietal cell atrophy, decreased gastric acid production, and increased TGF-alpha. It is associated with cytomegalovirus in children and Helicobacter pylori in adults. GI distress and edema are the most common symptoms. Endosopy with biopsy is the gold standard for diagnosis. Menetrier's disease is treated with supportive therapy, cetuximab, and surveillance.



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Characteristics

Hypertrophied Gastric Rugae
Hiker-trophy Stomach Rug
Histologically, Ménétrier’s disease can be mistaken for other disorders that also present as hypertrophic gastropathy. Mucosal hypertrophy is characterized by the thickening of gastric mucosa into giant, brain-like rugae. This is due to hypertrophy of mucus-producing cells in the gastric epithelium.

Protein Loss
Down-arrow Mr. Protein
Mucus is normally secreted by foveolar cells, found in the necks of the gastric pits that are the open part of the gastric glands. Non-inflammatory hyperplasia of these mucus producing cells leads to excess mucus production. For this reason, Ménétrier’s disease is a type of protein-losing gastropathy.

Parietal Cell Atrophy
Pirate Cell with @-trophy
Parietal cells secrete gastric acid (HCl) into the stomach through the gastric pits. These form a portion of the oxyntic glands (fundic glands). Patients with Ménétrier’s disease present with atrophic oxyntic glands and reduced or absent parietal cells.

Decreased Acid Production
Down-arrow Acidic-lemon
Decreased acid production occurs due to parietal cell loss via inhibition of acid secretion through TGF?-EGFR signaling. The increase of gastric pH can also occur because of the buffering capacity of excessive mucus production.

Increased TGF-alpha
Up-arrow Transformer-giant-flagger-Afro
Elevated TGF-? is found in the gastric mucosa of patients with Ménétrier’s disease. TGF-? is a ligand that binds epidermal growth factor receptor (EGFR) which upregulates cellular proliferation. Overexpression of TGF-? seems to defer gastric progenitor cells to surface mucous cell differentiation rather than shunting them towards becoming parietal or chief cells.

Associations

Cytomegalovirus (CMV)
Side-toe-mega-virus
Cytomegalovirus can be related to Ménétrier’s disease in children. It often resolves spotaneusly within 1 to 5 months with supportive treatment only.

Helicobacter Pylori
Helicopter-bacteria
Many patients undergo testing for H. pylori, and if found are treated for it. Eradication of H. pylori may induce spontaneous regression of the disease.

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Symptoms

GI Distress

GI-guy with flare-gun

The classic symptom triad of Menetrier disease is gastrointestinal symptoms, peripheral edema, and giant gastric folds. The most common GI symptoms include epigastric pain (65%), asthenia (60%), anorexia (45%), weight loss (45%), and vomiting (37.5%).

Edema

Edamame

Protein loss may lead to a decrease in oncotic pressure that can result in edema, ascites, and pleural effusions. Significant hypoalbuminemia may develop.

Diagnosis

Endoscopy with Biopsy

Endoscopy and Biopsy-needle

Histology is the gold standard for diagnosing Ménétrier's disease. Endoscopy (gastroscopy) will show giant rugal folds predominantly in the greater curvature of gaster, commonly in the fundus and body, which needs to be confirmed by biopsy. An increased mucous surface cell thickness >1 cm, diffuse thickening of the foveolar epithelium, lack of inflammatory cells, and dilated cystic gastric glands are often seen on histological specimens. Parietal and chief cell counts are often reduced as well.

Management

Supportive Therapy

Supportive IV Bags

Unfortunately, there is no cure for Ménétrier's disease. Supportive therpay with symptom control is the standard of care. This includes a high protein diet, proton pump inhibitors, and H. pylori eradication. Severe cases may require gastectomy.

Cetuximab

C-tux-mob

Cetuximab may be used in Ménétrier disease since it theoretically can target the TGF-alpha / EGFR overexpression. It is approved by the FDA on a compassionate need basis.

Surveillance

Surveillance-camera

Ménétrier’s disease might be a precancerous state. Reports have found a 2-15% increased lifetime risk in gastric adenocarcinoma. For this reason, surveillance with regular follow-ups and endoscopy is important.

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