Ménétrier's Disease

Synonyms: Menetriere's disease, giant hypertrophic gastritis, protein-losing gastropathy, hypertrophic gastropathy

This much misspelled disease was named for Pierre Eugène Ménétrier, a French physician (1859-1935) who first described it in 1888.[1]

It is a rare condition characterised by gross hypertrophy of the gastric mucosa, resulting in an increased production of mucus, reduced acid secretion and, due to selective serum protein loss across the gastric mucosa, severe hypoproteinaemia.[2]

A similar hypertrophic gastritis has been recognised with Helicobacter pylori infection and in children associated with cytomegalovirus infection (a self-limiting condition).[3] Recent work suggests that H. pylori isolated from patients with Ménétrier's disease is genetically different from that isolated from patients with other gastric diseases.

Aetiology

Research implicates overproduction of transforming growth factor-alpha with increased signalling of the epidermal growth factor receptor (EGFR) in the pathogenesis of the condition. Activation of the EGFR, a transmembrane receptor with tyrosine kinase activity, triggers a cascade of downstream, intracellular signalling pathways that leads to expansion of the proliferative compartment within the isthmus of the gastric mucosa (oxyntic) cells. The result is that production of gastric mucus is increased and production of gastric acid is decreased.[3] The existence of the disease in twins suggests that at least some cases have a genetic cause.[4]

Presentation

Symptoms include nausea, epigastric pain, weight loss and diarrhoea ± melaena.[5] The severe hypoproteinaemia can result in ascites and gross oedema.

Differential diagnosis

The condition needs to be differentiated from other forms of hypertrophic gastropathy such as Zollinger-Ellison syndrome, hypertrophic hypersecretory gastropathy, lymphocytic gastritis, or gastric carcinoma.[5, 6]

Associated diseases

One study reported two HIV-positive patients with Kaposi's sarcoma of the stomach who also had Ménétrier's-type changes in the gastric mucosa.[7]

Investigations

- The diagnosis of choice is gastroscopy with gastric mucosal biopsy. Characteristic histological changes include foveolar hyperplasia, cystic dilation of pits and reduced numbers of parietal and chief cells.[3] (Foveolar cells are mucous cells that cover the gastric mucosal surface and line the gastric pits.)
- Transabdominal ultrasound can provide additional information which helps to differentiate patients with giant gastric folds associated with malignancy from those with benign conditions.[8]
Management

- There is evidence that Ménétrier’s disease is linked to cytomegalovirus in children and H. pylori in adults and, if either of these infections is present, it should be eradicated. [3]
- Chronic disease is sometimes improved with H2-receptor antagonists or proton pump inhibitors. [9]
- Octreotide, a somatostatin analogue, has been used to good effect in some patients and may obviate the need for surgery. [10]
- Effective treatment has been achieved using a blocking monoclonal antibody specific for the EGFR. [11]
- Partial gastrectomy is occasionally recommended for persistent symptoms and particularly for uncontrollable protein loss. A total laparoscopic gastrectomy has also been reported. [12]

Complications

There is a well-documented link between Ménétrier’s disease and gastric carcinoma.

Further reading & references

5. Menetriere disease; Orphanet
9. Ishikawa T, Ando T, Obayashi H, et al; Helicobacter pylori Isolated from a Patient with Menetrier's Disease Increases Hepatocyte Growth Factor mRNA Expression in Gastric Fibroblasts: Comparison with Helicobacter pylori Isolated from Other Gastric Diseases. Dig Dis Sci. 2007 Nov 8;

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