USMLE 2 CK

GASTROENTEROLOGY

DISEASES OF THE ESOPHAGUS

General

The majority of diseases of the esophagus result in dysphagia. Dysphagia refers specifically to difficulty swallowing. Only a few of the diseases of the esophagus result in pain on swallowing, called odynophagia. Both dysphagia and odynophagia will result in weight loss if the symptoms persist for more than a few days. The mere presence of dysphagia or odynophagia is not sufficient to help one establish a diagnosis.

In general, a barium swallow or barium esophagram is a good answer to questions asking for the best initial test. This is not an absolute answer, however, and if there are clear signs of obstruction, then the answer could also be upper endoscopy as the best initial test.

Achalasia [(1)]

Pathogenesis. Achalasia is the idiopathic loss of the normal neural structure of the lower esophageal sphincter (LES). The LES is usually contracted to prevent the acidic gastric contents from refluxing backward into the esophagus. For swallowing to occur, there is normally a relaxation process of the LES in order to allow food to pass into the stomach. Inhibitory neurons are stimulated, blocking the impulses that cause constriction. In achalasia, these inhibitory neurons have been lost, as well as the ability to relax the LES. The vast majority of cases are of unknown etiology. A very small number can be from Chagas disease, gastric carcinoma, or disease that can infiltrate into the area, such as lymphoma.

Clinical Presentation. Achalasia presents with progressive dysphagia to both solids and liquids simultaneously. Achalasia has no relationship with alcohol or tobacco use. This is different from esophageal cancer, which not only usually presents with dysphagia to solid foods that progresses to difficulty swallowing liquids, but is more common in older patients with a long history of alcohol and tobacco use.

Diagnosis. Although a chest x-ray may show an air-fluid level in the dilated esophagus, plain radiography is insufficiently accurate to be very useful. Barium esophagography is very accurate and shows dilation of the esophagus, which narrows into a “bird’s beak” at the distal end. The most accurate rest overall is esophageal manometry.

Treatment. The best initial therapy is with pneumatic dilation. Pneumatic dilation should be effective in 80 to 85% of patients. Botulinum toxin injections into the LES are used in those patients not willing to undergo pneumatic dilation, or in whom it has failed. Although the botulinum toxin is relatively benign, the main limiting factor in its use is a need for additional injections in a few months. Fifty percent will relapse in 6 to 9 months, and all patients will need re-injection after 2 years. If both pneumatic dilation and botulinum toxin injections fail, then surgical myotomy is performed. Myotomy is performed laparoscopically and results in reflux in 20% of patients as a complication of therapy.

Esophageal Cancer [(2)]

Pathogenesis. Esophageal cancer is linked to the synergistic, carcinogenic effect of alcohol and tobacco use for cases of squamous cell cancer in the proximal two thirds of the esophagus. Adenocarcinoma is found in the distal third of the esophagus and is associated with long-standing gastroesophageal

reflux disease and Barrett esophagus [6]. The rate of development of cancer from Barrett esophagus is between 0.4 and 0.8% per year.

Clinical Presentation. Esophageal cancer presents with progressive dysphagia first for solid food, then for liquids. Weight loss is prominent. Rarely, halitosis, regurgitation, and hoarseness occur. Hypercalcemia may arise, as it can with most cancers.

Diagnosis. Although a barium swallow can be done first, endoscopy is mandatory because this is a diagnosis that requires a tissue biopsy. CT scanning detects the degree of local spread, and bronchoscopy detects asymptomatic spread into the bronchi.

Treatment. The only truly effective therapy for esophageal carcinoma is surgical resection if the disease is sufficiently localized to the esophagus. Only 25% of patients are found to be operable. A total of 10 to 20% will die from the surgery. Five-year survival is between 5 and 20%. Chemotherapy with 5-fluorouracil is combined with radiation to control locally metastatic disease.

Schleroderma (Progressive Systemic Sclerosis) [3]

Pathogenesis. As many as 80 to 90% of patients with scleroderma will develop diminished esophageal peristalsis from the atrophy and fibrosis of the esophageal smooth muscle.

Clinical Presentation. Although there is dysphagia, the main clue to the diagnosis is simply the presence of gastroesophageal reflux symptoms in a person with a history of scleroderma. The LES will neither contact nor relax and basically assumes the role of an immobile open tube.

Diagnosis. Barium studies are generally unnecessary. The most accurate diagnostic test is motility studies.

Treatment. Therapy is with proton-pump inhibitors, such an omeprazole. Metoclopramide is a promotility agent that has some modest efficacy.

Diffuse Esophageal Spasm and Nutcracker Esophagus

Pathogenesis. Esophageal spastic disorders are idiopathic abnormalities of the neural processes of the esophagus. Fundamentally, diffuse esophageal spasm and nutcracker esophagus are the same disease. The only different may be in the manometric pattern.

Clinical Presentation. These patients present with intermittent chest pain and dysphagia. The pain can simulate that of a myocardial infarction, but it bears no relationship with exertion. There is no relationship with eating, ruling out odynopaghia. The pain can be precipitated by drinking cold liquids.

Diagnosis. Barium studies may show a “corkscrew” pattern at the time of the spasm. The most accurate test is manometric studies, which will show high-intensity, disorganized contractions. Because the contractions are disorganized, they do not lead to the forward flow of food and peristalsis.

Treatment is with calcium-channel blockers, such as nifedipine, and nitrates.

Rings and Webs [(4)]

Pathogenesis. Schatzki’s ring and Plummer-Vinson syndrome reveal thin, epithelial membranes made out of squamous epithelial cells. Neither of them is progressive in nature, distinguishing both of these conditions from achalasia.

Schatzki's ring is more common and leads to intermittent dysphagia and is not associated with pain. It is also more distal and located at the squamocolumnar junction proximal to the lower esophageal sphincter.

Plummer-Vinson syndrome is more proximal and is located in the hypopharynx. The dysphagia is sometimes with liquids as well. Plummer-Vinson is associated with iron-deficiency anemia and has occasionally been associated to cancer.

Diagnosis. Both disorders are best diagnosed with a barium swallow or barium esophagram.

Treatment. Plummer-Vinson syndrome may response to treatment of the iron deficiency. Both are treated with dilation procedures.

Esophagitis [(7)]

Pathogenesis. Esophagitis is a generalized term referring to either infection or inflammation of the esophagus. The most common infection is from *Candida albicans*. When *Candida* esophagitis occurs, it is almost exclusively in patients who are HIV positive with a CD4 count below 200/mm^3, usually even less than 100/mm^3. Diabetes mellitus is the second most common risk for developing *Candida* esophagitis. Much rarer infectious etiologies of esophagitis are herpes simplex, cytomegalovirus, and aphthous ulcers. Medications and the ingestion of caustic substances have also been associated with the development of esophagitis.

Clinical Presentation. *Candida* esophagitis presents with progressive odynophagia. Although the swallowing is painful, food is still able to pass until the disease is extremely advanced. The major difference between the pain of esophagitis and the pain of spastic disorders is that in esophagitis, the pain is only on swalling, whereas with spastic disorders the pain occurs intermittently

without even needing to swallow. Esophagitis pain is simply from the mechanical rubbing of food against an inflamed esophagus as it passes by.

Diagnosis and Treatment. If the patient is HIV positive, the diagnosis is confirmed simply by seeing a response to empiric treatment with fluconazole. If the fluconazole doesn't work, then endoscopy should be performed.
Because esophagitis can also result from ingestion of medications and caustic substances, the direct effect of contact between the mucosa and the pill causes inflammation rather than infection. As with most other toxin-mediated damage to an organ, the diagnosis is based on the presentation and finding the toxin in the history. The most common pills that cause esophagitis are alendronate, risedronate, vitamin C, potassium chloride, doxycycline, NSAIDs, and iron sulfate. Pill esophagitis is managed by simply swallowing pills in the upright position and drinking enough water to flush them into the stomach.

Zenker Diverticulum [(5)]

Pathogenesis. Zenker diverticulum is the outpocketing of the posterior pharyngeal constrictor muscles at the back of the pharynx.

Clinical Presentation. This is generally a very slowly developing problem that occurs in older patients. These patients have bad breath and difficulty initiating swallowing because it is such a proximal lesion. Patients also complain of having to repeatedly clear their throats and waking up with undigested, regurgitated food on their pillow. This is particularly unpleasant because the food was usually eaten several days ago.

Diagnosis. The diagnosis is made on barium studies.

Treatment. Endoscopy and the placement of nasogastric rubes are contraindicated because of the risk of developing perforation of the pharynx. Patients with Zenker diverticulum are treated with surgical resection.