CHAPTER 1

The Patient with Dysphagia or Odynophagia

Dysphagia is the sensation of food hindered in its passage from the mouth to the stomach. Dysphagia is differentiated from odynophagia (pain on swallowing) and from globus sensation (perception of a lump, tightness, or fullness in the throat that is temporarily relieved by swallowing). The act of swallowing has four phases: the oral preparation phase, the oral transfer phase, the pharyngeal phase, and the esophageal phase. An abnormality of any of the phases can produce dysphagia. Dysphagia is usually divided into two categories: (i) oropharyngeal: disorders of the oral preparation, oral transfer, or pharyngeal phases of swallowing; and (ii) esophageal: dysfunction of the esophageal phase of swallowing (Table 1.1). The etiology and evaluation of oropharyngeal and esophageal dysphagia differ, whereas odynophagia also has a distinct underlying differential diagnosis (Figure 1.1).

Clinical presentation

History

The patient's history helps define whether symptoms are oropharyngeal or esophageal in location and structural or neuromuscular in origin. If dysphagia occurs within one second of swallowing or is associated with drooling, choking, coughing, aspiration, or nasal regurgitation, an oropharyngeal process is likely. Conversely, an esophageal cause is more likely if dysphagia occurs more than one second after swallowing, if there is retrosternal pain, or if there is regurgitation of unchanged food. Dysphagia perceived in the cervical area may result from either oropharyngeal or esophageal disease. Dysphagia perceived below the suprasternal notch is nearly always diagnostic of an esophageal source. Structural esophageal disorders generally produce solid food dysphagia with progression to liquid dysphagia only if luminal narrowing becomes severe. Patients with neuromuscular disorders of the esophagus usually report both

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Table 1.1 Differential diagnosis of dysphagia and odynophagia

Oropharyngeal dysphagia
Neurologic disorders
Cerebrovascular accident
Parkinson disease
Amyotrophic lateral sclerosis
Brainstem tumors
Multiple sclerosis
Bulbar poliomyelitis
Myogenic disorders
Myasthenia gravis
Muscular dystrophies
Polymyositis
Local mechanical lesions
Inflammation (pharyngitis, abscess, tuberculosis, radiation, syphilis)
Neoplasm
Congenital webs
Extrinsic compression (thyromegaly, cervical spine hyperostosis, adenopathy)
Radiation or caustic damage
Foreign body
Upper esophageal sphincter(UES) disorders
Primary cricopharyngeal dysfunction
Cricopharyngeal bar
Zenker diverticulum
Esophageal dysphagia
Motor disorders
Achalasia
Scleroderma or other rheumatologic conditions
Spastic motor disorders, such as jackhammer esophagus or diffuse esophageal spasm
Esophagogastric junction outflow obstruction
Chagas disease
Ineffective esophageal motility
Fragmented esophageal peristalsis
Intrinsic mechanical lesions
Benign stricture (peptic, caustic, radiation)
Schatzki ring
Carcinoma
Eosinophilic esophagitis
Esophageal webs
Esophageal diverticula
Benign tumors
Foreign bodies
Extrinsic mechanical lesions
Vascular compression – aberrant subclavian artery or thoracic aortic aneurysm
Mediastinal abnormalities
Cervical osteoarthritis
Foreign body
Functional dysphagia

Table 1.1 (*cont'd*)

Odynophagia
Mechanical
Trauma
Caustic ingestion
Foreign body
Esophageal malignancy
Inflammatory
Pill-associated ulceration – commonly doxycycline, nonsteroidal anti-inflammatory agents, slow-release potassium supplements, bisphosphonates
Infectious
Cytomegalovirus (CMV), herpes simplex virus (HSV), human immunodeficiency virus (HIV), Candida, Mycobacteria

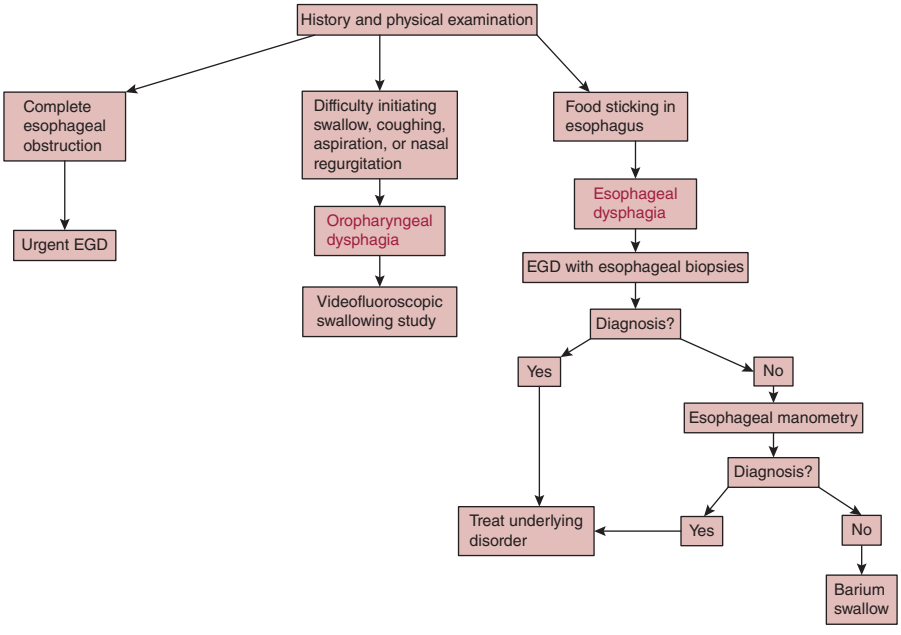

Figure 1.1 Evaluation of dysphagia or odynophagia. EGD, esophagogastroduodenoscopy.

liquid and solid dysphagia from symptom onset. Both structural and neuromuscular oropharyngeal disorders produce early liquid dysphagia. Patients with oropharyngeal dysphagia are commonly evaluated and co-managed with otolaryngologists or neurologists.

In patients with odynophagia, risk factors for opportunistic infection should be assessed and a careful medication history taken.

Physical examination

The head and neck must be examined for sensory and motor function of the cranial nerves, masses, adenopathy, or spinal deformity. Evidence of systemic

disease, including sclerodactyly, telangiectasias, and calcinosis in scleroderma; neuropathies or muscle weakness from generalized neuromuscular disease; and hepatomegaly or adenopathy due to esophageal malignancy should be sought. The presence of oral candidiasis suggests fungal infection as a cause of odynophagia. Weight and general nutritional status should be assessed in case dysphagia has resulted in malnutrition.

Differential diagnosis

Esophageal dysphagia

Obstructive esophageal lesions

Esophageal dysphagia is most commonly caused by structural lesions that physically impede bolus transit. Patients with esophageal strictures secondary to acid peptic damage may present with progressive dysphagia after a long history of heartburn. These strictures usually are located in the distal esophagus, but more proximal strictures may develop at the transition point to columnar mucosa in patients with Barrett esophagus. A Schatzki ring, a thin, circumferential mucosal structure at the gastroesophageal junction, causes episodic and nonprogressive dysphagia. Eosinophilic esophagitis should be considered in younger patients who present with intermittent solid food dysphagia or food impaction, particularly in those with a history of allergic or eosinophilic disorders. Patients with esophageal malignancies report progressive symptoms, often starting primarily with solid food dysphagia but progressing to dysphagia for liquids. Relatively recent onset of symptoms and presence of alarm symptoms should prompt rapid evaluation to rule out a malignant etiology. Other mechanical lesions (e.g. abnormal great vessel anatomy, mediastinal lymphadenopathy, and cervical vertebral spurs) can cause dysphagia.

Motor disorders of the esophagus

Primary and secondary disorders of esophageal motor activity represent the other main etiology of esophageal dysphagia. Primary achalasia is an idiopathic disorder characterized by dysmotility of the esophageal body and failure of lower esophageal sphincter (LES) relaxation on swallowing with or without associated LES hypertension. Primary achalasia can be categorized into three types using esophageal manometry. Type 1 achalasia is characterized by poor or absent esophageal peristalsis along with incomplete relaxation of the LES. Type 2 is characterized by pan-esophageal pressurization, and type 3 by spastic esophageal contractions.

Conditions that mimic primary achalasia include secondary achalasia, a disorder with identical radiographic and manometric characteristics caused by malignancy at the gastroesophageal junction or by paraneoplastic effects of a distant tumor, and Chagas disease, which results from infection with

Trypanosoma cruzi. Some patients have esophagogastric junction outflow obstruction, which can be defined by esophageal manometry and may be due to incompletely expressed achalasia or mechanical obstruction. Other spastic esophageal disorders, such as jackhammer esophagus or diffuse esophageal spasm, have also been associated with dysphagia. Many patients with esophageal dysphagia have ineffective motility or fragmented esophageal peristalsis, which are best diagnosed with esophageal manometry. Systemic diseases (e.g. scleroderma and other rheumatic diseases) also cause dysphagia because of reduced rather than spastic esophageal motor function.

Odynophagia

Oropharyngeal odynophagia most commonly results from malignancy, foreign body ingestion, or mucosal ulceration. Esophageal odynophagia usually is a consequence of caustic ingestion, infection (e.g. *Candida albicans*, herpes simplex virus, cytomegalovirus), radiation damage, pill esophagitis, or ulcer disease induced by acid reflux (see Table 1.1).

Diagnostic investigation

Patients who present with complete obstruction with inability to handle oral secretions should undergo urgent upper endoscopy. Contrast radiography is not only associated with an aspiration risk; lesions found on radiography may be obscured by the contrast media. Airway protection is mandatory, so there should be a low threshold for endotracheal intubation or use of an esophageal or gastric overtube.

In the absence of complete obstruction, the history further dictates the next step in investigation. For dysphagia of presumed esophageal origin, upper endoscopy is the test of choice for evaluation and possibly treatment of anatomic abnormalities. Strictures or Schatzki rings can be dilated at the time of endoscopy. If no structural lesion is found, biopsies from the proximal-to-mid and distal esophagus should be taken to exclude eosinophilic esophagitis. If no etiology is identified on upper endoscopy, esophageal manometry should be performed to determine if an esophageal motility disorder is present. Barium swallow radiography can also be helpful to evaluate esophageal transit or to reveal subtle anatomic abnormalities.

Oropharyngeal dysphagia is best evaluated by a video-fluoroscopic swallowing study. Videofluoroscopy of mastication and swallowing of thin liquids, thick liquids, and solids is helpful in examining the coordination of the swallowing process in patients with suspected neuromuscular disease and potentially directing therapy. In some instances, specialized manometry can reveal abnormal upper esophageal sphincter (UES) relaxations. Transnasal or peroral endoscopy also may reveal vocal cord dysfunction but is rarely diagnostic in this setting.

Because mucosal lesions are common, endoscopy is the procedure of choice for odynophagia. If endoscopy is nondiagnostic, esophageal manometry can be useful to identify motility disorders than can cause odynophagia, such as achalasia or diffuse esophageal spasm.

Management

Dysphagia

Selected causes of oropharyngeal dysphagia, including Parkinson disease, hypothyroidism, polymyositis, and myasthenia gravis, may have specific therapies. Surgical myotomy may benefit patients with Zenker diverticulum or cricopharyngeal achalasia. A few limited studies suggest that myotomy also may be useful in treating selected cases of neuromuscular disease. For untreatable neuromuscular conditions, consultation with a speech pathologist may afford development of a rehabilitation program to improve swallowing. Techniques include altering food consistency, motor retraining, controlled breathing, coughing, and head positioning. When adequate nutrition cannot be maintained, alternative routes for enteral feedings such as a gastrostomy may be indicated.

Management of esophageal dysphagia depends on its cause. Benign strictures, webs, and rings can be dilated at the time of upper endoscopy. Rigid bougie dilators or controlled radial expansion (CRE) balloon dilators have equivalent efficacy in treating strictures. Patients with underlying gastroesophageal reflux disease should be treated with intensive acid suppression. Eosinophilic esophagitis may improve with dietary modifications such as the six-food elimination diet or topical steroids. Early malignancies may be surgically resected, whereas palliation via endoscopic stenting or chemoradiotherapy may be used for unresectable lesions. Achalasia can be treated with surgical myotomy or large-caliber endoscopic balloon dilation. Botulinum toxin injection into the LES is used rarely for palliation of symptoms in patients with achalasia who are not candidates for more definitive therapy. The newest endoscopic option for treatment of achalasia is per-oral endoscopic myotomy (POEM). Other primary esophageal motility disorders may respond to nitrates, calcium channel antagonists, and, in rare instances, botulinum toxin or surgical myotomy.

Odynophagia

Therapy for odynophagia secondary to opportunistic infection relies on antiinfective treatments, whereas pill esophagitis and caustic ingestion may be managed with medications such as proton pump inhibitors to reduce acid reflux or slurries to coat the irritated esophagus.

Complications

The most serious complication of oropharyngeal dysphagia is tracheal aspiration, with development of cough, asthma, or pneumonia. Esophageal dysphagia may also result in aspiration, especially in the case of complete obstruction, and with more chronic symptoms in malnutrition and failure to thrive because of reduced oral intake.

Complications from odynophagia are related to the underlying etiology. Bleeding or perforation from esophageal ulceration may occur.

Key practice points

Dysphagia

- Complete esophageal obstruction requires emergency endoscopy with attention to airway protection. Radiographic contrast studies should be avoided due to risk of aspiration and potential interference with subsequent endoscopy.
- Distinguish oropharyngeal from esophageal source with a thorough patient history. Patients with oropharyngeal dysphagia are often co-managed with otolaryngologists or neurologists, depending on the underlying disorder.
- Oropharyngeal symptoms are best evaluated by fluoroscopic imaging, whereas esophageal symptoms are best evaluated initially by endoscopy, followed by manometry to evaluate motility disorders and potentially barium swallow.

Odynophagia

- If a fungal etiology is likely, empirical antifungal treatment is reasonable.
- Endoscopy is the optimal test to evaluate odynophagia.

Case studies

Case 1

A 32-year-old man with a history of asthma presents to the emergency department with six hours of difficulty swallowing. The symptoms began after eating a steak and have progressed to the point where he can no longer swallow his own secretions. He states a prior history of similar symptoms two and six months earlier; both episodes resolved without medical intervention.

Physical examination reveals a healthy-appearing man who is sitting upright with a bucket into which he spits his saliva. No other abnormalities are noted. Laboratory tests are normal.

The patient is intubated. An upper endoscopy is performed, and a large piece of steak is removed from the distal esophagus. Multiple esophageal rings and longitudinal furrows are noted in the esophagus.

The patient is discharged. At follow-up endoscopy one month later, the patient has had no recurrence of symptoms, although he has been careful to cut his food into small pieces and chew his food thoroughly before swallowing. Biopsies show increased intraepithelial eosinophils in the proximal and distal esophagus.

Discussion and potential pitfalls

Esophageal food impaction requires urgent medical intervention. The inability to swallow one's own secretions is a red flag indicating complete esophageal obstruction. In these cases, it is important to avoid contrast radiography, which not only interferes with the endoscopic visualization but is also potentially hazardous due to the risk of pulmonary aspiration. Endoscopic removal of the food bolus should be conducted after ensuring airway protection with an esophageal overtube or by endotracheal intubation.

Eosinophilic esophagitis is commonly diagnosed in young men with a history of atopic disorders and recurrent food impactions. Suggestive features on endoscopy include multiple esophageal rings and longitudinal furrows. The condition can be diagnosed by taking multiple biopsies in the distal and mid-to-proximal esophagus. Patients with possible eosinophilic esophagitis may respond to proton pump inhibitor therapy, and consideration should be given to evaluation for food allergies and treatment with topical steroids.

Case 2

A 67-year-old man is referred to your office for symptoms of dysphagia that have been present for four years, which have not previously been evaluated. He describes progressive dysphagia to solids and liquids, intermittent chest discomfort, and a 15-pound weight loss over the past year. He is otherwise healthy and has no symptoms of heartburn or odynophagia.

Physical examination reveals a thin man whose examination is otherwise normal. Upper endoscopy reveals a dilated, tortuous esophagus. No mass or stricture is noted, and the endoscope passes through the esophagogastric junction with only mild resistance. Esophageal manometry illustrates aperistalsis of the esophageal body with nonrelaxation of the LES, consistent with type 1 achalasia. He is referred for consideration of surgical myotomy.

Discussion and potential pitfalls

Achalasia is classically diagnosed on esophageal manometry by nonrelaxation of the LES with dysmotility of the esophageal body. High-resolution manometry with esophageal pressure topography provides greater diagnostic accuracy and differentiation between the three subtypes of achalasia. The manometric findings of idiopathic achalasia are not specific and can also be seen with pseudo-achalasia or Chagas disease. Barium radiography classically shows a

tortuous or dilated esophagus with a "bird's beak" appearance to the LES. The ganglion cell degeneration found in achalasia is presumed to be immune mediated, as opposed to pseudo-achalasia where infiltration of the esophageal wall by tumor causes obstruction with proximal dilation. In patients who are older and who have relatively rapid progression of symptoms, the diagnosis of pseudo-achalasia as a result of a paraneoplastic process without direct tumor stenosis of the esophagogastric junction must also be considered.

Case 3

A 26-year-old man known to be HIV-positive presents with odynophagia. He has no prior acquired immunodeficiency syndrome (AIDS)-defining diagnosis and has not been taking highly active antiretroviral therapy (HAART). Odynophagia with both liquids and solids has been noted for two weeks leading to a 12-pound weight loss. Physical examination is notable for the absence of oral candidiasis and no abdominal tenderness or masses. Laboratory tests include a CD4 count of 28.

The patient is prescribed oral antifungal medication but has no relief of symptoms after seven days. On upper endoscopy, multiple small discrete ulcers are seen in the mid-to-distal esophagus. Biopsies and viral cultures do not show evidence of herpes simplex virus (HSV) or cytomegalovirus (CMV) infection. HAART is initiated, and the patient improves within five days.

Discussion and potential pitfalls

Immunocompromised patients are at risk of opportunistic infections. Odynophagia in this group of patients is commonly due to fungal infections (*C. albicans*), CMV, and HSV. Empiric therapy with an oral antifungal agent is a reasonable first step. If symptoms do not improve, investigation with upper endoscopy is indicated to evaluate for the presence of the viral infections. Idiopathic ulcerations associated with HIV are also a cause of odynophagia and are diagnosed by the exclusion of evidence of other opportunistic infection on biopsies of the ulcer. Patients may improve with initiation of HAART, but in some cases oral corticosteroids are also necessary for treatment.

Further reading

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