UES Restrictive Disorders

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Why Do I Have Coughing and Choking Right After Eating? Epidemiology and Pathophysiology of UES Restrictive Disorders

Suggested Response to the Patients

Frequent coughing and choking right after eating could be a sign of a swallowing disorder, also called dysphagia. The swallowing process is complex and involves the following different stages: Oral phase refers to sucking, chewing, and moving food or liquid down to the throat; pharyngeal phase is the transport of the bolus down the throat and closing off the airway to prevent food or liquid from entering the airway or to prevent choking; esophageal phase involves propagation food bolus downwards through the esophagus into the stomach due to its rhythmic contraction. An important muscular structure located at the top of the esophagus, called the upper esophageal sphincter (UES), isolates the pharynx from the esophagus. The opening of this sphincter is tightly timed to open when a bolus of food and liquid reach it. The sphincter is normally closed and then relaxes during pharyngeal swallowing and then closes again as the food moves down in the esophagus towards the stomach. Disease conditions that limit adequate opening of the UES during swallowing will result in bolus residue in the pharynx and therefore increase the risk of aspiration of food and liquid into the airway as well as into the nasal passage. If this happens, individuals will experience choking or coughing right after eating or drinking. A number of intrinsic disorders of the UES can cause diminished or failed UES opening, such as Zenker's diverticulum, cricopharyngeal bar, and cricopharyngeal achalasia, causing resistance to bolus flow from the pharynx to the esophagus.

Zenker's diverticulum is an esophageal pouch that forms at the back of throat at the junction of the pharynx and esophagus typically in older patients. The cricopharyngeal bar is a frequent incidental radiologic finding, which in many cases does not cause symptoms. It is present in 5–19% of patients who undergo pharyngeal radiography. Both Zenker's diverticulum and cricopharyngeal bar are related to the fibrosis of the UES that results in diminished compliance and restricted opening of the UES. Increased flow resistance during swallow results in high pressure between the pharynx and esophagus, which facilitates the pouch formation in the area where the muscle is weak. Both conditions are almost uniformly seen in

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E. Bardan, R. Shaker (eds.), *Gastrointestinal Motility Disorders*,
DOI 10.1007/978-3-319-59352-4_7

the elderly. The prevalence of Zenker's diverticulum in the United States ranges from 0.01 to 0.11% of the population. Cricopharyngeal achalasia is a consequence of impaired neural mediated relaxation of the UES. There are diverse causes of cricopharyngeal achalasia, such as stroke, Parkinson's disease, and Alzheimer's disease. The true incidence of the disease is unknown.

Brief Review of Literature

The UES is a complex muscle structure that is composed of the cricopharyngeus muscle (CP), the inferior pharyngeal constrictor muscle, and the proximal cervical esophagus in the pharyngo-esophageal junction [1–5]. It plays an important role in the swallowing process and marks the transition from pharyngeal deglutitive phase to the esophageal phase. Adequate UES opening is therefore essential for an effective swallow. UES opening requires coordination of several factors: UES relaxation, anterior laryngeal traction, UES distensibility, bolus propulsion, and bolus size [1–8]. Failed or diminished UES opening results in incomplete pharyngeal clearance, post-deglutitive residual, and potential post-deglutitive aspiration. Disordered UES opening can be the result of abnormal UES distensibility, such as Zenker's diverticulum, cricopharyngeal bar, or lack of neural relaxation, such as cricopharyngeal achalasia. Alternatively, it can be due to weak pharyngeal propulsion alone or in addition to failed UES relaxation. For this review, we focus on intrinsic UES restrictive disorders.

Zenker's Diverticulum

Zenker's diverticulum is protruding of the mucosa and submucosa through the posterior hypopharyngeal wall at an area of muscular weakness (Killian's dehiscence) between the lower fibers of the inferior constrictor muscle and the upper fibers of the cricopharyngeus. The first case of posterior pharyngeal diverticulum was described by Ludlow in 1767 [9]. Zenker and von Ziemssen did a systematic review of this entity one century later [10]. Since then, this kind of diverticulum was called Zenker's diverticulum. A complete understanding of the etiology of Zenker's diverticulum formation is not available yet. The disease is thought to be related to esophageal motor dysfunction.

It has been reported that the annual incidence of symptomatic Zenker's diverticulum is 2 per 100,000 people per year in the United Kingdom [11]. The prevalence of Zenker's diverticulum in the United States ranges from 0.01 to 0.11% in the population [12]. It is more common in males than females by a ratio of 1.5:1. It rarely occurs in patients younger than 40 and extremely rare under the age of 30. The median age of presentation is in the seventh to eighth decades of life [13, 14]. Congenital pharyngeal pouches have been reported, suggesting that a congenitally weakened Killian's triangle may be a contributing factor in some cases [15, 16]. There is geographic difference of the disease occurrence and it appears to be more common in North America, Northern Europe, and Australia than southern Europe and Asia [17].

Current combined videoradiographic and pharyngeal manometric data support the hypotheses that the formation of Zenker's diverticulum is due to a poorly compliant but normally relaxing UES, which cannot be fully distended during the process of sphincter opening [13, 18]. This leads to abnormal high intrabolus pressure during the phase of trans-sphincteric bolus flow. Pressure imparted to the area of relative muscle weakness (Killian's dehiscence) predisposes to posterior herniation of the pouch over many years [13]. One study compared the cricopharyngeus and inferior constrictor muscle strips in patients with Zenker's diverticulum to controls obtained at autopsy from non-dysphagic individuals. The results showed histologic changes in muscle fibers in Zenker's diverticulum, including increased collagen content, fibroadipose tissue replacement, and fiber degeneration [19, 20]. These morphologic changes in the cricopharyngeus muscle affect contractile and elastic properties of the muscle and account for its restricted opening. In vitro, isolated cricopharyngeus muscle strip from patients with Zenker's demonstrated diminished time to peak twitch, reduced

contractile velocity, and lowered amplitude contractions when compared with controls [21, 22]. The aging process might play a role because of the loss of tissue elasticity and the decrease in muscle tone.

Cricopharyngeal Bar

The cricopharyngeus muscle (CP) is a major component of the upper esophageal sphincter, where it spans between 2.5 and 4.5 cm in length to prevent reflux of gastric contents and allows bolus passage during swallowing [23]. Dynamic function and coordinated relaxation of CP muscle are essential for successful bolus transfer from pharynx to esophagus. Dysfunction of CP muscle encompasses a broad spectrum of clinical manifestations and pathologies. The spectrum of presentation ranges from asymptomatic to severe dysphagia. CP bar refers to the radiographic appearance of a posterior indentation of the esophageal lumen between cervical vertebrae 3 and 6 during barium swallow, partially occluding the lumen of the upper esophageal inlet, and is best visualized in the lateral view [24–26]. It mostly represents an incidental finding on radiographic study and usually does not cause symptom. It is rarely a cause for dysphagia.

A CP bar is present in 5–19% of patients who undergo dynamic pharyngeal radiography [27]. Approximately 13% of these patients have dysphagia [28]. It is almost always seen in elderly subjects [24].

The pathophysiology of the CP bar is not completely understood and several etiological factors have been implicated. CP bar can occur secondary to decreased compliance of CP muscle by fibrosis, incoordination, or congenital weakness of CP muscle [4, 25, 27, 29]. Recent studies of inflammatory myopathy and dysphagia noted increased prevalence of a CP bar and stenosis in patients with dysphagia due to polymyositis or dermatomyositis [30, 31]. CP bar, seen mostly in the elderly, is not a direct result of the aging process, but may be a consequence of the increased prevalence of the neuromuscular disorders or systemic and degenerative processes in the elderly [32, 33]. Investigation by manometry and videofluoroscopy showed normal UES relaxation, normal flow rate across the UES, normal UES resting tone, and hyoid and laryngeal movement in the subjects with CP bar [25, 34]. The major abnormalities in the patients with CP bar are reduced maximal dimensions of UES during the trans-sphincteric flow secondary to decreased passive compliance of UES, and increased intrabolus pressure in the hypopharynx. Thus, the increase in intrabolus pressure preserves normal trans-sphincteric flow rates even though the UES does not open normally [35]. This situation may contribute to the development of Zenker's diverticulum in some patients. Histologic alteration of CP bar from patients undergoing myotomy includes degeneration and regeneration in the muscle fibers of CP with interstitial fibrosis [36].

Cricopharyngeal Achalasia

Cricopharyngeal achalasia (CA) or UES achalasia is a condition characterized by incomplete relaxation of the UES, or by a lack of coordination of the UES opening with pharyngeal contraction. It can arise from intrinsic problems confined to the muscle or from underlying neurologic dysfunction causing high UES tone. The term of CA is somewhat a confusing entity and has been inappropriately used in many instances to describe the radiologic abnormality of incomplete UES opening, such as seen in CP bar. Indeed, manometric studies from pharyngoesophageal segment in CP bar have demonstrated normal UES resting tone and normal relaxation in response to deglutition. There is also no specific finding to correlate with failed UES relaxation in radiography.

The exact incidence of CA is unknown. The lack of epidemiologic data results from the significant controversy regarding the diagnostic criteria required for proper use of the term cricopharyngeal achalasia. The literature reports CA as the primary cause of or as a contributor to dysphagia in 5–25% of patients being evaluated for clinical symptoms of dysphagia [37].

The UES is a skeletal muscle structure and is innervated by excitatory neurons residing in the nucleus ambiguous. The activation of motor

neurons is through the neurotransmitter acetylcholine, by acting on nicotinic receptors at the neuromuscular junction. Resting tone in UES is dependent on tonic input from excitatory neurons. Inhibition of tonic firing of excitatory neurons results in UES relaxation during deglutition. The central generator of swallowing resides within the medulla of the brain stem [38]. During deglutition, normal relaxation of the UES depends on complete and adequate inhibition of muscle tone and accurate coordination with pharyngeal activity in a swallow event. UES relaxation has to occur at a correct time, which is during superior laryngeal excursion and before opening by an average 0.1 s [39]. The UES relaxes during the apogee of UES movement, facilitating the entry of bolus into the UES. Destruction of the neuronal circuit of swallowing, which could involve any of the followings, medullary interneurons, efferent pathways carrying signals away from cortical swallowing centers, and afferent pathways transmitting sensory information to the central generator, may result in UES spasm and impairment of relaxation [3, 5, 39].

Primary CA refers to the abnormality that leads to the persistent spasm or failure of relaxation of the cricopharyngeus muscle that is confined to the muscle, with no underlying neurologic or systemic cause. In many instances, failed UES relaxation is secondary to neurologic disorders such as cortical stroke, lateral medullary stroke, Parkinson's disease, cerebral palsy, amyotrophic lateral sclerosis, myasthenia gravis, Arnold-Chiari malformation, multiple sclerosis, inclusion body myositis, and post-polio syndrome.

Four abnormal patterns of CP activities during deglutition have been observed: (1) incomplete relaxation that blocks the passage of the food bolus into the cervical esophagus; (2) abnormally short duration of complete relaxation; (3) abnormal hypertonic cricopharyngeus during the normal interval of inhibition; and (4) lack of coordination between the pharyngeal propulsion and the cricopharyngeal relaxation [39].

Histologic analysis of surgical specimens in CA patients has shown both striated muscle fibrosis and hypertrophy in the cricopharyngeus muscle [40].

What Are the Symptoms If My UES Does Not Open Normally During Swallowing and How Do You Diagnose It? Clinical Features and Diagnosis of UES Restrictive Disorders

Suggested Response to the Patients

Clinical symptoms for individuals with impaired UES opening vary. Depending on the level of UES restriction and whether or not other contributing factors to dysphagia coexist, e.g., weak pharynx, individuals may be totally asymptomatic to varying degree of difficulty swallowing. Common complaints may include coughing or choking right after eating or drinking, gurgling sound or voice after eating, and extra effort or time needed to chew or swallow. Other symptoms may include regurgitation of undigested food, feeling a lump in the throat, or recurrent pneumonia. As a result, individuals may have poor nutrition, dehydration, risk of aspiration, and chronic lung disease.

The major diagnostic tool is the barium swallow with videofluoroscopy. Individual eats or drinks food or liquid with barium in it, and then the swallowing process is viewed on an X-ray. Endoscopic evaluation of the pharynx and esophagus is to rule out complications and other intraluminal etiologies that may count for or contribute to dysphagia. Esophageal manometry is a tool to evaluate pressure changes that occur during swallowing. It is also used to assess the function of the UES.

Brief Review of Literature

Zenker's Diverticulum

Classical symptoms of Zenker's diverticulum are progressive oropharyngeal dysphagia, and regurgitation (often hours after ingestion) of undigested food debris due to food entrapment in the diverticulum. Eighty percent of the patients have complained of regurgitation of undigested food [22, 41]. Patients may present with chronic cough, chronic aspiration, foul breath, audible gurgling in the throat, sensation of a lump in the

throat, and hoarseness. Weight loss can happen in patients with long-standing dysphagia. The duration of symptoms prior to presentation varies from weeks to many years.

Squamous cell carcinoma complicating a pouch has been reported with an incidence between 0.4 and 1.5% [42, 43]. Chronic inflammation due to food stasis may attribute to the malignant changes. Malignancy should be suspected if there is a sudden change in the severity of symptoms or development of alarm symptoms (hemoptysis, hematemesis, or local pain). Other rare complications include bleeding [44, 45], benign ulceration of the mucosa within the pouch probably secondary to acid reflux or aspirin use [46], bezoar formation, and fistula formation. Due to the risk of perforation during endoscopy or passage of nasogastric tube in patients with known Zenker's diverticulum, it is advisable to intubate the esophagus under direct visualization.

The mainstay diagnosis is the barium swallow with videofluoroscopy. This dynamic study provides information about the size and location of the Zenker's diverticulum. In addition, it can help to detect pharyngeal dysfunction that might contribute to the patient's dysphagia. The esophagus should also be carefully examined in the radiographic study since coexistent pathology might account for the patient's dysphagia or regurgitation. Endoscopic techniques have limited diagnostic capability, as the opening of the pouch is not always apparent endoscopically. If a constant filling defect is seen radiographically, endoscopy is needed to rule out malignancy. Esophageal manometry is usually not required.

Cricopharyngeal Bar

Most of the time, CP bar is an incidental finding on pharyngeal radiography. It usually does not cause any symptoms, but when it becomes symptomatic, oropharyngeal dysphagia is the most frequent complaint. Depending on the swallow function, symptoms can vary from diet modification and/or prolonged mealtime to cough, aspiration, weight loss, or non-oral feeding. The CP bar is more frequently associated with dysphagia when there is a marked obstructive bar with narrowing of the UES lumen [47], when a Zenker's diverticulum is present, or when the patient has current pharyngeal weakness [25].

The diagnosis of CP bar includes videofluoroscopic, endoscopic, and manometric evaluation. CP bar is seen in the barium swallow as a posterior indentation in the barium column between cervical vertebrae 3 and 6 that persists throughout the swallow [24]. Recent interest in high-resolution manometric study of the UES and pharynx has improved our understanding of the motility alteration in CP bar. Manometry is not an essential for diagnosis, but will show an increase in intrabolus pressure suggesting increased flow resistance [48]. The UES relaxation and pharyngeal contraction are normal. A CP bar is difficult to appreciate on endoscopic examination; however, endoscopic evaluation is essential to rule out malignancy or other causes of dysphagia.

Cricopharyngeal Achalasia

The clinical presentation of CA is nonspecific and quite variable. Symptoms may have an abrupt or gradually progressive onset going on for months or years. Most patients complain of food sticking or catching in the lower part of the neck. Solid dysphagia seems more common than liquid dysphagia. "Stringy" foods like noodles or vegetable leaves seem to be particularly challenging [39]. Patients may also experience heartburn, choking, and odynophagia. Less common symptoms include dysphonia, globus sensation, and pressure in the neck during deglutition. Pulmonary symptoms like aspiration pneumonia usually result from aspiration of ingested food retained in the hypopharynx above a non-relaxing UES. In severe dysphagia, weight loss, starvation, and dehydration could occur.

Videofluoroscopic swallow remains the mainstay for diagnosis in the patients with symptoms suggestive of CA. It can demonstrate reduced opening of the pharyngoesophageal segment and dilated pharynx with holdup of the contrast bolus. Videofluoroscopy can also detect other disturbances in function, such as abnormal tongue strength or movement, impaired hypolaryngeal elevation, nasopharyngeal regurgitation, or aspiration. However, as mentioned before, there is no

specific radiologic finding indicative of failed UES relaxation. Besides CP relaxation, UES opening also relies on anterior laryngeal elevation, UES distensibility, bolus propulsion, and bolus size. Any or combined abnormalities of these conditions can result in impaired UES opening and hypopharyngeal bolus retention.

When appropriately utilized, esophageal manometry can be helpful in the diagnosis of CA by demonstrating impaired UES deglutitive relaxation and inappropriate contraction during the normal period of motor quiescence [49]. It can assess the coordination of UES relaxation with hypopharyngeal contraction during swallow. Typical manometric findings in CA include elevated deglutitive UES nadir pressure, reduced interval of UES relaxation, and elevated hypopharyngeal intrabolus pressure. One shortcoming of the manometric study is that it cannot assess for the presence of many other conditions that can cause symptoms similar to CA; therefore, it is not sufficient for diagnosis of CA by itself without additional information from radiologic study to rule out other causes.

Endoscopic evaluation is generally not helpful in the diagnosis of CA. In some occasions, tight entrance to the esophagus at the level of UES may raise the suspicion of CA; however, this finding is nonspecific. The main role of endoscopy is to rule out other conditions that may cause similar symptoms.

What Are the Treatment Options Available? Therapy for UES Restrictive Disorders

Suggested Response to the Patients

Treatment depends on the symptom and cause of the swallowing problems. There is no treatment required for asymptomatic patients. Mild dysphagia could be managed by modifying diet, avoiding food that causes problems, or changing the consistency of the diet. In individuals with Zenker's diverticulum, management depends on the local expertise, patient's age, and size of the diverticulum. Intervention could be open surgical repair or endoscopic repair. The latter has been increasingly adopted as a main treatment option among otolaryngology specialists in the United States since it is proven to be less invasive and has similar efficacy compared to surgery. Other treatment options for UES restrictive disorders include endoscopic dilation, botulinum toxin injection, and surgical myotomy. The purpose of these treatments is to relieve the UES obstruction. Endoscopic dilation and botulinum toxin injection are effective treatment options, but may need to be repeated at different intervals to achieve long-term effect.

Brief Review of Literature

Zenker's Diverticulum

Zenker's diverticula require intervention only if they produce symptoms. Small asymptomatic diverticula do not need treatment, as the risk of severe adverse complications, cancer, and aspiration is low. Open surgeries, which include CP myotomy alone, diverticulectomy, diverticulopexy, or diverticular inversion, all with or without current CP myotomy, have long been the conventional treatments with a high success rate, but are associated with high morbidity and mortality [50–54]. Since Zenker's diverticulum mainly affects elderly patients accompanied by multiple comorbidities, less invasive treatments are favored. In recent years, endoscopic repair of Zenker's diverticulum has been found to be a viable safe and effective alternative to surgery and gained widespread acceptance. When compared to open stapler-assisted diverticulectomy and CP myotomy, endoscopic staple-assisted diverticulostomy (ESAD) are associated with shorter operative times, shorter postoperative hospital stays, quick resumption of oral intake, and few complications, such as recurrent laryngeal nerve injury and bleeding [54, 55]. In many centers, EASD is performed as an outpatient procedure in appropriately selected patients. Flexible endoscopic approach consists of cutting

the septum between the diverticulum and the esophageal lumen, as the septum contains part of the cricopharyngeal mask [56–58]. The objective is to create a common room between the sac of the diverticulum and the esophagus, so that food can pass more easily into the esophagus. In the meanwhile, it helps to reduce the local pressure of the cricopharyngeal muscle. It has been reported that symptom relief or improvement was achieved in 89–96% of patients under EASD with recurrence ranging from 0 to 9% [55]. Factors that most often precluded a successful endoscopic approach were a patient's inability to open their mouth fully, extend their neck completely, or a shallow diverticula sac (<3 cm) that precludes full engagement of the entire CP muscle in the common wall by the stapler [55].

Cricopharyngeal Bar

The goal for treatment of a symptomatic CP bar is to increase the UES diameter during swallowing. If the CP bar does not cause symptoms or the bar is not the culprit for dysphagia, there is no need to treat it. Treatment options include endoscopic dilation, botulinum toxin injection, and surgical myotomy. Since this patient population is usually elderly with multiple comorbidities and high risk for perioperative complications, nonsurgical interventions are more preferred than surgical treatments. Though botulinum injection and CP dilation have been reported to be highly effective and safe, CP myotomy has remained as the gold standard treatment of CP bar.

Botulinum toxin A injection to the cricopharyngeus muscle under direct vision has been utilized since 1994 with success rate ranging from 43 to 100% [59]. Repeated injections are often necessary to achieve or maintain a good effect. Botulinum toxin injection works best in patients with impaired relaxation of the CP muscle, and is partially or not effective in structural stenosis of UES caused by persistent hypertrophy or restricting fibrosis, which is usually the case in patients with CP bar [25]. Diffusion of the toxin to adjacent muscle may worsen dysphagia or cause vocal cord dysfunction. Controlled trials are needed to determine the safety and efficacy of the use of botulinum toxin.

Dilatation of the cricopharyngeus muscle may be performed using either a balloon dilator or bougie dilator (Savary–Gilliard dilator). Both techniques have been broadly used clinically and proved to be safe and effective, although half of the patients experienced short-term recurrence and required repeated dilation over many years in order to maintain symptomatic improvement [34, 60–62]. It has thus been suggested that dilatation might be used as a first-line intervention, prior to more definitive management. Balloon dilation of the UES is a low-risk option that serves best in patients with fibrosis of the CP, which is usually the case in patients with CP bar.

CP myotomy has been a traditional treatment option for CP dysfunction. It helps to normalize UES opening and may improve pharyngeal contraction [63, 64]. It serves best in patients with structural UES disorders that constrict its opening, such as CP bar and Zenker's diverticulum. UES opening showed better improvement with CP myotomy than with dilation or botulinum toxin [64]. The success rates range from 50% in patients with dysphagia secondary to neurogenic etiologies to 98% in Zenker's diverticulum [65, 66]. In a report by Dauer et al., including 14 patients that underwent CP myotomy, 5 of 7 with idiopathic CP bar were completely asymptomatic postoperatively, while all of the 3 patients with concomitant systematic neurologic disorders had postoperative Functional Outcome Swallowing Scale (FOSS) score greater than 3 [67]. CP myotomy can be performed as open or endoscopic approach. The two techniques have a similar success rate, but endoscopic CP myotomy is associated with shorter operative times, more rapid postoperative recovery, and lower risk of major complications.

Cricopharyngeal Achalasia

Dietary modifications are usually the initial step if the symptoms are provoked by certain foods. These foods should be avoided or their consistency modified. For patients with solid and pill

dysphagia, liberal use of liquid wash may stimulate sensory input that drives the swallow central pattern generator toward more normal function.

Various forms of therapies have been employed for CA. Mechanical dilation of the CP muscle through balloon or tapered bougies has been proven to be safe and effective in all age groups. Objective responses to such therapy include improved pharyngoesophageal segment opening on videofluoroscopy and reduction in basal UES pressure. In adults, dilation diameters 16–20 mm usually result in immediate symptom improvement, though some require repeated dilation at varying intervals [39]. Potential risks of dilation include perforation or bleeding in the pharynx or esophagus; however, these risks can be minimized by appropriate videofluoroscopy or endoscopic pre-dilation assessment.

Botulinum toxin injection was reported to provide temporary relief of symptoms. By diminishing acetylcholine levels, the toxin interferes with nerve impulse transmission and causes flaccid paralysis of muscles [68]. The injection into the cricopharyngeus muscle can be via endoscopy or percutaneous EMG-guided needles. Onset of subjective benefit is usually by day 7, with duration of benefit varying by 3–4 months [39]. Repeated injection is usually required to maintain the effect. Botulinum toxin injection has better symptomatic response in isolated cricopharyngeal dysfunction without other impairment in the swallowing mechanism. Complications from botulinum toxin injection are low, but there is a risk of spreading the toxin into adjacent muscles, which could result in paradoxical worsening of dysphagia or aspiration.

Surgical treatment with cricopharyngeal myotomy is the curative care of CA. Mechanical division of the cricopharyngeus muscle essentially alleviates the symptoms caused by tonic contraction of the UES. The procedure can be performed via an open (transcervical) or endoscopic approach. The potential risk for complications is higher than that of nonsurgical approaches, including infection, hemorrhages, inadequate myotomy, recurrent laryngeal nerve injury, and pneumonia. Outcomes of cricopharyngeal myotomy tend to be poor when significant pharyngeal weakness is also present.

What Are the Other Conditions That Could Be Confused with UES Opening Dysfunction? Disorders That Need to Be Distinguished from UES Opening Dysfunction

Suggested Response to the Patients

Structural abnormalities of the esophagus just below the level of the UES or in the proximal esophagus, such as esophageal webs or rings, may cause dysphagia that needs to be distinguished from impaired UES opening. Barium swallow and endoscopic evaluation can help to differentiate the diagnosis. Treatment option for proximal esophageal webs and rings is mechanical dilation.

Brief Review of Literature

An esophageal web is a thin, non-circumferential membranous tissue covered with squamous epithelium that protrudes into the lumen. Esophageal webs could be congenital or acquired. The congenital webs are usually located in the middle or distal esophagus, while acquired esophageal webs most commonly occur anteriorly in the cervical esophagus below the cricoid, causing narrowing of the esophageal lumen. The prevalence of cervical webs in patients undergoing barium swallow studies is reported to be 5.5–8% [69, 70]. It appears to predominantly affect white individuals and mostly in female patients [71]. It can occur in all age groups. Esophageal webs associated with iron-deficiency anemia, glossitis, koilonychia, and esophageal or pharyngeal carcinoma are known as Plummer-Vinson syndrome or Paterson-Kelly syndrome [71]. Esophageal webs have also been reported to be associated with extracutaneous manifestations of bullous dermatologic disorders such as epidermolysis bullosa [72], bullous pemphigoid [73], pemphigus vulgaris [74], and immunologic disorders in chronic graft-versus-host disease [75], as well as Zenker's diverticulum [76] and gastroesophageal reflux disease [77].

Most patients with cervical webs are asymptomatic. In symptomatic patients, the characteristic complaint is solid food dysphagia. The

severity of dysphagia is directly related to the luminal obstruction. Some patients may even present with acute food impaction. Other complaints include nasopharyngeal reflux, aspiration, and spontaneous perforation. The esophageal webs are usually diagnosed by barium swallow and upper endoscopy. A frequent videofluoroscopic finding is that impaired transit of a swallowed tablet or marshmallow at a subtle narrowing site of the post-cricoid region coincides with subjective experience of dysphagia. Endoscopic diagnosis of esophageal webs can be difficult because the proximal location of a web makes it difficult to detect. An esophageal web appears as a smooth, thin membrane that is eccentric under endoscopic examination [71].

Asymptomatic esophageal webs do not require any intervention. For patients with mild dysphagia, the initial step is diet modification to avoid certain foods that can trigger symptoms. Lifestyle modification including cutting food into small pieces and chewing carefully can help to eliminate symptoms. Mechanical dilation with through-the-scope balloon dilator or a large bougie dilator can be used to rupture the ring. In patients with underlying medical conditions such as iron-deficiency anemia or chronic graft-versus-host disease, treatment should be aimed at the underlying medical condition after dilatation.

An esophageal ring is defined as a concentric, smooth, thin extension of mucosa or muscular structure. It can be found anywhere along the esophagus, but the most common location is in the distal esophagus, such as Schatzki ring. The pathogenesis of esophageal ring is related to acid exposure and eosinophilic esophagitis (EoE) [78, 79]. The clinical presentation is typically solid food dysphagia. The diagnosis can be made with barium esophagram and endoscopy. The treatment is mechanical dilation combined with acid-suppressive treatment. One of the differential diagnoses of proximal esophageal rings is eosinophilic esophagitis (EoE). Endoscopic findings that suggest EoE include stacked circular rings, linear furrows, whitish papules, and small-caliber esophagus [80, 81]. Esophageal biopsies should be obtained to confirm the diagnosis. The treatment of EoE involves dietary, acid suppression, tropical steroid, and mechanical dilation. The details of EoE are presented in a separate chapter in this book.

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