EXTERIORIZED GIANT ZENKER`S DIVERTICULUM: CASE REPORT

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ABSTRACT

Zenker's diverticulum (ZD) also known as cricopharyngeal (CP), pharyngoesophageal or hypopharyngeal diverticulum is an outpouching of mucosal and submucosal layers originated from the pharyngoesophageal junction. Radiological evaluation of deglutition, modified barium swallow radiography may reveal the retentive character of the diverticular sac and confirm the diagnosis. Many different types of surgical approaches have been developed over the decades with modifications focusing on the need to reduce intra-operative or post-operative complications and overall morbidity. The authors present the case of a 76-years-old woman with spontaneous apparition of an exteriorised tumor in the oral cavity after an excessive cough effort. Surgical treatment is elective, preventing the occurrence of complications that may endanger the patients life and improve the quality of life. Surgical therapy was open neck resection of the Zenker's diverticulum with esophageal myotomy and pharynx reconstruction.

Keywords: Zenker's diverticulum, diverticulectomy, myotomy, opentechique, surgery

Introduction

Zenker's diverticulum (ZD) also known as cricopharyngeal (CP), pharyngoesophageal or hypopharyngeal diverticulum is an outpouching of mucosal and submucosal layers originated from the pharyngoesophageal junction. It is an acquired herniation through the posterior wall of the pharyngoesophageal junction known as Killian's triangle, which is a weak point formed between the horizontal fibers of the cricopharyngeus muscle and the oblique fibers of the inferior pharyngeal constrictors, also known as Killian's dehiscence. Being formed on the dorsal wall of the hypopharynx it is more

correctly classified as a pseudo diverticulum (1).

ZD was first observed by Abraham Ludlow MD, during an autopsy in 1769, who described an abnormal dilation of the posterior wall. A more detailed description was made in 1877 by Zenker and Von Ziemssenn, german pathologists which characterized this disease and elucidated that the diverticulum formed due to “forces within the lumen acting against restriction”. Killian's triangle was described by the renowned surgeon Gustav Killian in the early 1900's (2).

Over time, different therapies for Zenker diverticulum were imagined. In 1796 Ludlow had made the first description of the surgical treatment, but was reported by Weeler in

1882. In 1912, Schmid communicated the diverticulopexy, with lower morbidity and mortality compared with the resection. In 1917, Mosher had introduced the technique that uses a rigid endoscope, with the section of the septum of the diverticulum, which was later improved by Dohlman in 1960 and Collard in 1993, as the performances of the endoscopes got better. In 1958, Harrison added myotomy in the surgical protocol of Zenker diverticulum (3,4).

Zenker classified esophageal diverticula, in general, in pulsion and traction diverticula. This entity was categorized after Negus in 5 evolutive stages: stage I: mucosal protrusion; stage II: horizontal evolution; stages III, IV: parallel evolution with the esophagus, which is pushed to the anterior; stage V: retroclavicular evolution in the mediastinum;

Pathophysiology

It is believed that poor upper esophageal sphincter compliance is the presumed pathophysiologic mechanism of action. While important debate exists with respect to the mechanism of ZD development, the most implicated phenomena is the dysfunction of the cricopharyngeal muscle and incoordination of the inferior pharyngeal constrictor muscle, leading to an increased intraluminal pressure, resulting in a gradual, progressive herniation in a relative weak area of muscle when compared with surrounding areas (5). Cook et al. suggested the CP dysfunction occurs due to muscle fibres degeneration and replacement with fibroadipose tissue compromising upper esophageal sphincter (UES) elasticity, sphincter opening, and the generation of elevated pressure gradient in the hypopharynx (6,7).

Several studies revealed the relationship between gastro-esophageal reflux disease (GERD) and Zenker diverticulum. Some authors consider that the acid reflux can theoretically ascend to the pharyngeal level determining mucosal injury at the Killian triangle site and cricopharyngeal muscle hypertrophy (8,9,10).

Symptoms

Zenker's Diverticulum has a preponderance in affecting males more often than females, with a 1.5:1.0 ratio. The annual incidence has been estimated at 2 per 100.000. This rare condition most commonly presents itself in the seventh and

eight decade of life and can be associated with various symptoms. The duration of symptoms may vary from a few weeks to years. The leading symptom is represented by the progressive oropharyngeal dysphagia, occurring in more than 90% of patients. There are 2 mechanism by which the diverticulum may cause dysphagia: incomplete opening of the upper extrinsic sphincter and extrinsic compression of the cervical esophagus by the diverticulum itself. In the early stage of ZD development transient dysphagia may be noted (11). Other symptoms may include regurgitation of undigested food, halitosis. These symptoms may occur when the diverticular sac becomes large enough to retain contents such as mucus, sputum and food. Gurgling in the throat, dysphonia, noisy swallowing, coughing, hoarseness, chest pain, globus sensation, recurrent respiratory infections and weight loss, small, depressable swelling on the left side of the neck known as Boyce sign, may occur depending on the severity of dysphagia. Cervical borborygmus, particularly in the presence of a palpable mass in the neck is the most pathognomic of ZD (12). Clinical examination with inspection, palpation and auscultation are often normal in patients with ZD.

Diagnosis

It is well known that imaging studies are the primary means of diagnosis of ZD. This pathology may be considered based on the history of dysphagia and regurgitation. Radiological evaluation of deglutition, modified barium swallow radiography may reveal the retentive character of the diverticular sac and confirm the diagnosis. Flexible endoscopy may reveal the characteristic of mucosa but it is not indicated as you can accidentally enter and perforate the diverticular pouch. Manometric pressure recordings reveal abnormalities of the upper esophageal sphincter during swallowing. Ph-metry reveals the correspondence with GERD, Barret's disease. ZD may also be diagnosed by transcutaneous ultrasonography while ingesting water, making the diverticular sac increase in size, reducing the definition of its margins, and showing the heterogeneous ecogenity of the sacular contents. The ultrasound findings help differentiate ZD of a thyroid mass. Although the

diagnosis is considered based on patients' history, dysphagia and regurgitation and it is confirmed by esophageal studies (13,14,15).

Management

The elective and curative treatment for DZ is surgery. For the asymptomatic patients surgery is not necessarily needed, never the less long term monitoring is indicated.

Many different types of surgical approaches have been developed over the decades with modifications focusing on the need to reduce intra-operative or post-operative complications and overall morbidity. Surgical treatment of ZD can be classified by the approach as either open, via an external neck incision, or endoscopical, via the mouth. Current approach is considered to be transoral rigid endoscopy with open cricopharyngeal myotomy. This has become the most popular treatment method for Zenker's diverticulum due to the fact that it is less invasive, shortens hospital stay, decreases anesthesia time and has fewer complications.

External approach consists of four different types of surgical procedures. In some cases a two stage surgery may be needed if there is granulation tissue formation in which the excision of the on-forming ZD surrounding tissue may be needed. Other techniques include single step surgery for ZD removal, CP myotomy without the excision of the ZD and CP myotomy with the excision or suture of ZD (16-19).

Complications

In the case of untreated ZD the most common complications are ulceration and perforation of the diverticular sac, upper gastrointestinal bleeding, diverticulum infections, endobronchial fistulas, aspiration pneumonia. ZD may associate esophageal carcinoma.

Postoperative complications can be immediate such as hematoma, recurrent laryngeal nerve lesions, faringo-cutaneous fistula, mediastinitis, and late such as reappearance of dysphagia and lack of improvement in symptomatology (20).

Case Report

A 76-years-old woman from urban area was brought by ambulance from home to the ENT emergency room with spontaneous apparition

of a tumor in the oral cavity after an excessive cough effort.

The patient experienced anxiety, was not cooperative, denied smoking or alcohol consumption, and also denied any preceding symptoms including dysphagia or weight loss. Personal pathology history includes high blood pressure and no surgical procedures prior to the presentation in the emergency room.

At presentation, physical examination revealed a sacciform tumoral mass of approximately 20 cm exteriorized in the right corner of the mouth surpassing it with about 7 cm (Figure 1). Larynx was normal conformed, vocal cords with normal mobility and normal glottic space. Other clinical findings of the head and neck region were unremarkable. Blood tests were unremarkable. A pharyngolaryngeal fibroscopy revealed the possible origin of the sacciform tumor in the esophageal mouth. CT scan of the neck and thorax revealed that the esophageal diverticulum was located on the right side of the wall of the esophagus, immediately above the upper thoracic aperture with a narrow parcel of approximately 0.7 cm with intracavitary flapping in the esophagus, pharynx and mouth. No other pharyngeal-esophageal structures appeared on CT scans sections with morphology or pathological significance.

The patient was informed about the therapy options and surgical benefits and risks were explained. Informed consent was signed by the patient.

Surgical therapy was preceded by endoscopic examination of the esophagus to evaluate the point of origin of the ZD and it was followed by cricopharyngeal myotomy with diverticulectomy through right cervicotomy.

The patient was placed in the supine position with the head tilted towards left and underwent general anesthesia with oro-tracheal intubation. The rigid esophagoscope was inserted and the eviscerated exteriorized diverticulum was revealed with the base of insertion at 15 cm from the upper dental arc. The endoscope was fixated in this position. A right latero-cervical incision was made at the anterior margin of the sternocleidomastoid muscle. Subplatysmal flaps were elevated, deep cervical fascia was incised just anterior to the sternocleidomastoid

and was extended several centimeters above and below the incision. The sternocleidomastoid muscle was retracted laterally and the omohyoid muscle was exposed and divided. Dissection was carried down ventrally to the carotid sheath, which was retracted laterally, while the thyroid gland and the larynx were retracted medially.

The diverticulum implantation base was highlighted by light coming from the esophagoscope at this moment and a lateral esophagotomy was performed with the traction and inversion of the diverticulum, followed by diverticulotomy at its implantation base (Figure 2). A myotomy of the cricopharyngeal muscle was performed prior to the diverticulum resection. The diverticulum was sent for pathological examination (Figure 3). The nasogastric tube was fitted and the hypopharynx defect was repaired followed by the suture of anatomical plans (Figure 4).

Post-operatively the patient received antibiotic, antiinflammatory and antalgic therapy and evolution was favorable. Nasogastric tube was suppressed five days after the surgery. We performed a contrast radiography which showed a normal anatomical esophagus. 10 days postsurgery the skin sutures were removed. The result of the histopathological examination confirmed the diagnostic of Zenker's diverticulum with no other evidence of malignancy. Larynx and pharynx were inspected with flexible fiberoptic laryngoscopy and did not reveal abnormalities regarding vocal fold motion, pharyngeal hematoma or edema.

Figure 1. Sacciform tumoral mass of approximately 20 cm exteriorized in the right corner of the mouth surpassing it with about 7 cm

Figure 2. Lateral esophagotomy with diverticulotomy at its implantation base.

Figure 3. Excised diverticulum sent for pathological examination.

Figure 4. Hypopharynx and anatomical suture.

Discussions

Discussions need to be made regarding the particularities of the case since there were some atypical manifestations of ZD. The presented case is of a rare case of giant bilobate Zenker's diverticulum, exteriorized in the right corner of the mouth due to prolonged coughing effort. The diverticulum of the pharyngeal-esophageal junction is a pulsation diverticula formed at a weak point of the pharyngeal muscles, which in exceptional cases can be exteriorized. Even if the literature describes a preponderance in males our patient was female.

We want to emphasize the need for emergency treatment given by the life-threatening potential. Surgery consisted of a two-step approach, first rigid endoscopy followed by diverticulotomy with miotomy by open technique. Choosing the surgical technique is very important and directly related to the diverticulum stage. This must be communicated to the patient during the pre-therapeutic discussion. Also, the intra-operative and post-operative complications and risks must be explained to the patient and the patient must understand and agree by signing the informed consent.

Open approach through right cervicotomy was more difficult considering that it is usually preferred to approach ZD through the left cervicotomy. The incision is usually placed in the left side of the neck for several reasons, including the relatively straighter, possibly more stretch-resistant left recurrent laryngeal nerve compared with the more horizontal placed right recurrent laryngeal nerve, as well as the gradual tracking of the cervical esophagus from the center to the left as it descends into the lower neck and upper chest (21).

Conclusions

Surgical treatment is elective, preventing the occurrence of complications that may endanger the patient's life and improve the quality of life. Several techniques have been used in the management of ZD. However, the right one to choose depends on the stage of the diverticulum, size, morphological impairment in endoscopic

approach, surgical team expertise and factors linked to the general status of the patient, along with the informed consent.

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Conflict of interest

The authors declare that there is no conflict of interest.

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