The esophageal rapunzel syndrome: a case of a trichobezoar in the esophagus

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Abstract

Rapunzel syndrome is a form of trichobezoar, a rare form of bezoar, especially seen in individuals with hair pulling (trichotillomania) and hair eating (tricophagia) habits, that extends from the pylorus into to the duodenum, jejunum and even the colon. We report the case of a 37-year-old woman with a trichobezoar in the esophagus, causing esophageal rupture that required an urgent surgical intervention.

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Introduction

Bezoar is the name given to the masses formed by the accumulation of an indigestible substance anywhere in the gastrointestinal tract. Bezoars are called according to the substances they contain, such as phytobezoars (vegetable or fruit fibres), lactobezoars (milk and its products), trichobezoars (hair), diospyrobezoars (dates), pharmacobezoars (especially ferrous solfate and antiacid drugs), lithobezoars (pieces of stone), plasticobezoars (plastics) and others (sand, gum, mushroom, *etc.*). The most common form is phytobezoar.[1] Trichobezoar is a rare form of bezoar casused by the accumulation of swallowed head and other body hairs in the gastrointestinal tract, most commonly seen in individuals with hair pulling (trichotillomania) and hair eating (tricophagia) habits. Rapunzel syndrome was first described by Vaughn in 1968,[2] and is defined as the extension of the trichobezoar from the pylorus into the duodenum, jejunum and even the colon, even if the most common site in the stomach.[3]

Based on the current literature, trichobezoars in the esophagus are very rare, and isolated esophageal trichobezoars are uncommon, generally related to tube feeding or esophageal disorder (achalasia, epifrenic diverticulum). We report the case of a patient admitted to our hospital for abdominal pain with a voluminous trichobezoar in the esophagus, that required an urgent partial esophageal resection after three unsuccessfull endoscopic procedures, complicated by esophageal rupture in the last one. We conied the definition *Esophageal Rapunzel syndrome* for this case.

Case Report

A 37-year-old female patient was admitted to our hospital with complaints of abdominal pain and loss of wieght in the last year. She reported a history of recurrent abdominal pain and diarrhea, treated with symptomatics. She also referred the habit of eating her hairs (tricophagia) since the age of 7, that required an esophagogastoduodenoscopy (EGD) for the removal of a trichobezoar in the esophagus 4 months ago. She has never consulted a psychiatric for her mental disorder. On admission, she denied nausea and vomiting. The abdomen was not distended. Rebound tenderness, and abdominal guarding and rigidity were not detected. Laboratory tests were all within the normal limits. Three consecutive EGDs were performed trying to remove the trichobezoar seen in the gastroesophageal junction. Unfortunately the last procedure was complicated by the esophageal rupture. An urgent computed tomography (CT) scan of the thorax and abdomen showed a right pleural effusion, a right pneumothorax, and a mass lesion at the gastroesophageal junction that enlarged the esophagus (Figure 1). A right chest tube was immediately placed and an

empiema was diagnosed that required 4 antibiotics. The patient drank methylene blue solution, and a leakage from the chest tube was observed which required an urgent thoracotomy. Exploration revealed a perforated esophagus with an approximately 8 cm lesion extending to the esophagogastric junction in the distal part of the esophagus (Figure 2A-B). Gastric skeletonization by laparotomy and partial esophageal resection by right thoracotomy were performed. The operation was terminated after performing an end-to-side esophagogastrostomy. Approximately 8 cm-long trichobezoar was removed (Figure 2C-D). On the 6th postoperative day, chest tube, abdominal drains and nasogastric tube were removed. The patient was discharged in good clinical condition 14 days after the surgical intervention.

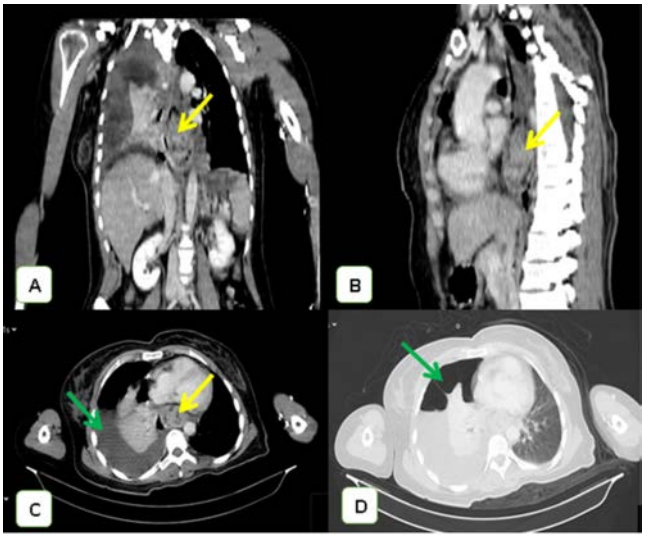

Figure 1. CT scan of the thorax and abdomen. Coronal (A), sagittal (B) and mediastinal window (C) view showing the trichobezoar (yellow arrow), the empyema (C) and the pneumothorax (D) (green arrow).

Discussion

Trichotillomania is currently considered as a type of impulse control disorder[4] followed by swallowing the hair (trichophagia), that can have severe consequences including surgical emergencies.[5,6] As a consequence, early diagnosis and prompt management are crucial.

Trichobezoar occurs when a trichotillomanic patient eats his or her hair after extraction leading to surgical emergency.[7,8] Rapunzel syndrome is a rare form of trichobezoar generally resulting from psychiatric disorders like depression, anxiety, obsessive-compulsive disorder and body dysmorphic disorder,[9,10] that always require a psychiatric referall.[11] In addition, causes such as mental retardation and family stress can be considered as risk factors for trichobezoar formation.[12] The prevalence is unclear. Depending on the case series, the symptoms often begin in the twenties and it is more common in women.

Due to the slow growth rate of the trichobezoar, the mass of the bezoar increases gradually, and clinical symptoms appear late in most of the cases. The most common symptoms and findings detected are palpable abdominal mass (88%), followed by abdominal pain (70%), nausea-vomiting (63%), weight loss and fatigue (39%), constipation or diarrhea (32%), and rarely hematemesis (6%).[13] Low level of hemoglobin has been reported in about 62% of the cases.[14] The diagnosis of trichobezoars is based on imaging evidence. The gold standard technique is EGD.[15] Plain radiographs can usually show a non-specific mass. Ultrasound can show the echogenicity of trichobezoars. CT of the abdomen is an effective imaging method to clearly reveal the presence, location and distribution of bezoars. Endoscopy gives the definite diagnosis. Although barium radiographs are used as an imaging method, they are not recommended as they may cause obstruction or perforation.

Serious complications may be encountered when the diagnosis of trichobezoar is delayed. The most common complication is perforation of the stomach and small intestine. Invagination, pancreatitis, intestinal obstruction, hematemesis, gastric ulcer, obstructive jaundice and peritonitis are possible but rare complications.[16] In the case of untreated trichobezoars, a mortality rate up to 30% has been reported, while perforation and peritonitis have been shown as the causative factors.[17]

Treatment includes surgical removal of the bezoar and professional psychotherapy.[9] While endoscopic treatment can be performed for small-sized ticobezoars, the recommended treatment for the management of cases with large trichobezoars is removal of the mass by laparotomy or laparoscopy, according to the facility and experience.[18] However, treatments such as extracorporeal shock wave, lithotripsy, intragastric application of enzymes (pancreatic

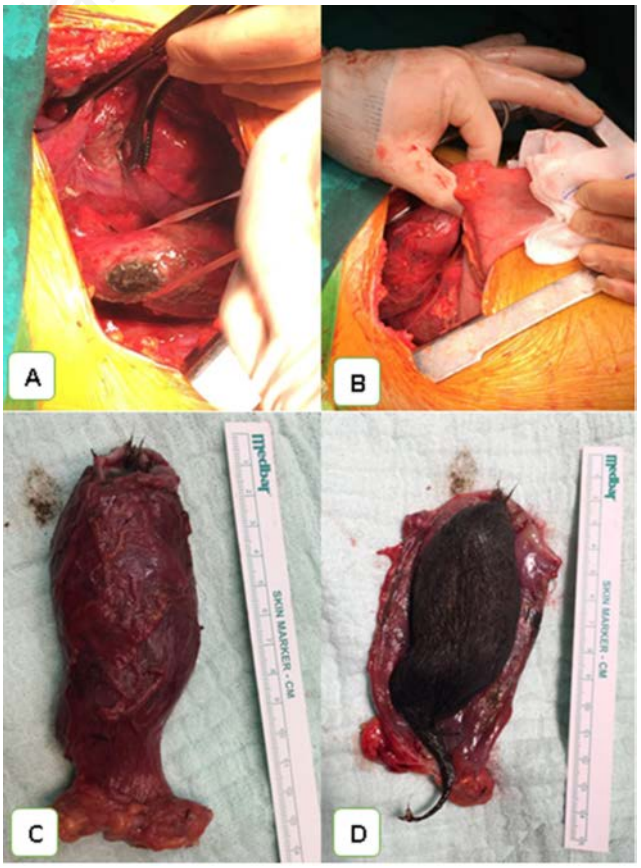

Figure 2. Intraoperative extraction of the trichobezoar from the distal esophagus. Perforated esophagus (A), isolated trichobezoar (B), resected esophagus (C), trichobezoar (D).

lipase, cellulose) and drugs (metoclopramide, acetylcysteine) may provide varying rates of treatment success.[19] Endoscopy is not recognized as an effective therapeutic option as it needs frequent introduction of the endoscope leading to esophagitis, pressure ulceration and esophageal perforation. Gorter *et al.* showed that endoscopic removal of trichobezoars was successful in only about 5% of the patients, and they found that 100 (92.5%) of 108 patients were treated with laparotomy.[20] In our case, esophageal perforation developed after attempting to remove the trichobezoar endoscopically.

Conclusions

To the best of our knowledge, this is the first case of esophageal trichobezoar. We conied the definition *Esophageal Rapunzel Syndrome* to describe this condition for its similarity for the clinical history, symptoms, physical and pathological features of the bezoar, diagnosis and treatment with the well-known *Rapunzel syndrome*. A prompt diagnosis is essential to avoid severe complications, including esophageal rupture, that can occur also during EGD and requires an urgent surgical intervention. Based on our experience, patient with trichotillomania should be always referred for psychiatric consultation to avoid urgent surgical consequences.

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