

Rapunzel syndrome associated with bowel obstruction and upper GI bleeding: case report and literature review

Síndrome de Rapunzel asociado a obstrucción intestinal y hemorragia de vías digestivas altas: reporte de caso y revisión de literatura

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Abstract

Rapunzel syndrome is an unusual and rare disease that occurs in young people, caused by intragastric accumulation of ingested hair (gastric trichobezoar) that keeps stuck in the gastric lumen and extends to the small intestine, associated with trichophagia. We report the case of a 25-year-old female patient with a history of trichophagia who is admitted in our institution with abdominal pain, distention, nausea, weight loss and concomitant upper gastrointestinal bleeding. Preoperative diagnosis was made by prior abdominal computed tomography scan with duodenal extension. Success surgical laparotomy and multimodal psychiatric follow up was made. This entity must be considered as a differential diagnosis with Acute Abdomen with Bowel obstruction and upper gastrointestinal bleeding.

Keywords: Bezoars. Intestinal obstruction. Gastrointestinal hemorrhage. Feeding and eating disorders. Anxiety disorders.

Resumen

El síndrome de Rapunzel es una enfermedad inusual y rara que se presenta en personas jóvenes, caracterizada por un acúmulo de cabello ingerido (tricobezoar gástrico) y confinado generalmente a la cámara gástrica que se extiende al intestino delgado, asociada a tricofagia. Reportamos el caso de una paciente de 25 años con antecedente de tricofagia que ingresa a un centro de salud en curso de dolor, distensión abdominal y datos de obstrucción intestinal asociado a hemorragia digestiva alta. Con documentación preoperatoria por tomografía computada abdominal de un tricobezoar con extensión duodenal, el cual se extrae por laparotomía. La paciente se remite para manejo multimodal por servicio de psiquiatría. Es una afección para tener en cuenta como diagnóstico diferencial en los cuadros de abdomen agudo quirúrgico, de la mano con obstrucción intestinal y sangrado gastrointestinal.

Palabras clave: Bezoares. Obstrucción intestinal. Hemorragia gastrointestinal. Trastornos de alimentación y de la ingestión de alimentos. Trastornos de ansiedad.

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Date of reception: 02-08-2021

Date of acceptance: 26-08-2021

DOI: 10.24875/CIRUE.M21000495

Cir Cir (Eng). 2022;90(6):834-838

Contents available at PubMed

www.cirurgiaycirujanos.com

Introduction

Bezoars are masses of undigested material stuck in the gastrointestinal tract. Their shape varies depending on the type of material ingested, and the phyto-bezoar is the most commonly found type¹. Typically, there are four main types of bezoar according to the material they are made of: phytobezoar with fruit and vegetable fibers, trichobezoar (TB) with compacted hair strands, pharmacobezoar made up of oral medications (tablets), and lactobezoar comprised of milk by-products^{2,3}. 90% of TBs have been associated with psychiatric and behavioral disorder linked to trichotillomania and trichophagia, primarily occurring in young women. Their diagnosis is based in imaging findings with barium radiology studies, abdomen computerized tomography, or upper digestive endoscopy. They present a constellation of symptoms that are not very specific, where abdominal pain is the most common symptom, followed by nausea and vomiting, anorexia, anemia, bowel obstruction, and to a lesser degree, upper digestive bleeding due to ulceration of the gastric mucosa⁴. Rapunzel syndrome, initially described by Vaughan in 1968, is a rare complication of the formation of TB, where the mass of hair strands spans distal to the antra pyloricum toward any portion of the small intestine, and its morphology shows a distal elongation that takes up the entirety or part of the small intestine, up to the ascending cecum and colon^{5,6}. This article reports a case of Rapunzel Syndrome associated with bowel obstruction and upper digestive bleeding data.

Clinical case

Female, 25 years old, with history of trichophagia seeking care for a clinical presentation for 2 months with weight loss of 10 kg, epigastralgia radiated to the right hypochondrium, associated with abdominal dilation, nausea, vomiting, and the episodes of hematemesis in the 12 hours before being admitted. In her initial evaluation, she was dehydrated, pale, diaphoretic, she had tachycardia, with a finding of painful palpable mass in the epigastrium with signs of peritoneal irritation. The hemogram is compatible with macrocytic and hypochromic anemia, leucocytosis associated with neutrophilia, and the rest of the blood chemistry evaluated (biliopancreatic profile, kidney function, and arterial gases) are within normal range. A serial abdomen X-ray was performed, showing a dilation of the gastric chamber in the standing position with hydro-aerial levels and presence of distal gas.

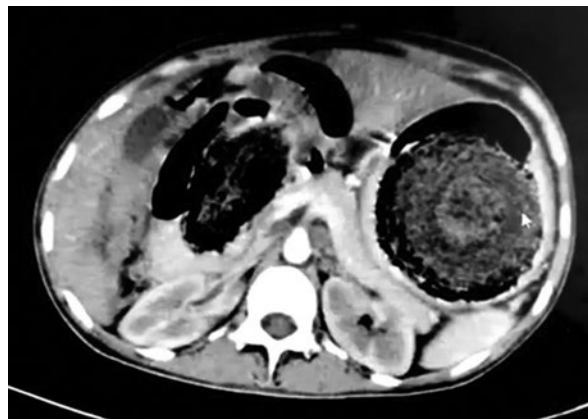


Figure 1. Total contrast computerized tomography of the abdomen showing a solid lesion with uneven outline in the stomach lumen.

A complete abdominal ultrasound showed a calcified intraabdominal mass of unknown etiology. A contrast tomography of the abdomen evidences dilation of the gastric chamber occupied by density material with soft parts, of uneven shape, which compromises large part of the gastric fundus and body, as well as the antra pyloricum and the duodenum (Fig. 1).

A exploration laparotomy showed the dilation of the gastric fundus and gastric body with an uneven palpable mass. An anterior gastrotomy (Fig. 2) is performed showing that the TB occupies 90% of the stomach lumen up to the first part of the duodenum, and for this reason it is manually removed (Fig. 3).

Afterward, an intragastric exploration shows ulceration of the mucosa and signs of recent bleeding. A gastrorrhaphy in two planes is performed with absorbable Polyglactine 910 multifilament sutures caliber 2-0, and the abdominal cavity is closed without any drainage devices. The patient improved satisfactorily, returns intestinal transit with effective peristalsis, and tolerates food by mouth. Strict psychiatric treatment is prescribed, and after 48 hours of monitoring, the patient is discharged.

Discussion

The first historic report of a TB dates back to the 18th century, when Baudamant described a 16-year-old boy with this condition. Trichotillomania, described for the first time in 1889 by Dr. Hallopeau as the desire of removing one's own hair, together with trichophagia, are considered concomitant disorders in up to 90% of TB cases, predominantly found in females with a history of major depression, bipolar



Figure 2. Anterior gastrotomy with exposure of the intraluminal foreign body.



Figure 3. Extraction of trichobezoar that spreads to the first portion of the duodenum.

affective disorder, schizophrenia, anxiety disorder, and post traumatic stress disorder history⁷ Approximately, 10% of patients with trichotillomania also present trichophagia as part as their ritual behavior⁸.

The spread of TB from the stomach to the small intestine is present in 49% to any level of the jejunum, to the duodenum in 18%, to the ileum in 21%, to the colon in 5%, and with origin in the jejunum and spread to the colon in 4% of the 72 reported cases

of Rapunzel Syndrome⁹. The gastric mucin coats the bezoar hair and provides a bright and rustic surface which, associated with the decomposition and fermentation of fat facilitated by the proteolysis of the hydrochloric acid cause compaction, solidification, and denaturalization of the hair proteins, providing dark pigment regardless of the color of the implanted strand¹⁰. We have adopted the universal definition of the Rapunzel Syndrome, which involves: 1) a TB with distal elongation, 2) spread to the small intestine, and 3) with signs that suggest bowel obstruction. The TB usually has a prepyloric location, but it can also compromise other levels of the intestinal tract less frequently, such as the duodenum and the proximal segments of the jejunum, even an obstruction of Vater's ampulla that conditions for jaundice, pancreatitis, or even appendix symptoms due to an obstruction of Gerlach's valve^{11,12}.

Clinically, over half of the patients that were seen in the emergency room with a TB had a history of chronic abdominal symptoms (57%), which included early satiety, weight loss, abdominal pain, abdominal distension, and intermittent vomiting. The most common finding in the physical exploration was a palpable abdominal mass (43%), bilious vomit, bowel obstruction found in X-rays (29%), and anemia caused by chronic gastrointestinal bleeding with required blood product transfusion before the surgical procedure (14%)^{13,14}.

The preferred test to confirm the preoperative diagnosis of the TB is an endoscopy of the upper digestive tract. We chose imaging studies, such as a double contrast computerized tomography scan of the abdomen and baric contrast abdomen X-ray to highlight the uneven endoluminal outline of the TB. In the abdomen X-Ray in the supine position a radio-lucent area can be seen in the gastric chamber with a thin elongation towards the duodenum, documented as a "comma"^{2,15-17}.

The upper digestive endoscopy is the most reliable diagnostic method, but the rate of therapeutic effectiveness is only 5%¹⁸. The preferred treatment for Rapunzel Syndrome is surgical extraction by laparotomy, as the laparoscopic method is considered less effective^{19,20}.

Smaller bezoars can be receive a conservative treatments, in general, with liquid diet for a brief period of time, and a prokinetic agents to promote gastric emptying. The chemical solution with cellulase is effective in up to 85% of patients with small bezoars, taken in the form of tablets or administered directly into the stomach in liquid form through an endoscope

or a nasogastric tube. Nasogastric washing promotes the physical dilution of small bezoars. Carbonated beverages dissolve over half of phytobezoars and over 90% when combined with endoscopic techniques²¹. The endoscope breaks down bezoars in smaller fragments. The fragmentation of bezoars is possible using endoscopic manipulation tools. Bezoar fragments can be pushed towards the small intestine or be removed through the mouth, always providing an overtube in order to facilitate frequent passes of the endoscope and ensuring airway protection. Endoscopic fragmentation and extraction yield positive results in 85-90% of small gastric bezoars, with the best effectiveness rates when combined with proteolytic enzymatic therapy²²⁻²⁶.

Bezoars in Rapunzel Syndrome is resistant to enzymatic degradation and conservative management. The frequent complications derived from the endoscopic approach make the surgical approach the preferred method for the definitive management of the symptoms and their complications. TBs require surgery more frequently than phytobezoars. Gastric bezoars are usually extracted through an anterior gastrotomy with exploratory laparotomy^{27,28}. Laparoscopic management is associated with less complications related to the wound, with more documented cases of peritoneal contamination due to spilling, and therefore, new techniques have been proposed, such as the *Iftikhar Jan* technique, which decreases the probability of contamination of the peritoneal cavity during the TB extraction process with the use of a provisional gastrotomy attached to a supraumbilical utility incision, two 5 mm infraumbilical and right-flank trochars. Small intestine bezoars are removed through an enterotomy or they can be propelled by applying transmural pressure to the cecum, where issues are rare due to the larger diameter of the colon. If a surgical procedure is considered, the existence of synchronous bezoars in more than one location should be ruled out^{29,30}.

Conclusions

TBs have low incidence, and Rapunzel Syndrome is an extremely rare and infrequent variant with varied clinical presentations. It is a challenging preoperative diagnostic, which is normally incidentally documented. The definitive treatment is almost exclusively surgical, where laparotomy is preferred, with an effectiveness of approximately 99% of cases, in addition to other

less invasive laparoscopic, endoscopic, and pharmacological techniques without the same results.

We have performed an extensive literature review starting from the case described, explaining similarities in the preferred surgical approach, with an uncommon presentation of upper digestive bleeding linked to the erosion caused by permanent contact of the TB with gastric mucosa. It is evident that, in our region, there has not been any thorough documentation of this disorder, and therefore, it must be included in differential diagnosis of surgical acute abdomen that begin with bowel obstruction and gastrointestinal bleeding. It is expected that this review will continue to contribute to the development of clear management algorithms, as well as to the unanimity in diagnostic criteria, which is far from being a universal concept. We support a multidisciplinary management in combination with the psychiatry specialty, due to the high prevalence of concomitant behavioral disorders and psychiatric pathologies.

Acknowledgements

The authors acknowledge the Research Committee of Universidad Libre for guiding the structuring of each section of this article.

Funding

The authors have not received any sponsorship or financing means in order to write this article.

Conflicts of interest

The authors have no conflicts of interest for the publication of this article.

Ethical disclosures

Protection of people and animals. The authors declare that no experiments were carried out on humans or animals for this research.

Data confidentiality. The authors declare that they have followed their center's protocols on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

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