

CASE REPORT

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Rapunzel syndrome debuts as a gastric perforation

Alberto De Anda Coronado, Anya Yamilia Adame Gutierrez,
Gerardo Enrique Muñoz Maldonado

ABSTRACT

Rapunzel syndrome refers to a rare disease that consists of a variant of gastric trichobezoar that has an extension distal to the small bowel. Most cases occur in young women with an underlying psychiatric illness, such as trichotillomania and trichophagia. The trichobezoar should be considered as a differential diagnosis of abdominal pain in young patients with the presence of associated psychiatric pathology since its early diagnosis allows avoiding complications. We present the case of Rapunzel syndrome in a young female patient with no documented history of trichotillomania or trichophagia who presented with gastric perforation. The patient underwent surgery, and a 25 cm trichobezoar was extracted from the stomach. She was discharged home on the 3rd postoperative day. Multidisciplinary management with general surgery, gastroenterology, and psychiatry divisions is essential for offering an integrative treatment to these patients.

Keywords: Gastric ulcer, Perforation, Trichobezoar, Trichophagia

How to cite this article

Coronado ADA, Gutierrez AYA, Maldonado GEM.
Rapunzel syndrome debuts as a gastric perforation.
Int J Case Rep Images 2022;13:101309Z01AC2022.

Article ID: 101309Z01AC2022

doi: 10.5348/101309Z01AC2022CR

INTRODUCTION

A bezoar is a foreign body composed of non-digestible material located in any portion of the digestive tract, being more common its location in the stomach [1]. The word bezoar derives from the Persian *Pâdzahr* which means antidote [2].

Trichobezoar is the most common type of bezoar. There are five types of bezoars according to their composition; trichobezoar: composed of human hair; phytobezoar: composed of fruit and vegetable fibers such as non-digestible cellulose; lactobezoar: composed of poorly diluted milk; pharmacobezoar: composed by ingestion of medicines and finally foreign bodies or substances different from the previous ones such as wood chips (lignobezoars), resins (resinobezoar), starch (amilobezoar), among others [3].

Unlike the rest of the bezoars, trichobezoars are common in young women with psychiatric disorders such as trichotillomania or trichophagia. Rapunzel syndrome is a rare and more severe form of gastric trichobezoar that has an extension into the small intestine [4]. Rapunzel syndrome can be an uncommon cause of chronic abdominal pain with rare complications such as intestinal obstruction and perforation.

We present the clinical case and subsequent management of Rapunzel syndrome in a patient with no documented history of trichotillomania or trichophagia who debuted with gastric perforation.

Alberto De Anda Coronado¹, MD, Anya Yamilia Adame Gutierrez¹, MD, Gerardo Enrique Muñoz Maldonado², MD, PhD, FACS

Affiliations: ¹PGY-3 General Surgery, Hospital Universitario Dr. José Eleuterio González, Monterrey, Nuevo Leon, Mexico; ²Chairman General Surgery Department, Hospital Universitario Dr. José Eleuterio González, Monterrey, Nuevo Leon, Mexico.

Corresponding Author: Gerardo Enrique Muñoz Maldonado, Chairman General Surgery Department, Hospital Universitario Dr. José Eleuterio González, Monterrey, Nuevo Leon, Mexico; Email: deandaalberto92@gmail.com

Received: 16 February 2022

Accepted: 02 April 2022

Published: 12 May 2022

CASE REPORT

A 27-year-old female patient, with a surgical history of two cesarean sections, presented to the emergency department of a tertiary level hospital in the metropolitan area of Monterrey, Nuevo Leon, Mexico; with a 2-year history of abdominal pain, nausea, intermittent vomiting, and early satiety. She denied other accompanying features such as changes in bowel habit, hematemesis, or melena and had no urinary tract or gynecologic symptoms.

On physical examination she had a heart rate of 120 beats per minute with a blood pressure of 90/60 mmHg, an axillary temperature of 37.2 °C, distended abdomen, auscultation showed the absence of peristaltic sounds, acute peritoneal signs such as abdominal rigidity, and generalized pain of intensity 9/10 evaluated with visual analog scale, the rest of the examination was without relevant data. The paraclinical examinations showed white blood cells of 32,000/mm, arterial gasometry showed an acidosis of 7.31 pH, metabolic type. Abdominal and chest X-rays were requested, which showed a J-shaped stomach, and intragastric mass as well as subdiaphragmatic free air (Figure 1). It was decided to admit her to the operating room by the General Surgery service, performing a laparotomy revealing a gastric perforation in the lesser curvature at the level of the body on the anterior gastric face (Figure 2) with the presence of a foreign body inside the stomach lumen compatible with trichobezoar, performing a wide gastrotomy extracting trichobezoar of 25 × 20 cm (Figure 3), which occupied the entire gastric chamber presenting extension to the second portion of the duodenum.



Figure 1: Chest X-ray showing pneumoperitoneum.

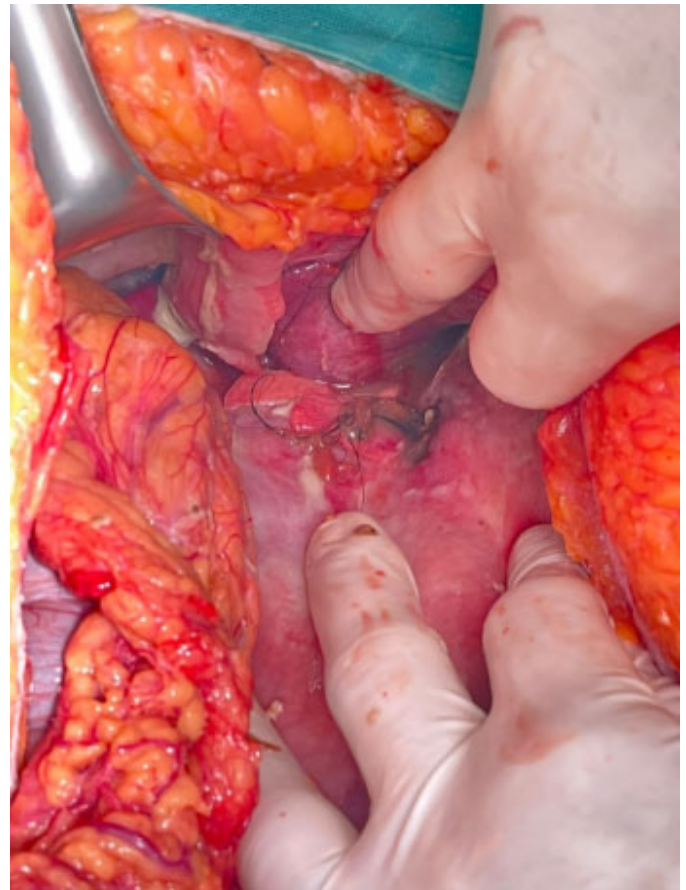


Figure 2: Gastric ulcer located at the lesser curvature.



Figure 3: Gastric and duodenum shaped trichobezoar.

During her postoperative recovery, the patient evolved favorably, being discharged on the fifth postoperative day with diagnoses of trichotillomania, gastric perforation by gastroduodenal trichobezoar, generalized anxiety disorder, and depression, being in control and follow-up by the Psychiatry Service.

DISCUSSION

The oldest reports of this condition date back to the 12th century BC in India; however, the first officially documented trichobezoar was reported by Bardeman in 1779 as a mass of hair found in the autopsy of a patient who died of gastric perforation and peritonitis [5].

The presence of a trichobezoar is rare, most cases being in young women under 30 years of age with underlying psychiatric disease, such as trichotillomania, although it is not uncommon for them to deny an underlying pathology.

Human hair is resistant to digestion and intestinal peristalsis, so continuous and prolonged ingestion causes its impaction in the lumina. Other factors associated with the formation of bezoars include altered gastric motility, decreased acidity, bacterial colonies, lack of surface area for propulsion, and entrapment of the fibers in the folds of the gastric mucosa [6].

Trichobezoar mainly affects the stomach; however, on rare occasions it goes beyond the pylorus and manages to extend to the duodenum and jejunum, being called Rapunzel syndrome, named after the eponymous heroine of a German fairy tale written by the Brothers Grimm, first described by Vaughan et al. in 1968 [7].

The clinical picture in these patients usually presents until the trichobezoar grows considerably to obstruction, the most common symptoms include a palpable abdominal lump in the epigastrium, chronic abdominal pain, nausea, vomiting, constipation, alopecia areata, and even in some patients halitosis, weight loss, anorexia, hematemesis, and iron deficiency anemia can be found. Complications, although rare, include ulcer formation, perforation, peritonitis, intussusception, malnutrition, jaundice, and pancreatitis.

Radiological diagnosis can be made with simple abdominal radiography, gastrointestinal series with contrast, and abdominal computed tomography in which the trichobezoar can be evidenced as a filling defect in the stomach and small intestine, being possible to delimit its extension.

Upper endoscopy plays an important role in the diagnosis of trichobezoars and Rapunzel syndrome since it allows direct visualization, as well as a possible therapeutic intervention; however, it is rarely a definitive treatment due to the large size that gastric trichobezoars can have [8].

Surgical removal remains the cornerstone in the treatment of trichobezoars and Rapunzel syndrome, it is performed by gastrotomy or enterotomy as in the present

case; however, recent studies describe the successful laparoscopic removal of trichobezoars in pediatric patients.

CONCLUSION

Trichobezoar is infrequent and Rapunzel syndrome is a very rare condition, yet it should be considered as a differential diagnosis in young women with psychiatric history who report palpable abdominal tumor, abdominal pain, nausea, vomiting, early satiety, or other gastrointestinal manifestations. Early diagnosis avoids complications that can become serious. Intentional questioning of a patient's history of trichotillomania or trichophagia is essential for diagnosis. Multidisciplinary management and the long-term psychiatric follow-up are important parts of treatment and prevention of recurrence.

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Author Contributions

Alberto De Anda Coronado – Design of the work, Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Anya Yamilia Adame Gutierrez – Analysis of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Gerardo Enrique Muñoz Maldonado – Design of the work, Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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