CASE REPORT

Gastric trichobezoar and Rapunzel syndrome: case report of a very rare condition

Kouskos E[1], Polychroni D[2], Rizou M[1], Andronikou A[1], Petrellis V[3]

[1]Surgical Department
[2]Department of Obstetrics and Gynecology
[3]Department of Radiology
General Hospital of Mytilene, Lesvos Island, Greece

Abstract

Background: Rapunzel syndrome is a rare trichobezoar variant extending from the gastric cavity into the small bowel.

Case description: We report the case of a 22-year-old woman who presented with epigastric pain, nausea, and loss of appetite within the preceding five weeks. She had a palpable mass in the epigastric area with mild localized tenderness. Her abdominal computed tomography scan showed a distended stomach and duodenum, with a heterogeneous solid material, suspicious for a bezoar. Upper gastrointestinal endoscopy revealed a large, densely packed trichobezoar occupying the gastric cavity and extending through the pylorus. Endoscopic removal of the bezoar was unsuccessful. The patient underwent a 6cm-long gastrotomy, and the 150cm-long bezoar, extending from the stomach to the jejunum, was uneventfully removed. The patient was referred postoperatively to a dietitian and psychiatrist for management of her trichotillomania and trichophagia.

Conclusion: Trichobezoars are commonly found in young females with a history of trichotillomania and trichophagia and are associated with psychiatric disorders. HIPPOKRATIA 2023, 27 (1):25-27.

Keywords: Trichobezoar, Rapunzel syndrome, trichotillomania, trichophagia

Corresponding author: Kouskos Efstratios, Surgical Department, General Hospital of Mytilene, Lesvos Island, Greece, e-mail: skouskos@hotmail.com

Introduction

Bezoars are concretions of undigested material found in the stomach. They are differentiated depending on the type of retained foreign bodies[1]. This material may include plant fibers (phytobezoar), hair (trichobezoar), medications (pharmacobezoar), paper, milk protein, etc[1,2]. Trichobezoars are formed by ingesting and accumulating hair- or hair-like fibers in the gastrointestinal tract, especially in the stomach[1,3].

Rapunzel syndrome is a rare form of trichobezoar. This entity occurs as gastric bezoars get bigger, beyond the small bowel, producing long, tail-like hair extensions[1].

Bezoars usually cause no specific symptoms and, as a result, may imitate other diseases such as tumors[1]. By the time they reach a large size, they can be related to severe gastrointestinal complications such as perforation, peritonitis, pancreatitis, constipation, appendicitis, jaundice, steatorrhea, protein-losing enteropathy, and intussusception[4].

Trichotillomania is a psychiatric condition characterized by compulsive behaviors like self-induced and repetitive hair-pulling, in combination with obsessive thoughts, mood disorders, and anxiety[1]. Almost half of trichotillomania cases have skin-picking disorder as a comorbid condition[5]. It affects up to 4 % of the population, mostly children and adolescents[2].

In this article, we report a rare case of Rapunzel syndrome in a young female patient with a very long trichobezoar that was successfully treated surgically.

Case presentation

A 22-year-old woman with clinical signs of malnutrition presented to the emergency department of our institution with dull abdominal pain and distention in the epigastric area. She also complained of nausea, occasional vomiting, and loss of appetite for at least the preceding five weeks. She reported normal bowel movements and denied fever or diarrhea. Her past medical history was unremarkable, and her vital signs were stable.

Physical examination revealed a soft abdomen and a well-defined, relatively mobile, large epigastric mass extending from the left upper quarter to the umbilicus. Bowel sounds were audible. There were no other signs indicating peritonitis. There was no blood on digital rectal examination.

Laboratory results revealed a reduced hemoglobin level of 9.8 g/dl and an elevated white cell count with 82.3 % neutrophils. C-reactive protein was elevated at 12.9 mg/l. Urea, electrolytes, liver function tests, and coagulation

profile were within normal range. The abdominal computed tomography (CT) scan demonstrated a distended stomach and duodenum with a large, well-circumscribed mass with internal air loculi occupying almost the entire stomach and extending into the duodenum (Figure 1). Neither gastric obstruction nor perforation was evident. Images were highly suggestive of gastric bezoar. Given the characteristic radiological findings, the patient underwent upper gastrointestinal endoscopy under conscious sedation. The procedure confirmed the diagnosis, revealing Rapunzel syndrome. The mass was found under the esophagogastric junction extending through the pylorus to the duodenum (Figure 2). The hairball, covered by mucus, had a glistening shiny surface. Due to the large size, repeated attempts of endoscopic removal of the tangled bezoar were unsuccessful.

Based on imaging, we decided to proceed with open laparotomy. Through a limited midline supraumbilical incision, we performed a 6cm-long anterior gastrotomy. A huge gastric trichobezoar and its tapering tail were completely removed without complications (Figure 3, Figure 4). The proximal part of the bezoar was found molded into the shape of the stomach and duodenum, while its total length was more than 150 cm. The postoperative course of the patient was uneventful. She was discharged in satisfactory condition on the fourth postoperative day and referred for outpatient psychiatric and dietitian consultation. Psychiatric evaluation diagnosed her with anxiety, trichotillomania, and trichophagia, with onset dating back to her final high school year. She was assigned to a psychiatric follow-up and support to avoid recurrences.

Discussion

Rapunzel syndrome is a rare form of an occluding trichobezoar, where the gastric bezoar with a tail extends to the duodenum, ileum, or even the colon[3,6,7]. The large hair quantity assumes the stomach shape as a single mass[4]. Underlying mental disorders that are usually associated with the syndrome are anxiety and post-traumatic stress, leading to trichophagia and trichotillomania[8]. The

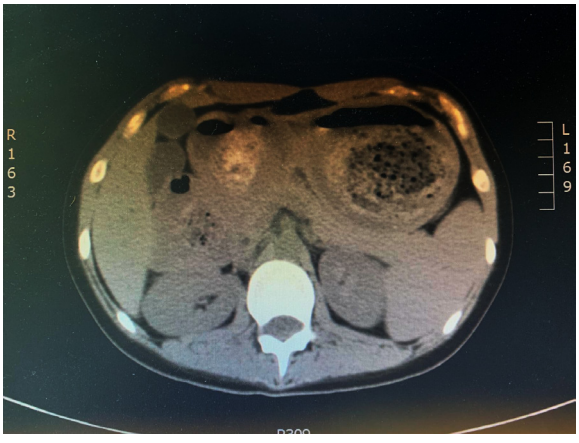

Figure 1: Axial computed tomography image demonstrating a large heterogenous intraluminal mass obstructing the entire lumen of the stomach.

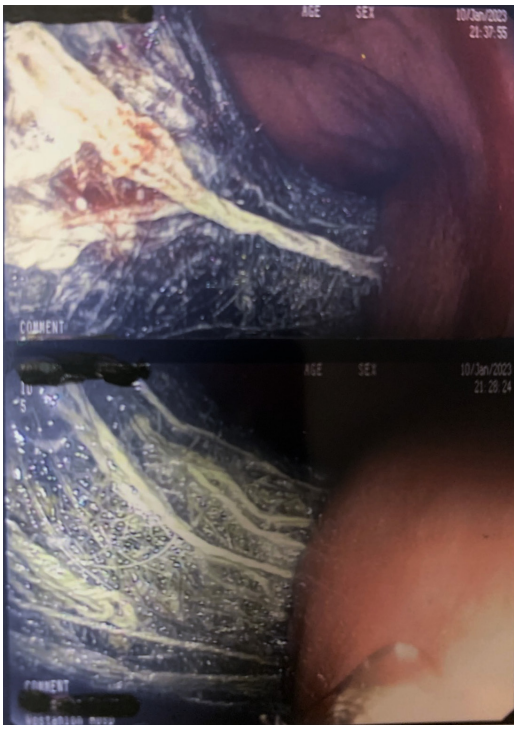

Figure 2: Endoscopic images during the upper gastrointestinal endoscopy, which confirmed gastric trichobezoar extending from the esophagogastric junction to the duodenum.

syndrome was first reported by Vaughan ED in 1968[9]. The term originates from Grimm Brothers' fairy tale of a very young princess locked in an inaccessible tower and a young prince, trying to save her, climbs up her long tresses[4].

The accurate incidence of trichobezoars is unknown[3]. It is much more common in female patients between the ages of 10 and 19 years old, attributed to the traditional long hair[3,4]. Young women with emotional disorders, behavioral disturbances, and mental retardation are prone to continuous habitual ingestion of hair in the gastrointestinal tract[2,10,11].

The formation of a bezoar is extremely rare, and it occurs in only 1 % of patients suffering from trichophagia. Trichobezoar is related to the indigestible composition of hair, which becomes entangled in the gastric folds. Hairballs that are resistant to digestive enzymes and escape the peristaltic propulsion continue accumulating into bigger mass size[9,12]. Gastric acid denatures and oxidizes the hair, and gastric mucus covers the mass[8]. As confirmed from our case report, the bezoar has a shiny, dark look.

Differential diagnosis may prove difficult due to nonspecific symptoms and the rarity of the syndrome. The personal history of trichotillomania and trichophagia and dietary habits could be helpful clues in the diagnostic process. Symptoms usually start developing as the hair mass sizes up[8]. Most patients are diagnosed with weight and appetite loss, halitosis, abdominal pain, abdominal mass, and gastrointestinal obstruction[9,10]. A well-defined, firm, mobile epigastric mass is revealed at physical ex-

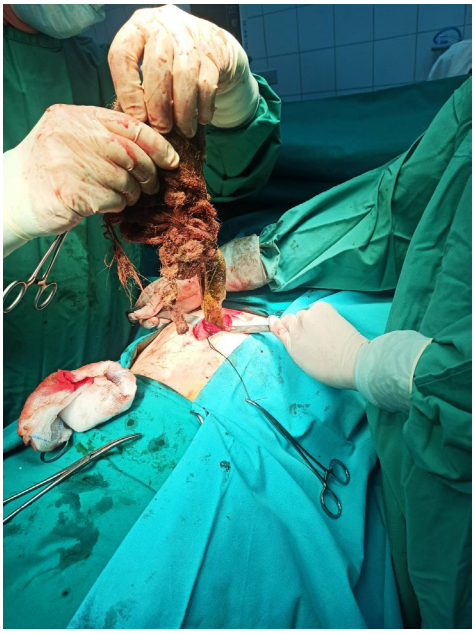

Figure 3: Intraoperative view showing trichobezoar removal through limited gastrotomy.

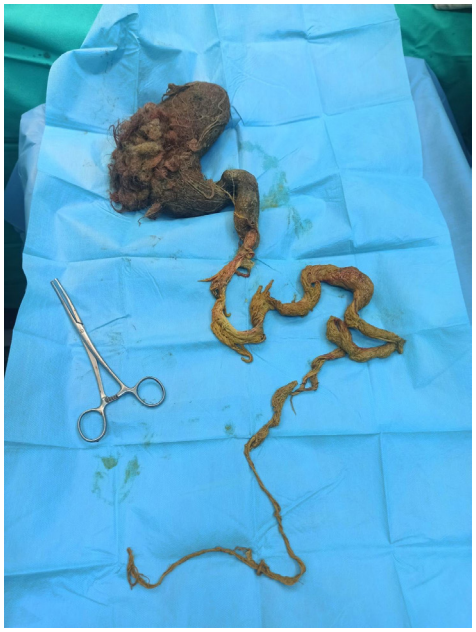

Figure 4: A stomach-shaped trichobezoar of about 150 cm in length extending to the small bowel.

amination in 85 % of patients[3,6]. Blood tests may demonstrate anemia, electrolyte disorders, malabsorption of iron, and hypoproteinemia[3].

Early diagnosis is crucial since neglected obstructing or eroding bezoar may lead to complications such as bleeding, ulceration, and perforation[8,13]. Abdominal CT scan reveals a heterogenous intragastric lesion consisting of entrapped air bubbles, food debris, and concentric rings[2,11]. However, upper gastrointestinal endoscopy remains the diagnostic gold standard method[3,8]. It provides direct visualization of the bezoar and can be used for diagnostic and therapeutic purposes[13]. In our study, a CT scan showed an oval intraluminal mass in the distended stomach and duodenum, and endoscopy revealed the large size of the bezoar.

The treatment of the bezoar focuses on the complete removal of the hair mass. The treatment approach depends on the bezoar's size, density, and localization[3,8]. Small trichobezoars can be managed by endoscopic removal, endoscopic fragmentation, gastric lavage, enzymatic therapy, or a combination of these methods[8,14]. Larger bezoars and Rapunzel syndrome usually require laparotomy with longitudinal gastrotomy, as performed in our case[13]. Although a minimally invasive approach with satisfactory postoperative outcomes has been reported by a few authors, the open conventional method is still the preferred method[13].

A recurrence of the syndrome has been reported. The lack of postoperative care and/or the untreated underlying trichotillomania may result in the disorder's relapse[1]. Since this entity is extremely rare, long-term psychiatric/psychological therapy and dietitian consultation seem to be prerequisites for the prevention of recurrence[8].

Conflict of Interest

None declared by authors.

References

1. Balawender K, Pliszka A, Możdżeñ K, Kłos M, Ogorzałek A, Boroñ D, et al. Trichopsychodermatology: trichotillomania and trichophagia leading to Rapunzel syndrome. Postepy Dermatol Alergol. 2022; 39: 739-743.
2. Sivasubramanian BP, Ashokkumar M, Afzal M, Samala Venkata V, Dhanasekaran UD, Palit SR, et al. Rapunzel Syndrome in a Teenage Girl: A Case Report. Cureus. 2022; 14: e29975.
3. Delimpaltadaki DG, Gkionis IG, Flamourakis ME, Strehle AF, Bachlitzanakis EN, Giakoumakis MI, et al. A rare giant gastric trichobezoar in a young female patient: Case report and review of the literature. Clin Case Rep. 2021; 9: e05152.
4. Naik S, Gupta V, Naik S, Rangole A, Chaudhary AK, Jain P, et al. Rapunzel syndrome reviewed and redefined. Dig Surg. 2007; 24: 157-161.
5. Madhusudhan KS, Vaishnav M, Joy D. Rapunzel Syndrome. J Gastroinest Surg. 2022; 26: 1111-1112.
6. Chin X, Ng JY. Acute Presentation of Rapunzel Syndrome and a Review of Bezoars. Cureus. 2021; 13: e20785.
7. Vaughan ED Jr, Sawyers JL, Scott HW Jr. The Rapunzel Syndrome: An unusual complication of intestinal bezoar. Surgery. 1968; 63: 339-343.
8. Schuler L, Hodel M, Stieger C. The Rapunzel syndrome: a hairy tale. Surg Case Rep. 2023; 9: 49.
9. Jackman J, Nana GR, Catton J, Christakis I. Gastric perforation secondary to Rapunzel syndrome. BMJ Case Rep. 2021; 14: e240100.
10. Daoud R, Tlili A, Fendri S, Akrout A, Trigui A, Boujelbene S. Gastric trichobezoar: An uncommon cause of epigastric pain: A case report. Clin Case Rep. 2022; 10: e05653.
11. Kwon HJ, Park J. Treatment of large gastric trichobezoar in children: Two case reports and literature review. Medicine (Baltimore). 2023; 102: e33589.
12. Lalith S, Gopalakrishnan KL, Ilangovan G, Jayajothi A. Rapunzel Syndrome. J Clin Diagn Res. 2017; 11: TD01-TD02.
13. Haggui B, Hidouri S, Ksia A, Oumaya M, Mosbahi S, Messaoud M, et al. Management of trichobezoar: About 6 cases. Afr J Paediatr Surg. 2022; 19: 102-104.
14. Mazzei A, Centonze A, Aloi IP, Bertocchini A, Baldassarre E. Rapunzel Syndrome: Endoscopy, Laparotomy, or Laparoscopy? J Indian Assoc Pediatr Surg. 2021; 26: 66-67.