CASE REPORT

The Rapunzel syndrome - a case report: 
trichobezoar as a cause of intestinal perforation

Andréa P. Faria,1 Ivy Z. Silva,1 Aneli Santos,1 Sylvio G. A. Avilla,2 Antônio E. Silveira3

Abstract

Objective: to report a rare case of patient with gastric trichobezoar extended through the small bowel, suggestive of Rapunzel syndrome.

Case report: a 7-year old girl with history and exams suggestive of peritonitis in whom laparotomy demonstrated trichobezoar and jejunal perforation. She also presented alopecia and psychological disorder, which were not totally eradicated despite the support given.

Conclusion: a multidisciplinary view of the patient would have increased the suspicion rate for the pathology, providing an early diagnosis and treatment before complication was evident.


Introduction

The word bezoar is derived from the Arabic “badzehr” or from the Persian “padzahr”, and it means antidote.1,2

Today, bezoars are defined as foreign bodies formed in the stomach and/or small bowel due to an accumulation of swallowed substances. These substances may be vegetable fibers (phytobezoars), animal fats, antacids, resin, plastic, latex, and hair (trichobezoar). Gastric trichobezoar, presenting a “tail” extended through the small bowel, is rare and proper of Rapunzel syndrome, first described by Vaughan et al. in 1968.3,4 In ancient times, bezoars found in animals and in human beings were used in charms.1,2

Although trichotillomania (irresistible will to pull out the own hair) was only described in 1889 by Hallopeau, the first report of a trichobezoar case occurred in 1779, by Baudamant; the first surgical excision was performed by Schonborn in 1883.2,5

The most common complication associated with gastrointestinal trichobezoar is intestinal obstruction,4 and, less frequently, small bowel perforation.2 The treatment of Rapunzel syndrome aims at the complete elimination of trichobezoar from the gastrointestinal tract. When the small bowel is compromised, the trichobezoar removal by enterotomy is suggested,4 associated or not with gastrostomy.1

1. Undergraduate student, Pontifícia Universidade Católica do Paraná (PUC/PR) School of Medicine.
2. Pediatric Surgeon, HIPP-PR, and Assistant Professor of Pediatric Surgery, PUC/PR.
3. Pediatric Surgeon, HIPP-PR, and Professor of Pediatric Surgery, PUC/PR.
Case report

Patient MSA, female, 7 years old, admitted to the Hospital Infantil Pequeno Príncipe, in Curitiba, state of Paraná, Brazil, presented with complaint of vomiting, abdominal pain, fever, and lack of fecal or intestinal gas elimination for 3 days prior to admission. History reported by the father (since the patient herself did not cooperate) describes a status of abdominal pain for 8 months, similar to colic, intermittent, of low intensity, not accompanied by vomiting. The onset of pain in this patient is related to the date of the litigious divorce of her parents. Initially, the patient was evaluated and treated, without improvement, as if the pain was caused by gastritis. The pain increased in both intensity and frequency, and for 7 days prior to admission, her clinical status worsened progressively. The patient began to present high fever, food-related vomiting, and, later on, bilious vomiting, diffuse abdominal distension, and lack of fecal or intestinal gas elimination. At that moment, the patient was directed to the service of pediatric surgery, Hospital Infantil Pequeno Príncipe, for evaluation.

On physical examination, the patient was agitated and uncooperative. Moreover, she presented a general compromise of her state, toxic facies, moderate paleness of the skin and mucosae, alopecia in bilateral temporal region, tachycardia, fever, distended abdomen, sore at superficial palpation, lacking hydro-aerial noises and signs of peritonitis. Other organs did not present alterations.

The patient was hospitalized with diagnosis of peritonitis of unknown cause.

Complementary exams. A) Hemogram: hemoglobin = 7.8 g/dl; leukocytes = 21,000/mm³; basophils = 43%; neutrophils = 25%; eosinophils = 0%. B) Radiological examination of the abdomen: diffuse distention of the intestinal loops; possibility of free fluid in the cavity, preperitoneal line blurring; and epigastric mass. C) Abdominal echographic examination: free fluid in the peritoneal cavity and presence of grumes, which is compatible with peritonitis; presence of an extended solid mass inside the stomach and duodenum, suggesting hypothesis of tumor or foreign body. Due to the patient’s clinical status, we did not perform a contrasted study, and an explorative laparotomy was indicated.

Description of the surgical procedure. A) Inhaled general anesthesia; extended supraumbilical transverse incision; opening of the abdominal wall by planes, in order to access the peritoneal cavity. B) Presence of large amount of purulent secretion; friable, distended, and edematous intestinal loops. C) Aspiration of purulent secretion and exteriorization of all small bowel loops. D) Presence of intestinal perforation in the jejunum, 15 cm distal from Treitz angle; presence of a hairball, emerging through the perforation orifice. E) By palpation, we localized an extended mass occupying the proximal jejunum, the whole duodenum, and the stomach. F) Through the already existing perforation, by traction, we were able to remove the extended trichobezoar (Figure 1). G) Resection of the jejunal segment (Figure 2); enteroanastomosis, mechanically cleaning the cavity, and closing it by planes. Although the clinical indication for the treatment of this patient (peritonitis and perforation) was to associate gastrotomy, enterotomy, and intestinal resection, gastrotomy was not performed, since, in this case, the bezoar removal was made possible by the already existing jejunal perforation, a relatively easy process. We judged that this trauma would be less harmful than the one caused by another lesion (even being elective) in the gastrointestinal tract.

Figure 1 - Trichobezoar: gastric portion and tail, which extended through the intestine
The immediate postoperative care was difficult, since the patient presented septic shock evolving to multiple organ failure. She was submitted to a strict hydroelectrolytic support in the intensive care unit, where she remained for 7 days. In the eighth day, she began to be fed orally and to receive psychological advice. Then the patient was directed to the surgical and psychological outpatient unit for 8 months. Alopecia and the habit of eating hair returned in the 6th postoperative month. Despite medical and psychological guidance, the patient did not return for control, and in the 8th month after the surgery, we lost contact with her.

Discussion

Although bezoars are rare, trichobezoar is the form most commonly found in childhood.\(^1\)

Rapunzel syndrome received this name out of the long-haired heroine of the Grimm brothers’ fairytale.\(^1\) It was first described by Vaughan et al. in 1968,\(^3,4\) and it is constituted of a trichobezoar of rare appearance: only nine cases have been described until June 1996.\(^4\)

This syndrome is usually found among teenage girls with psychological disorders, and it is manifested as follows: abdominal discomfort or nausea and vomiting, anorexia, weight loss, and habit of eating hair (intentional alopecia may be a frequent clue for the diagnosis).\(^1\) Radiological exams of the abdomen, either simple or contrasted, may confirm the diagnosis. Ultrasonography, computed tomography, and high digestive endoscopy assist the diagnosis and the preoperative evaluation.\(^1,4\) Although the manifestations of this syndrome vary according to each patient, an usual triad consists of abdominal mass, bezoar’s tail extended from the stomach to the small bowel, and gastrointestinal symptoms.\(^1\)

The formation of trichobezoar starts with the intake of small bits of hair that do not progress to the gastrointestinal tube, getting accumulated and gathered.\(^2\) This condition can evolve to the following complications: ulceration, partial obstruction, total obstruction, necrosis, intestinal perforation, or peritonitis.\(^1,2,4,5\) The dimensions of the bezoar seem to have a relation with the morbidity and mortality evaluation, but not with the evolution of the clinical status and its complications. Therefore, the pathological findings of the complications are not related to the size of the trichobezoar.\(^1\)

The treatment aims at the complete removal of bezoars from the gastrointestinal tract. Small bezoars may be removed by endoscopy, associated or not with laser and shock wave (lithotripsy).\(^6\) Phytobezoars may be eradicated with enzymes, fragmentation, or medications. The treatment must begin as non-invasive, but surgery is the best prognosis in cases of severe clinical status that do not respond to conservative handling, as well as in cases presenting complications.\(^6\) There are no reports of spontaneous resolution of obstructions caused by trichobezoar.\(^6\) Depending on the clinical status, Rapunzel syndrome may be treated with gastrotomy and/or enterotomy (in cases of intestinal obstruction), or extended resection of the intestine associated with gastrotomy and enterotomy (in cases of necrosis, perforation or peritonitis).\(^1\) We must emphasize that gastrotomy is not indicated in all cases; if the patient presents intestinal obstruction, which happens in most cases,\(^4\) enterotomy may be performed instead.\(^1\) The treatment of trichotillomania, nowadays characterized as an obsessive-compulsive disorder, may be performed with the administration of clomipramine or desipramine, both tricyclic antidepressants, - the first is the most efficient and frequently prescribed drug.\(^5\) Evaluation and psychological support are important and necessary in order to provide a better life quality to the patient, and to prevent complications and recidivism.

In the suspicion of bezoar, the diagnosis must be established as early as possible, in order to provide an effective treatment, which benefits the patient.
References


Correspondence:
Dra. Andréa Palazzo Faria
Rua Euclides Bandeira, 500 - ap. 1301
CEP 80530-020 – Curitiba, PR, Brazil
Phone: + 55 41 253.7652
Fax: + 55 41 253.7790 / 254.5892