

Rapunzel syndrome

A case report and literature review

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Abstract

The presence of trichobezoars is a rare condition which usually occurs in young women who have histories of trichotillomania and trichophagia. Nowadays, the majority of cases occur in patients with a history of gastric surgery, or pyloric function alteration. They may be clinically asymptomatic for months or years or may present acute symptoms accompanied by severe complications.

This article presents the case of a pregnant patient who was diagnosed with a case of Rapunzel syndrome a is presented. This complex variety of gastroduodenal trichobezoar involves the stomach, duodenum and intestine. The article also reviews the literature about the Rapunzel syndrome.

Keywords

Trichobezoar, Rapunzel syndrome, pregnant woman.

INTRODUCTION

Generally, bezoars are intraluminal accumulations of indigestible material. They have been classified, according to their predominant composition, into phytobezoars (vegetable and fruit residue), lactobezoars (milk residue especially in premature babies who consume concentrated formulas), pharmacobezoars (cellulose residue of patients with “polypharmacy”) and trichobezoars (an accumulation of hair). Trichobezoars usually occur when hair is eaten by people with mental retardation, psychiatric disorders or personality disorders such as trichophagia and trichotillomania. Rapunzel syndrome is a rare and complex disorder in which a gastric bezoar extends into the duodenum and small intestine increasing the risk of complications such as obstruction, perforation and peritonitis (1, 2).

Since the initial description of this syndrome by Dr Vaughan in 1968 fewer than 40 cases have been reported in the literature. Thirty percent of these have been in India. In

our environment this is an uncommon entity about which there are few reports (2,9).

CASE REPORT

The patient was a 20 year old female from Bogotá with a history of chronic gastritis which had been documented by extra-institutional endoscopy one a half years prior to hospitalization. The patient suffered from a personality disorder for which she had not received any medication and had dyspeptic syndrome refractory to treatment. She had been seen at several different medical institutions in the previous six months because of worsening gastrointestinal symptoms. When she was referred to our institution she had clinical symptoms of burning epigastric pain associated with nausea and retching which had been diagnosed as peptic acid disease after three days of development. She was treated with antacids and proton pump inhibitors in an outpatient clinic, but without improvement. Instead, she

presented worsening symptoms with episodes of hematemesis. For this reason she was referred to our institution (Hospital Cardiovascular Child Cundinamarca - Soacha).

During her physical examination the patient was alert. She had a mucocutaneous pallor and signs of dehydration, tenderness in the epigastrium and left upper quadrant without signs of peritoneal irritation or palpable masses, but with obvious abdominal distension.

Initial paraclinical studies included a CBC which showed hemoglobin 7.5g/dl, hematocrit 26%, a platelet count of 155,000, and a leukocyte count of 6,800 (65% neutrophils). The patient was positive for Beta Subunit human chorionic gonadotrophin, so an obstetric ultrasound was performed. It showed a multiple dichorionic-diamniotic (DiDi) pregnancy of 7.5 weeks. The patient was hospitalized because of the clinical history reported and laboratory findings. A transfusion of 2 units of blood and crystalloid volume resuscitation were performed. An upper GI endoscopy showed a large (trichobezoar) foreign body occupying the whole gastric space and which extended to the duodenum and beyond. Balloon enteroscopy (Olympus SIF-180 Q), performed to complement the endoscopic assessment, showed an extension of the bezoar to the jejunum.

Due to the critical condition already discussed, and the requirement that the bezoar be extracted, endoscopic management was initially attempted. It obtained a large number of different materials including hair, plastic, basketwork, shoe laces and other items. However, because of the size and diversity of materials, endoscopic removal was considered to be too difficult technically. The patient was referred for surgery where a laparotomy and a gastrotomy were performed. A foreign body which had extended beyond the angle of Treitz was removed (Figure 1).



Figure 1. Bezoar extending to intestine.

Following the operation the patient and her pregnancy were successfully managed with proton pump inhibitors and analgesics. She left the hospital five days after surgery with outpatient follow-up by surgical, gynecological and psychiatric services.

DISCUSSION

The term bezoar is derived or translated from the Arabic word *badzhe*, the Persian word *padzhar* and the Hebrew word *beluzer* which all mean antidote. It was thought that bezoars had healing powers against the poisoning. Most cases have been reported in young women under 30 with a peak incidence between 15 and 20 years of age (2, 10). Patients with bezoars often have personality disorders, psychotic features, delusions, limited adaptive skills (mental retardation) and changes in mood (1-3, 5-7).

Although there are some data on cases in the twelfth century BC, apparently Sushruta and Charak first described bezoars in the second and third century BC (7, 8, 10). The first surgery for this illness was performed in 1883 by Dr. Schonborn (5,10).

Pathophysiologically, trichobezoars are formed because ingested materials are retained in the gastric folds and, due to the small contact area, they can resist the stomach's propulsion to expel them (1, 2, 10).

The most common clinical manifestations include palpable epigastric masses in 70% of cases, abdominal pain in 37%, nausea and vomiting in 33%, weight loss in 38%, and diarrhea and anorexia in 32% (2,10). The most frequently reported complications include intestinal obstructions, pancreatitis due to irritation and swelling of the ampulla of Vater, and intestinal bleeding. There have even been perforations with peritonitis (1, 2, 10).

The Rapunzel syndrome is a rare but potentially fatal trichobezoar. The name is an allusion to the tale written in 1812 by the Brothers Grimm about a young woman named Rapunzel. She lets her long hair down from the tower where she is imprisoned to the prince she loves to climb up and save her. This syndrome was originally described by Vaughan et al. in 1968 as a strange variety of trichobezoar, which extended down the pylorus into the duodenum (2, 7, 10). It is a rare entity of which there are few reports in literature and for which there is no formal consensus on diagnostic criteria. However Naik et al. have proposed the following diagnostic triad (2):

DIAGNOSTIC CRITERIA FOR RAPUNZEL SYNDROME

1. Trichobezoar with a tail
2. Extension of this tail at least until the jejunum

3. Obstructive symptoms

As mentioned, the risk factors for this syndrome include,

1. Risk factors for all trichobezoars: mental illness, adjustment disorder, trichotillomania and trichophagia. It should be noted that almost all reported cases have been young women with long hair.
2. Predisposition to the formation of bezoars due to changes in gastric or intestinal anatomy, commonly after gastroduodenal surgery or disorders of gastrointestinal tract motility and gastroparesis (2, 4, 10).

Diagnosis is not easy because the patient usually denies or hides information about trichophagia or trichotillomania making clinical suspicion difficult. Diagnostic methods used in these cases include ultrasound in which an indication is high gastric intraluminal echogenicity with posterior acoustic shadow (2, 6, 14). CAT scans have proven to be better able to describe the site and size of the bezoar. They also allow differentiation between a bezoar and a neoplasm (2, 6, 7). Contrast studies with barium surrounding a mass, generally floating in the suspension, allows us to rule out a tumor. In addition, in some cases the barium becomes trapped in the interstices of the bezoar producing a classic "honeycomb" image (2, 6). Finally, the ideal method is endoscopy because it allows diagnosis in early stages, has a diagnostic performance 4 times greater than do imaging studies, and because it has therapeutic potential in less complex cases (1, 2, 5, 6, 10).

The initial goal of treatment in these patients is the extraction of the bezoar and prevention of recurrence with psychiatric therapy (6, 11, 12). Unlike other bezoars, trichobezoars are resistant to dissolution using enzymes such as catalase, coca-cola, and prokinetics such as metoclopramide which have been used successfully in other bezoars (5, 10). Thus, the definitive treatment for these patients is mechanical removal either endoscopically or surgically. They can be fragmented with lasers, extra corporeal lithotripsy, endoscopic scissors, polypectomy loops or electrohydraulic lithotripsy, but they usually require several long and wasteful sessions especially in cases of large bezoars. Consequently, a surgical procedure is the most appropriate treatment for these patients. These cases have low rates of morbidity and mortality, especially those who undergo a laparoscopic procedure which is the ideal approach (1, 2, 5, 10, 13). In the case we are reporting, due to the length and size of the bezoar, surgical treatment was required after failed endoscopic removal. Particularly striking in this case was the rapid formation of the bezoar as the patient had had an endoscopy 18 months before admission.

In the literature of the world on this topic there is not another report of this syndrome in a pregnant woman.

Consequently, there is no consensus agreement, proposal or recommendation for the therapeutic approach for these patients. However, given the need for extraction and the existing experience which has been reported for laparoscopies for pregnant women, laparoscopy is considered to be the ideal approach for these patients, although this also depends on the months of pregnancy and associated diseases peculiar to each patient.

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