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CLINICAL CASE

Rapunzel syndrome with double simultaneous trichobezoar in a teenager: Clinical Case Report

Síndrome de Rapunzel con doble tricobezoar simultáneo en una adolescente: Reporte de caso

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Abstract

Introduction: Trichobezoars are an intraluminal accumulation of ingested hair. The Rapunzel syndrome refers to the presence of gastric trichobezoars which extend to the small intestine together with trichotillomania and trichophagia, that occur predominantly in psychiatric patients of pediatric age. **Objective:** To analyze the clinical course and resolution of this syndrome in a case report. Likewise, we provide information about the family environment and psycho-emotional context of the patients and help the reader identify similar circumstances in their clinical practice. **Case report:** Female 14-year-old patient with history of trichotillomania and trichophagia of two years of evolution, who consulted for epigastric pain associated with weight loss, nausea, and postprandial fullness. During the physical examination, the patient was found to have bald patches in the scalp along with a palpable mass that seemed to be confined to the gastric limits. Imaging studies revealed gastric occupation due to a bezoar formation. The patient was treated surgically with laparotomy and gastrostomy, and two simultaneous trichobezoars were removed from the patient´s stomach and duodenum, the patient also underwent psycho-emotional professional counseling. **Conclusion:** Rapunzel´s syndrome, far for being a merely surgical entity, also requires psychoemotional assessment to prevent it recurrence and limit its severity.

Keywords:
Intestinal Obstruction;
Trichophagia;
Trichotillomania;
Trichobezoar

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Introduction

While the term bezoar in Western culture refers to a hidden treasure, for Islamic cultures its symbolism refers to a powerful antidote. On the other hand, in the peninsular Mayan tradition, the belief that bezoars provide good fortune to those who find them in the gastrointestinal tract of their prey, commonly, deer, is maintained[1]. However, outside the ethnic or symbolic context, in the field of human health, trichobezoars are the most common and they are made of indigestible masses of ingested hair that requires surgical management[2-4].

The Rapunzel syndrome is related to gastric trichobezoars which extend to the small intestine[5,6]. Trichobezoars are due to trichotillomania, the habit of pulling the hair, and to trichophagia, which corresponds to the act of eating hair[7]. The name of this syndrome is a reference to Rapunzel, the main character of a fairy tale of the same name, whose hair was held in a long and resistant thick braid.

Rapunzel syndrome appears mainly in young women, 90% of the cases are women and 80% of them are younger than 30 years[6,8,9]. Its low prevalence justifies the fact that it usually appears in other cases that commonly focus on organ compromise and resolution of acute symptoms[7,10,11]. Therefore, the presentation of this case, in addition to analyzing aspects related to the onset and management of this syndrome, also provides data about the family and psychoemotional environment that may be related to the disease, thus, in the future the risk can be identified in patients with similar circumstances through the analysis of these data.

Case report

Female patient, 14 years old, without a relevant surgical or pathological history, no language, intellectual or cultural barriers that could affect communication. The patient went to emergency service due to a two-week history of acute abdominal pain exacerbations; the pain was mainly in the epigastrium area, with irradiation to the mesogastrium and left hypochondrium, associated with a constant feeling of postprandial plenitude, nausea without vomiting and weight loss of 8.8 lb (4 kg) in fifteen days. Two weeks before, the patient had episodes of watery small-volume diarrhea, with an average of three stools a day between periods of constipation over 48 hours, without fever. She was treated as an outpatient with antispasmodics, prokinetic agents and antibiotics, she also reported that she took non-steroidal anti-inflammatories and opioids, which were given by a relative.

Since there was no progression, the patient was referred to a general hospital, where the presence of alopecic areas on the scalp and nail borders that indicated nail biting were recorded. During the abdominal auscultation, it was possible to observe an absence of peristalsis, tympanic percussion, and a stomach walls pattern was identified by palpating. The patient reported pain in the epigastrium and mesogastrium, there were no signs of peritoneal irritation.

In the laboratory tests, leukocytosis due to neutrophilia (86.60%) was identified and a reduction in the mean corpuscular hemoglobin (table 1). The simple radiography showed a ground glass image in the level of the gastric cavity (figure 1). The simple and contrasted computed axial tomography of the abdomen

Table 1. Pre-surgical and post-surgical hematic biometry results

Test	Pre-Surgery Results	Post-Surgery Results	Units	Reference values
Leukocytes	***13.80****	7.60	10 ^ 3 mm³	4.50-10
Neutrophils	***11.80****	4.90	10 ^ 3 mm³	*1.40-6.5*
Lymphocytes	1.40	1.70	10 ^ 3 mm³	1.20-3.4
Monocytes	0.50	0.70	10 ^ 3 mm³	0-0.70
Eosinophils	0.10	0.30	10 ^ 3 mm³	0-0.70
Basophiles	0.00	0.00	10 ^ 3 mm³	0-0.20
Erythrocytes	5.60	3.78	10 ^ 6 mm³	3.50-5.70
Hemoglobin	15.30	10.00	Gr/dl	10-15
Hematocrit	45.90	30.70	%	30-46
Corpuscular Volume	80.90	81.20	Fl	80-99
Median Corpus. Hgb	***27.00****	***26.40****	Pg	28-32

Data outside the normality ranges are shown in italics, bold and*; Vol = Volume; Hgb = Hemoglobin.

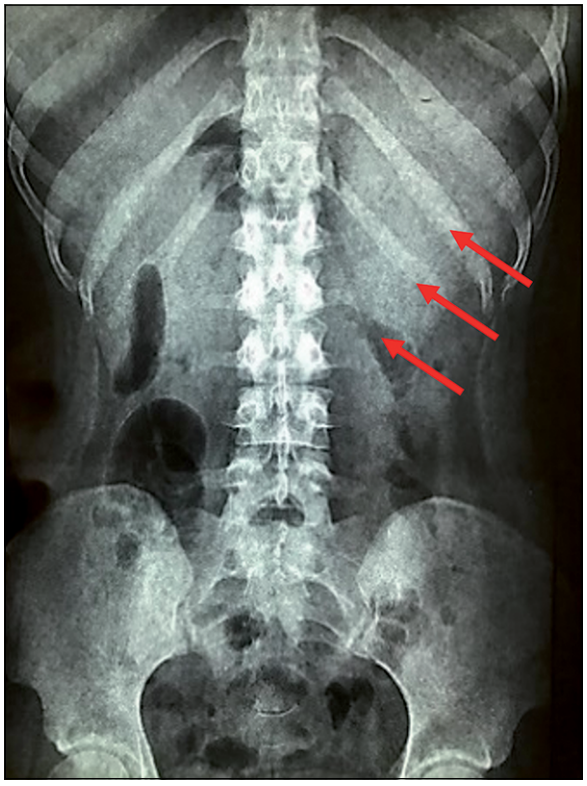

Figure 1. Simple radiographic image resembling a "stained glass" in gastric chamber level.

showed a dilatation at the level of the pylorus due to the obstruction of the gastric content by an obstructive mass (figure 2). In addition, it was also observed a dilation of the intestinal loops with hydro-aerial levels (figure 3) and the complete occupation of the gastric cavity by the trichobezoar (figure 4 and 5).

The management plan included the surgical extraction and examination by the psychiatry service. After the laparotomy and gastrostomy, two simultaneous

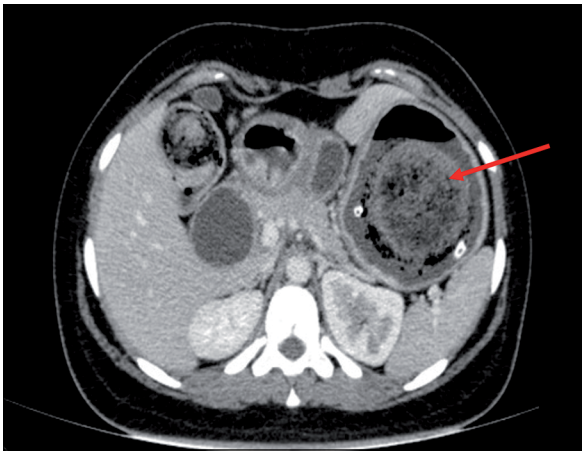

Figure 2. Barium-Contrasted axial tomographic image, where an ovoid image containing gas can be seen. The ovoid image is not enhanced by contrast medium, and similar images can be also observed next to the liver and jejunum.

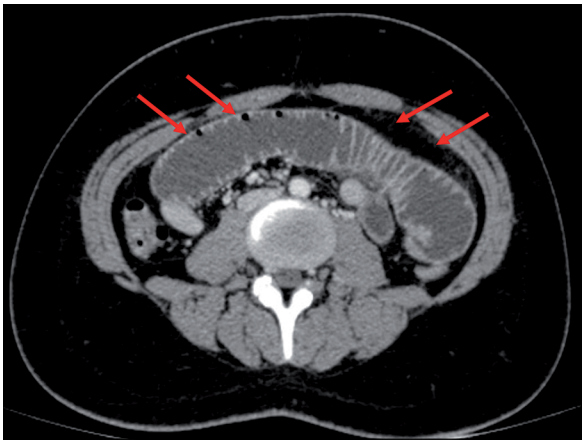

Figure 3. Barium-Contrasted axial tomographic image. Small bowel image shows dilation and a hydro aerial level is found, due to intestinal occlusion.

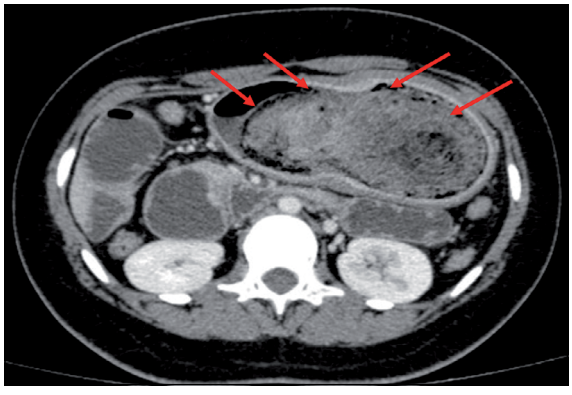

Figure 4. Barium-Contrasted axial tomographic image. Intraluminal total occlusion image is found, due to the presence of bezoar.

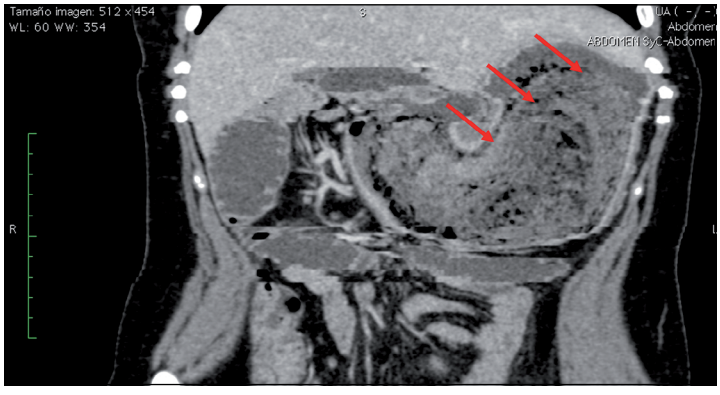

Figure 5. Barium-Contrasted axial tomographic image. Intraluminal total occlusion image is found, due to the presence of bezoar.

trichobezoars were identified: the first one was located in the stomach and duodenum (11.8 x 1.9 inches – 30 x 5 cm) and the second one in the intestines, 15.7 into the ligament of Treitz (1.9 x 1.1 inches – 5 x 3 cm). The patient was discharged 72 hours after the surgery.

During the psychiatric follow-up, it was possible to initiate an effective communication with the patient, who mentioned that she had good family and affective relationships, however, she mentioned that she was going through in what she defined as "constantly tense situation" with her biological parents due to the difficult relationship between them and towards her. The patient denied suicidal, self and hetero injuring thoughts.

Two years before, the patient developed to trichotillomania and trichophagia, as well as non-specific gastrointestinal distress. Back then, the patient was diagnosed with depression and anxiety, she was treated with a selective inhibitor of serotonin reuptake in order to solve the trichotillomania. The psychiatric follow-up and the adherence to the pharmacological treatment were suboptimal and there was no warning of the possible inherent risk of continuous hair intake, which was part of her disorder. The patient temporarily linked the onset of her disorder (trichotillomania) with the periods of high tension in her home.

The psychiatric and then psychologic evaluation, was consistent in both cases, since the appetite for pulling the hair, then the feel of putting it in the mouth and finally swallowing it (*verbatim* as explained and expressed by the patient with non-verbal language), they considered it as a pathological searching for a socially atypical, pleasant feeling associated with a presumably hostile environment, therefore her conduct was discarded as a behavior related to an anxiety disorder.

Currently, the patient continues the treatment without anxiolytic drugs, and she was also discharged by psychiatry. She is still under monitoring by psychologists, her prognosis is favorable as her has broadly understood the risk of continuing her previous behavior.

Discussion

The presentation of this case allowed an approach to the familiar and psycho-emotional environment underlying trichotillomania, with trichophagia, which also produced the double simultaneous trichobezoar. This surgical finding is uncommon, in relation to other cases, where the presence of multiple trichobezoars is limited. Recurrent trichobezoar is more common than multiple ones[12].

The patient presented most of the signs and symptoms found in clinical symptoms of intestinal obstruction, however, the alopecic areas were an indicator of trichotillomania and possible trichophagia. The typical clinical symptoms supported the diagnostic suspicion, thus, even when the neutrophilia in the white series was not suggestive for this type of pathology, the computed tomography confirmed the suspicion[7,13].

The surgery was the chosen therapeutic method in this case, even when the literature suggests that the treatment to remove trichobezoars has not been standardized, cases with multiple trichobezoars that were treated with surgery were successful[14-16].

Recurrence prevention, which affects 20% of the patients, it is important to consider the psycho-emotional environment of the patient. In a case study of Frey et al., the reaction of the patient towards the dysfunctional relationship between the parents was identified as the triggering factor of trichotillomania[17].

Although the exact proportion of cases of trichotillomania along with trichophagia is unknown, it is estimated that half of the patients with trichophagia develop trichobezoars, therefore, it is important to identify this risk in patients with trichotillomania. The gastrointestinal obstruction could have been prevented if the patient and family were warned since there were no intellectual or maturity limitations that could hinder the comprehension of the consequences of her behavior[15,18].

The psychiatric treatment of trichotillomania should not be considered as optional since it is important to identify the main determinants associated with the disorder in each case because the integral treatment depends on it[19-21].

In conclusion, this case showed that the treatment of Rapunzel syndrome does not end with the surgical extraction of the trichobezoar, it is required to explore underlying psycho-emotional causes of trichotillomania and to determine the importance of advising patients and relatives about the risks of the relapse which can help control the recurrence.

Ethical responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights 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 correspondence author.

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Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

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