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CASE PRESENTATION

Rapunzel syndrome: psychological bondage and organic simulation

Síndrome de Rapunzel: atadura sicológica y simulación orgánica

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Abstract

Introduction. Rapunzel syndrome is an uncommon condition that manifests as trichobezoar, which are hair bundles in the stomach or small intestine that can mimics other surgical illnesses. Multiple complications can arise from delayed diagnosis and treatment.

Clinical case. A 10-year-old female patient with trichotillomania and trichophagia, with abdominal pain and nonspecific symptoms of intestinal obstruction of eight months of evolution. Physical examination revealed epigastric tenderness and a solid mass was palpable in the mesogastric and epigastric region. An abdominal ultrasound showed gastric trichobezoar that extended into the small intestine. A gastrotomy was performed and the trichobezoar was extracted with satisfactory evolution of the patient. The comprehensive approach allowed knowing the psychological bond due to possible child abuse**.**

Results. The patient had a satisfactory evolution and was discharged on the fifth day of hospitalization. She is currently being monitored by psychology, child psychiatry and pediatrics.

Discussion. This clinical case highlights the importance of recognizing situations that seldom present in pediatrics, which may have a psychological aspect due to the presumption of child abuse, and which present as a recurrent organic condition simulating other frequent abdominal pathologies in childhood; all of which may lead to an unwanted outcome due to diagnostic delay.

Conclusion. The comprehensive management of the pediatric patient is mandatory to recognize situations of presumed child abuse, in the face of a recurrent surgical conditions.

Keywords: trichotillomania; bezoars; stomach; duodenal obstruction; differential diagnosis; child psychiatry.

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Resumen

Introducción. El síndrome de Rapunzel es una entidad infrecuente, que se presenta como un tricobezoar a causa de una aglomeración de cabello acumulado dentro del tracto gastrointestinal, por lo que simula otras patologías quirúrgicas.

Caso clínico. Paciente femenina de 10 años de edad, con tricotilomanía y tricofagia, dolor abdominal y síntomas inespecíficos de obstrucción intestinal de ocho meses de evolución. Al examen físico se encontró abdomen con distensión y masa palpable en epigastrio y mesogastrio. La ecografía permitió hacer el diagnóstico de tricobezoar gástrico extendido hasta el intestino delgado, por lo que se llevó a cirugía para gastrotomía y se extrajo el tricobezoar, con evolución satisfactoria de la paciente. El abordaje integral permitió conocer la atadura sicológica por posible maltrato infantil.

Resultado. La paciente tuvo una evolución satisfactoria y se dio egreso al quinto día de hospitalización. Actualmente se encuentra en seguimiento por sicología, siquiatría infantil y pediatría.

Discusión. El caso clínico denota la importancia en reconocer situaciones de presentación infrecuente en pediatría, que puedan estar asociadas a alteraciones sicológicas o presunción de maltrato infantil y que se presenten como una condición orgánica recurrente que simule otras patologías abdominales frecuentes en la infancia. El retraso diagnóstico puede conducir a un desenlace no deseado con complicaciones.

Conclusión. Se hace mandatorio el manejo integral del paciente pediátrico y aumentar la sensibilidad para reconocer situaciones de presunción de maltrato infantil, sobre todo en pacientes con una condición orgánica quirúrgica recurrente.

Palabras claves: tricotilomanía; bezoares; estómago; obstrucción duodenal; diagnóstico diferencial; siquiatría infantil.

Introduction

Trichobezoar is a clump of hair that accumulates and remains within the gastrointestinal tract. They are formed because human hair is resistant to digestion and intestinal peristalsis due to its smooth surface. The decrease in friction and propulsion of the bezoar with the gastric contents leads to the hair becoming intertwined, forming clumps, with retention in the folds of the upper digestive tract.

Gastric trichobezoar is an extremely rare pathology [1], occurring in 0.06 to 4% of the population [2]. When it extends to the duodenum or jejunum, it is known as Rapunzel syndrome. Rapunzel syndrome is a rare entity, observed mainly in female pediatric school patients who have some emotional disorder [3-5].

Patients with bezoars usually manifest nonspecific gastrointestinal symptoms such as abdominal pain, anorexia, and vomiting [6,7]. The most frequent presentation is a condition of intestinal obstruction, although rare, it has been reported that 41% develop complications secondary to trichobezoar [5], gastric perforation (10%), intestinal intussusception (1.8%) or cholangitis (less than 0.9%) can occur [4,6,8]. The objective of this article was to present the clinical case of a pediatric patient, with a five-year history of trichotillomania and trichophagia, which evolved into Rapunzel syndrome, and the comprehensive approach that allowed the identification of a psychological affectation.

Clinical case

A 10-year-old female patient was brought to the emergency department at the Napoleón Franco Pareja Children's Hospital - Casa del Niño, in Cartagena de Indias, Colombia, due to an eight-month history of a mass in the epigastrium, associated with morning food vomiting, an average of two

moderate amounts a day, and diffuse abdominal pain, of moderate intensity according to the Wong-Baker scale [9], which increased with food intake. In the physical examination on admission, normal vital signs were found, weight of 27.6 kilos and height of 131 cm (Z -1), for a body mass index (BMI) of 16 kg/m^2 (Z -1 and 0), areas of alopecia in the fronto-temporo-parietal region, abdomen with distention and palpable mobile mass in the epigastrium and mesogastrium, measuring 7x12 cm and hard consistency, in addition to a wall defect at the right inguinal level, with reducible, isothermal and painless content.

A total abdominal ultrasound was ordered, which revealed a gastric trichobezoar that occupied the entire gastric chamber. The hemogram and blood biochemistry were normal. Upper digestive tract endoscopy confirmed the diagnosis of trichobezoar, which almost occupied the entire gastric chamber, with obstruction of the pylorus and involvement of the second portion of the duodenum.

It was decided to perform an exploratory laparotomy where distension was evident in the gastric chamber and small intestine. During the gastrotomy, a trichobezoar approximately 12 centimeters long was identified and removed, extending from the lower third of the esophagus to the second portion of the duodenum (Figure 1). After verifying the absence of residual trichobezoar, gastrorrhaphy was performed.

The patient and her family received support from psychology and child psychiatry, who found the behavioral link with the organic commitment through the story of the patient's mother, who reported on the state of family breakdown due to domestic violence and possible sexual abuse of the minor by a close relative. The care route for possible child abuse and sexual violence was triggered, which included infectious profile



Figure 1. Gastric trichobezoar. A. Trichobezoar was found occupying the entire gastric chamber and extending to the second portion of the duodenum. B. The trichobezoar produced an obstruction that required urgent removal.

laboratories with serology for syphilis, hepatitis B virus, herpes type II, acquired immunodeficiency virus, and *Neisseria gonorrhoeae*, which were negative. The mother stated that she had known about the patient's habit of trichotillomania and geophagia since she was 5 years old. Mental health performed psychoeducation, cognitive-behavioral therapy and operant behavior reduction techniques.

The patient had a satisfactory evolution and was discharged on the fifth day of hospitalization. She is currently being monitored by psychology, child psychiatry and pediatrics.

Discussion

The patient in this clinical case did not have an apparent premorbid personality that would explain the compulsive behavior of trichotillomania and trichophagia. The chronic evolution of the patient is relevant, with the compulsive behaviors that led to the occurrence of the gastrointestinal condition. The clinical presentation initially simulated other clinical conditions, which were addressed by the treating physicians in the process prior to the consultation at the final care institution, where the presenting condition was initially addressed as abdominal pain without surgical cause.

It has been reported that patients with bezoars present among the symptoms abdominal pain and vomiting (88%) or abdominal distension (47%). Uncommon manifestations that may delay diagnosis have also been described, such as sepsis symptoms attributed to intestinal intussusception. In our case, the psychological condition due to separation anxiety disorder was possibly aggravated by the COVID-19 pandemic [10]. Another atypical presentation in adolescents with depression has been the finding of signs of peritoneal irritation due to perforation on the anterior surface of the stomach [11].

Although the average age of presentation is 7 years, with a predominance in the female sex (14 versus 3 in men) [5], it has also been reported in adults with strange manifestations such as hematemesis due to gastric perforation, localized peritonitis, coagulopathy, severe anemia and liver failure [12].

Diagnostic modalities include ultrasound, computed tomography (CT), and upper gastrointestinal endoscopy. An echogenic mass can be seen on ultrasound, but the accuracy and precision of this in such cases is not so high, while CT is the diagnostic tool of choice due to its high percentage of precision. Endoscopy checks for agglomeration of hair, which may be fetid due to the decomposition and fermentation of fats [4,7,10].

Treatment of trichobezoars usually involves endoscopic or surgical removal, via gastrotomy, and psychiatric intervention. In the absence of adequate treatment, the associated mortality rate is up to 30%, mainly due to complications due to gastrointestinal bleeding, obstruction or perforation [11]. In the study by Mirza [5] with 17 cases, more unusual complications occurred such as intussusception and intestinal necrosis, small bowel obstruction, intestinal perforation with massive rectal bleeding, and transient acute pancreatitis.

Another representation noted in the literature is trichosicodermatology, which reports that the behavior of trichotillomania and trichophagia habits is generally accompanied by states of depression and anxiety, and may be part of an obsessive-compulsive disorder [13,14]. Trichotillomania can mean and simulate conditions of presumptive child abuse [15], which if not recognized promptly can even lead to death [16-18]. However, it is also important to recognize situations that suggest trichotillomania, when in reality they are cases of abusers who pull children's hair, making people believe that it is a compulsive behavior of the minor [19,20].

The patient in the clinical case had a chronic condition of five years of evolution, with a presumption of unresolved child abuse, conditioned by a habit of trichotillomania and trichophagia, in which the origin of alleged child abuse that would explain the behavior of the patient was not psychological determined. On the other hand, the similarity with other organic pathologies that are accompanied by manifestations such as abdominal pain, emesis and weight loss, limited the timely

diagnosis. The comprehensive management in the care route of possible child abuse allowed the approach and protection of the minor in possible situations of transgression of their rights.

Conclusion

The review of the literature and the experience of the clinical case reported, direct us towards a comprehensive evaluation and management of the pediatric patient, especially when we face a surgical condition that involves inappropriate psychological behavior, as occurs in Rapunzel syndrome, which allow us to recognize non-obvious situations of presumed child abuse.

Compliance with ethical standards

Informed consent: Written informed consent was obtained from the patient's mother for publication of this case and images.

Conflict of interest: The authors of the study have no conflicts of interest to declare.

Use of artificial intelligence: The authors declared that they did not use artificial intelligence (AI)-assisted technologies (such as large language models, chatbots, or image creators) in the production of this work.

Fuding: Authors' own resources.

Author's contributions

- Conception and design of the study: Pastor Alexander Thomas-Olivares, Camila Andrea Otero-Payares, Dilia Fontalvo-Rivera.
- Acquisition of data: Camila Andrea Otero-Payares, Dilia Fontalvo-Rivera.
- Data analysis and interpretation: Pastor Alexander Thomas-Olivares, Camila Andrea Otero-Payares, Dilia Fontalvo-Rivera.
- Drafting the manuscript: Pastor Alexander Thomas-Olivares, Carlos Andrés López-López, Manuel Sebastián Mosquera-Cárdenas, Dilia Fontalvo-Rivera.
- Critical review and final approval: Camila Andrea Otero-Payares, Dilia Fontalvo-Rivera.

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