ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

https://ijrsms.com

International Journal of Recent Surgical and Medical Sciences

Case Report

Rapunzel Syndrome with Gastric Perforation: A Rare Case Report in a Three-year-old Child

Chhavindar Singh[1], Vinod Kumar Pandey[1], Prakhar Pratap[1]

[1]Department of General Surgery, Moti Lal Nehru Medical College, Prayagraj, Uttar Pradesh, India

***Corresponding author:**
Dr. Chhavindar Singh, Department of General Surgery, Moti Lal Nehru Medical College, Prayagraj, Uttar Pradesh, India.

bebosingh111@gmail.com

Received: 22 May 2023

Accepted: 30 August 2024

EPub Ahead of Print: 20 December 2024

Published: 23 January 2025

DOI
10.25259/IJRSMS-2023-4-4

Quick Response Code:

ABSTRACT

Rapunzel syndrome is a rare variant of trichobezoar in the stomach extending from the stomach into the small intestine causing a bowel obstruction and commonly associated with an underlying psychiatric disorder. The term comes from a story written by the Grimm brothers in 1812 about Rapunzel who was a long-haired maiden. We hereby, present the case of a 3 years old boy with Rapunzel syndrome due to a very large gastric trichobezoar. An abdominal computed tomography scan showed a heterogeneous mass occupying the whole stomach cavity with extension into the jejunum. A large dense mass of hair extending up to jejunum was removed. Following surgery, psychiatric consultation was sought to prevent recurrence. Trichobezoar as an entity should be considered in the differential diagnosis of abdominal pain and non-tender abdominal mass in young patients. A thorough assessment of psychiatric history is mandatory to address the underlying disease to prevent recurrence.

Keywords: Rapunzel syndrome, Trichotillomania, Trichobezoar, Bezoar

INTRODUCTION

A trichobezoar is a hair ball that forms in the proximal gastrointestinal tract (GI), and it is an extremely unusual illness that practically never affects anyone older than 30 years old.[1] There have been numerous reports of gastric trichobezoars published in the form of case studies and series. The majority of these studies describe the condition as abdominal masses, with or without the characteristics of intestinal obstruction. The term 'Rapunzel syndrome' describes a condition in which a trichobezoar has extended at least as far as the jejunum and is responsible for obstructing the intestines.[2]

Despite the fact that the name derives from the context of a fairy tale, the frequently surrounding issues might give it the connotation of a hairy tail or even a terrifying tail.[3] Trichobezoar most frequently manifests itself in young women, and these patients almost always have a pre-existing mental health condition. The low index of suspicion shown by the treating physician accounts for the late presentation of the vast majority of trichobezoar cases.[4]

Because the clinical picture in the early stages is typically very limited, diagnosis is frequently delayed as a result. Therefore, an awareness of trichotillomania, also known as the urge to pull out one's own hair, in conjunction with trichophagia, also known as swallowing hair, is required for an early diagnosis of trichobezoar in women who have psychiatric comorbidity. This is because these two conditions are associated with one another. In cases when the trichobezoar is not detected

in time, it continues to gain weight and expand in size until it entirely occupies the stomach. This places the patient at an elevated risk for gastric mucosal erosions or ulcerations and potentially gastrointestinal perforation.[1]

Additionally, sections of the trichobezoars' tail may rupture and migrate into the small intestine, which can result in severe consequences such as intestinal blockage, perforation, and peritonitis in advanced instances. These complications can be avoided by early detection and treatment.[5] Other complications, such as protein-losing enteropathy, intussusception, obstructive jaundice, pancreatitis, or even death in cases of unrecognised bezoars or delayed diagnosis, have also been reported in patients who have trichobezoars.[6–8]

There are less than 40 cases recorded in the literature, and the purpose of psychiatric treatment is to prevent future occurrences of the condition. In the cases reported in the medical literature, children with Rapunzel syndrome have been diagnosed at ages as young as three.[9]

CASE REPORT

A three-year-old male child presented with multiple episodes of fever, pain abdomen, intermittent vomiting, and abdominal distension for two days.

The patient did not have a history of type 1 diabetes mellitus, congenital defects, delay in milestones, and more. The baby had a history of pica disorder since he was two years old. The baby was immunised completely to date and was born normally via vaginal delivery, and was full term.

The patient presented with a sudden onset of vomiting for two days, with pain being intermittent in character. The vomiting precludes eating and consisted of gastric content and was projectile in nature. The vomiting was associated with pain and altered bowel habits.

It aggravated after eating, and the pain was relived after vomiting. The pain was sudden in onset for the past one day and progressive in nature. It was colicky in nature and severe in intensity. The pain was associated with constipation and generalised in terms of the site. The pain was relived after vomiting.

The patient also had a complaint of abdominal distension for two days, which was acute in onset and diffuse in the abdomen.

Clinical examination

The baby looked toxic and poorly nourished, with weight and height at 60th percentile with normal growth curves. The blood pressure was normal at 90/68 mm Hg, and the weight was 7 kg.

The muscle co-ordination was normal for age. Muscles were normal, and there was a normal growth curve. On inspection, the abdomen was distended, and the umbilicus was central and everted with tenderness, rigidity, and guarding present. Percussion of the abdomen had a tympanic note. On auscultation, no bowel sounds were heard.

Investigations

Ultrasonography (USG) showed that the stomach was overly distended, filled with thick hypoechoic contents and wall thickening. There is a focal area of breach in the lesser curvature of the stomach with minimal leak of fluid and air in the epigastric space. Similar contents are seen extending into the duodenum with wall thickening. The findings are suggestive of gastric and duodenal bezoar with contained gastric perforation.

Laboratory results showed anemia (hemoglobin 8.3 gm/dl) and leucocytosis (total leucocyte count is 17,000 c/mm3). Liver function tests were normal, except that aminotransferase (AST) was 600 IU/L. Kidney function tests were normal, with no derangement.

Management

The patient was managed operatively by emergency laparotomy with midline incision, with the surgical removal of trichobezoar via anterior gastrotomy incision. Intraoperative findings showed 1×1 cm perforation in lesser curvature with gross distension of the stomach. Closure of the gastrotomy incision and primary perforation site was done using the two-layered 3-0 polyglactin 910 suture. Peritoneal lavage was done, and the rest of the bowel inspected was found to be normal. The specimen was sent for histopathological examination.

Further, the patient was managed conservatively by intravenous fluids, antibiotics, analgesics and other supportive measures. The postoperative period was found to be uneventful. Oral feeds were resumed on day two, and the patient was subsequently discharged on day five.

The patient was given postoperative psychological evaluation and treatment.

The respective intraoperative images have been attached hereof as Figures 1, 2, and 3.

DISCUSSION

Trichobezoars are the most prevalent type of bezoar found in humans. They typically appear in preteen or teenage girls who have psychiatric co-occurring disorders or developmental delay.[10,11] Nevertheless, trichobezoars are also possible in males, as reported by Khanna K *et al.* describing to the

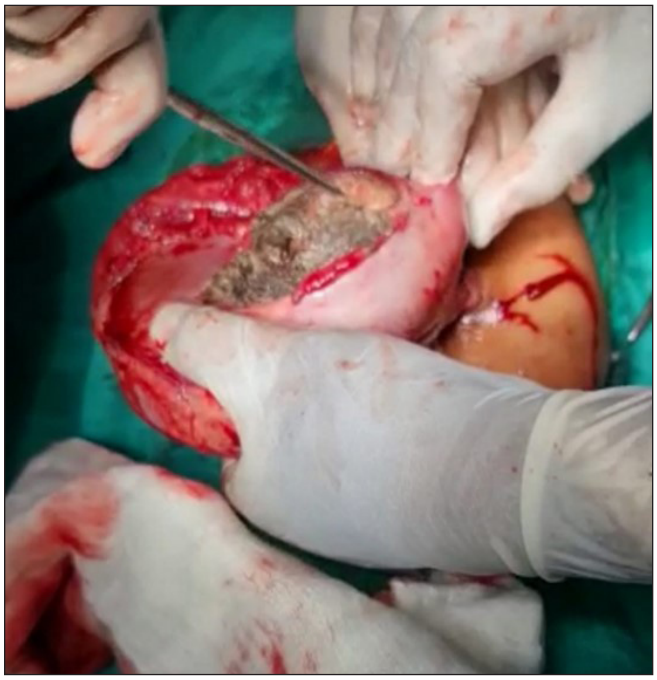

Figure 1: Gastrotoy revealed large trichobezoar.

Figure 2: Removal of trichobezoar through gastrotomy.

best of our knowledge, the youngest patient diagnosed with trichobezoar – a three-year-old boy[5] – similar to the case we describe.

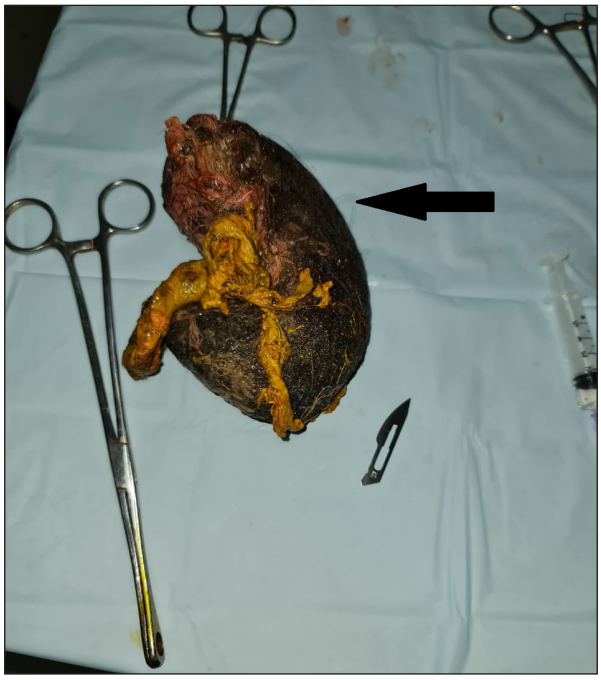

Figure 3: Trichobezoar.

In 1968, Altonbary AY *et al*. were the first to describe the condition known as Rapunzel syndrome. This condition is characterized by a stomach trichobezoar that has a tail that extends to the jejunum, ileum, or ileocecal junction.[6] The enormous quantity of hairs become tangled with one another and take the form of a stomach as a result. When a patient most frequently presents as an abdominal mass with characteristics that are indicative of intestinal obstruction, doctors generally suspect a malignant etiology until the suspicion is disproved by imaging. The majority of the time, victims are adolescent girls who suffer from trichotillomania or trichophagia.[12]

In our patient's instance, the symptoms first appeared at a very young age and were caused by hair that had grown all the way down to the small bowel. Due to their non-specific presentation or even the absence of symptoms in the early stages, the majority of trichobezoars may not be detected during the early stages of the condition. Due to the fact that a history of trichophagia is rarely obtained unless the patient is particularly probed about their condition, it is rarely kept as a differential diagnosis on clinical examination alone.[2]

A palpable abdominal mass was detected in 87.7% of the 131 trichobezoar cases that were collected over the course of this study. Other common manifestations included abdominal discomfort (70.2%), nausea and vomiting (64.9%), weakness and weight loss (38.1%), constipation or diarrhea (32%), and haematemesis (6.1%).[4]

The Rapunzel syndrome can cause a variety of consequences, including bouts of incomplete pyloric blockage, full

obstruction of the intestine, perforation, peritonitis, and even death.[9] Trichobezoars have recently been implicated as a potential cause of jejunal intussusception, appendicitis, nephrotic syndrome, and biliary obstruction, according to research published in medical journals.[13]

In virtually all instances, endoscopy can be diagnostic, although ultrasound does not have much to give in the way of diagnostic potential. A contrast-enhanced computed tomography (CT) scan will be able to delineate the extension of the trichobezoar. Endoscopic removal, dissolving, and mechanical fragmentation with a hydro jet are some of the treatment options for stomach trichobezoars. However, surgical removal is the only choice for larger masses that cause discomfort and extend into the small bowel.[2]

CONCLUSION

The Rapunzel syndrome is an extremely uncommon condition that can produce gastrointestinal symptoms in children. These can include abdominal pain, anorexia, or weight loss. When children present with digestive problems, celiac disease is rarely evaluated as a possible cause in the differential diagnosis. As a result, a delayed diagnosis is a rather typical occurrence. It is essential for an early diagnosis that there is increased knowledge regarding risk factors for trichotillomania and trichophagia. Some of these risk factors include developmental delay as well as anaemia as a source of pica. In most cases, the removal of big bezoars via laparotomy is the recommended course of treatment. Because of this, Rapunzel syndrome, despite the fact that it is very uncommon, should never be forgotten because its early detection prevents the onset of future difficulties.

Ethical approval

The research/study complied with the Helsinki Declaration of 1964.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of AI-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Gorter RR, Kneepkens CM, Mattens EC, Aronson DC, Heij HA. Management of Trichobezoar: Case Report and Literature Review. Pediatr Surg Int 2010;26:457–63.
2. Gupta A, Mittal D, Srinivas M. Gastric Trichobezoars in Children: Surgical Overview. Int J Trichology 2017;9:50–3.
3. George SV, Samarasam I, Mathew G, Chandran S. A Hairy Tail not a Fairy Tale – Rapunzel Syndrome. Indian J Surg 2013;75:80–1.
4. Al Wadan AH, Al Kaff H, Al Senabani J, Al Saadi AS. 'Rapunzel Syndrome' Trichobezoar in a 7-year-old Girl: A Case Report. Cases J 2008;1:205.
5. Khanna K, Tandon S, Yadav DK, Khanna V. Rapunzel Syndrome: A Tail too Long to Tell! BMJ Case Rep 2018;2018: doi :10.1136/bcr-2018-224756
6. Altonbary AY, Bahgat MH. Rapunzel Syndrome. J Transl Int Med 2015;3:79–81.
7. Naik S, Gupta V, Naik S, Rangole A, Chaudhary AK, Jain P, *et al*. Rapunzel Syndrome Reviewed and Redefined. Dig Surg 2007;24:157–61.
8. Koç O, Yildiz FD, Narci A, Sen TA. An Unusual Cause of Gastric Perforation in Childhood: Trichobezoar (Rapunzel Syndrome). A Case Report. Eur J Pediatr 2009;168:495–7.
9. Mohite PN, Gohil AB, Wala HB, Vaza MA. Rapunzel Syndrome Complicated with Gastric Perforation Diagnosed on Operation Table. J Gastrointest Surg 2008;12:2240–2.
10. Aybar A, Safta AM. Endoscopic Removal of a Gastric Trichobezoar in a Pediatric Patient. Gastrointest Endosc 2011;74:435–7.
11. Michail S, Nanagas V, Mezoff AG. An Unusual Cause of Postfundoplication Vomiting. J Pediatr Surg 2008;43:E45–7.
12. http://fyra.io. (Jejunal Trichobezoar in a 3-year-old Boy; Practice Path MD Pro Tip: Architects. Practical Dermatology. Available from: https://practicaldermatology.com/articles/2017-may/jejunal-trichobezoar-in-a-3-year-old-boy-practice-path-md-pro-tip-architects). https://practicaldermatology.com/articles/2017-may/jejunal-trichobezoar-in-a-3-year-old-boy-practice-path-md-pro-tip-....
13. Nwankwo E, Daniele E, Woller E, Fitzwater J, McGill T, Brooks SE. Trichobezoar Presenting as a Gastric Outlet Obstruction: A Case Report. Int J Surg Case Rep 2017;34:123–5.

How to cite this article: Singh C, Pandey VK, Pratap P. Rapunzel Syndrome with Gastric Perforation: A Rare Case Report in a Three-year-old Child. Int J Recent Surg Med Sci. 2024;10:125-8. doi: 10.25259/IJRSMS-2023-4-4