

Case Report

Every tangle has a story - Rapunzel Syndrome: A case report

Yousuf Aziz Khan,^{*1} Zainab Zaib,¹ Fizza Tufail,¹ Muhammad Kashif Rafiq,¹ Khyal Muhammad,²

1 Department of Surgery, Surgical Unit A, Ayub Teaching Hospital – Medical Teaching Institute (MTI), Abbottabad

2 Department of Paediatrics, Unit A, Ayub Teaching Hospital – MTI, Abbottabad

Cite as: Khan YA, Zaib Z, Tufail F, Rafiq MK, Muhammad K. Every tangle has a story - Rapunzel Syndrome: A case report. J Pediatr Adolesc Surg. 2024; 2: 109-11.

This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited (<https://creativecommons.org/licenses/by/4.0/>).

ABSTRACT

Background: Rapunzel syndrome is the rarest form of trichobezoar; a condition in which trichobezoar extends beyond the stomach into the small intestine. It is common among children and young girls with a history of psychiatric illnesses.

Case Presentation: A 10-year-old girl without any history of psychiatric illness, presented with abdominal pain and non-bilious vomiting for 6 months. Clinically she had a non-tender upper abdominal mass which, later at workup, was found to be a trichobezoar. She was surgically managed successfully.

Conclusion: Though uncommon, Rapunzel syndrome should be kept in the differential diagnoses especially in a young female patient with features of an upper GI obstruction and a non-tender, palpable epigastric mass.

Keywords: Rapunzel syndrome, Trichobezoar, Epigastric mass, Paediatric.

INTRODUCTION

Bezoars are compact masses of undigested foreign objects in the gastrointestinal tract. Trichobezoar is a dense ball of swallowed hair and constitutes around 6% of bezoars.[1] Rapunzel syndrome (RS) is a rare type of trichobezoar; the condition in which trichobezoar extends beyond the stomach into the small intestine.[2] Children and young girls with a history of psychiatric illnesses like pica syndrome/trichotillomania, anxiety disorders, depressive illnesses, and obsessive-compulsive disorder are usually affected in this condition. [3,4]

Clinically it presents with a variety of symptoms ranging from loss of appetite and a palpable painless mass in the abdomen to weight loss with a prolonged history of abdominal pain, vomiting, obstruction, and even perforation. It can also present with complications such as gastric ulceration, intestinal obstruction/perforation, intussusception, cholestatic jaundice, and acute pancreatitis, etc. [5-7]

The diagnostic tools are CT scan abdomen and upper GI endoscopy; the latter being the gold standard. Treatment options include endoscopy or surgery either laparoscopic

or open laparotomy. [5,7] We report the presentation and management of a 10-year old girl with Rapunzel syndrome.

CASE REPORT

A 10-year old girl was brought to the out-patient clinic with a 6 months history of abdominal pain and non-bilious vomiting. According to the mother, the patient also had anorexia, off and on irregular bowel habits, and upper abdominal distension during this period.

Clinically, her general physical examination was unremarkable except for mild pallor. Abdominal examination revealed a palpable, non-tender epigastric mass. Her laboratory workup showed a hemoglobin level of 10.6g/dL and a normal biochemical analysis including normal serum electrolytes, renal and liver function tests. Ultrasound abdomen showed an echogenic shadowing material in the gastric lumen suggesting the presence of bezoar. To confirm the diagnosis, an abdominal computed tomography (CT) scan with intravenous and oral contrast was performed. CT scan findings were suggestive of Rapunzel syndrome; showing a large hypodense filling defect with mottled air lucencies in the stomach, extending up to the proximal jejunum (Fig. 1).

Correspondence*: Dr. Yousuf Aziz Khan, Assistant Professor – Paediatric Surgery, Surgical Unit A, Ayub Teaching Hospital – MTI, Abbottabad.

Email: dr_yousufaziz@yahoo.com

Submitted on: 22-10-2021

© 2020, Khan et al,

Accepted on: 23-01-2022



Figure 1: CT Abdomen: showing a large hypo-dense filling defect with mottled air lucencies in the stomach

After optimization, an elective laparotomy was performed through an upper midline incision. Anterior gastrotomy was done and a large trichobezoar with a tail extending up to proximal jejunum was removed in-toto (Fig. 2). Double-layered primary gastric closure was performed and the abdomen was closed in layers. The postoperative course was smooth. She was allowed oral feeds on the 5th post-op day which were tolerated well. During her hospital stay, an initial psychiatric evaluation was also done, and was advised follow-up. The patient was discharged home on the 7th postoperative day. Unfortunately, they never showed up in person although, on a telephonic follow-up, she was doing well with no major complaints.



Figure 2: The extracted Trichobezoar mass

DISCUSSION

Rapunzel Syndrome (RS) is a rare trichobezoar affecting mainly young females. The first case of RS was described

REFERENCES

- De Melio J, Debrouwere T, Herman M. Trichobezoar. J Belg Soc Radiol. 2021; 105:31.
- Mouffak A, Salam S, Fahl A, Chbani K, Laoudiyi D, Ouzidane L. Rare cause of mechanical occlusion in children: a case report. Pan Afr Med J. 2021; 38:71.
- Reilly L. Interesting cases in Pediatric Radiology: ingestions, insertions, and aspirations. J Radiol Nurs. 2021; 40:14-22.
- Mazzei A, Centonze A, Aloï IP, Bertocchini A, Baldassarre E. Rapunzel Syndrome: endoscopy,

in 1968. [8] As per a recent review of the English literature, 40 cases have been reported until now. [9]

In the majority of cases, it has been associated with psychiatric illnesses but in our case, no such history was found; an initial psychiatric evaluation was also normal. A similar case without a background psychiatric illness was reported by Antunes et al. [10]

An ultrasound abdomen may show echogenic gastric shadows just like our case but cannot be used as a definitive diagnostic tool as its efficacy is 88% to 93%. Endoscopy is the gold standard imaging and treatment modality. CT scan with contrast is 90% sensitive and 57% specific and it delineates the extension of entangled hair as shown by the majority of cases. [11] We opted for a CT abdomen for definitive diagnosis as pediatric endoscopy was not available at our center.

The treatment of RS is the removal and depends upon the size and extension of the hair mass. The most commonly used modalities are endoscopic and surgery either laparoscopic or open approaches. Small-sized bezoars can be removed successfully with an endoscope. According to the literature, the success rate with laparoscopy is 75%. Success rate with a laparotomy and extraction is 99%; the mass can be removed in total making it the treatment of choice especially in RS. [7, 12]

In summary, though uncommon, Rapunzel syndrome should be kept in the differential diagnoses especially in a young female patient with and without any psychiatric illness who presents with features of an upper GI obstruction and a non-tender, palpable epigastric mass. Surgical removal is the gold standard and a long-term follow-up combined with a psychiatrist's support is recommended to prevent a recurrence.

Conflict of Interest: Nil

Source of Support: Nil

Consent to Publication: Author(s) declared taking informed written consent for the publication of clinical photographs /material (if any used), from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient, however it cannot be guaranteed.

Authors Contribution: Author(s) declared to fulfill authorship criteria as devised by ICMJE and approved the final version. Authorship declaration form, submitted by the author(s), is available with the editorial office.

Acknowledgements: None

- laparotomy, or laparoscopy? J Indian Assoc Pediatr Surg. 2021; 26:66-7.
5. Kumar M, Maletha M, Bhuddi S, Kumari R. Rapunzel Syndrome in a 3-year-old boy: A menace too early to present. J Indian Assoc Pediatr Surg. 2020; 25:112-4.
6. Mirza MB, Talat N, Saleem M. Gastrointestinal trichobezoar: An experience with 17 cases. J Pediatr Surg. 2020; 55:2504-9.
7. Michael G, Qureshi E, Miah M, Husain N. An unusual form of a trichobezoar causing a peculiar case of appendicitis: A case of Rapunzel syndrome. Cureus. 2020; 12:e7554. Available from: <https://doi.org/10.7759/cureus.7554>.
8. Prasad A, Jain A, Gupta A, Kamra A. Trichobezoar: ravenous for hair. Euroasian J Hepatol Gastroenterol. 2018; 8:97-8.
9. Radhakrishna V, Basavaraju M. Laparoscopic extraction of a Rapunzel: Safe and effective. Gut. 2020; 69:A1-A95.
10. Antunes H, Barroso C, Faria C, Correia-Pinto J. Images in pediatrics: Rapunzel syndrome: the pathway for a prompt diagnosis. Arch Dis Child. 2020; 105:298.
11. Claro M, Costa Santos D, Abreu Silva A, Deus C, Grilo J, Sousa D, et al. When eating makes you sick - Gastric stump obstruction caused by a phytobezoar. A case report and literature review. Int J Surg Case Rep. 2021; 79:263-6.
12. Kang SK, Kim SH, Cho YH. Successful laparoscopic removal of a huge trichobezoar in cases of Rapunzel Syndrome in children. J Acute Care Surg. 2021; 11:39-42.