

## CASE REPORT

### Death of a young girl with Rapunzel Syndrome

Mendis NDNA<sup>1</sup> & Illangarathne Banda YMG<sup>2</sup>

<sup>1</sup> Faculty of Medicine, University of Colombo, Sri Lanka

<sup>2</sup> Teaching Hospital, Kurunegala, Sri Lanka

#### ABSTRACT

**Introduction:** Rapunzel Syndrome is an uncommon condition where a trichobezoar is seen at autopsy. This consists of a ball of hair in the stomach, with strands of hair extending like a tail into the small intestine.

**Case History:** A post-mortem examination was performed on an eleven-year-old girl. History revealed chronic alopecia of over 6-8 months duration with loss of appetite and occasional abdominal pain of 3-4 months duration. She had collapsed suddenly and was pronounced dead on admission to hospital. Autopsy revealed peritonitis secondary to gastric perforation with a trichobezoar in the stomach weighing 600g.

**Discussion:** Trichobezoars usually remain symptomless until they become large. They are often palpable when they are substantial. It is common in young females, usually with an underlying psychiatric disorder. Various imaging modalities help in the detection of bezoars. The main complications are ulceration, perforation, and obstruction. This condition is rare, but late or missed diagnosis could be fatal.

**Conclusions:** Even though Rapunzel syndrome is an extremely uncommon variant of trichobezoar, awareness and early suspicion could prevent fatal complications.

**Keywords:** Alopecia, trichobezoar, Rapunzel's syndrome, gastric perforation, peritonitis

Corresponding Author: **Mendis NDNA**  
asela@fortox.cmb.ac.lk  
ORCID iD: <https://orcid.org/0000-0002-4690-9493>

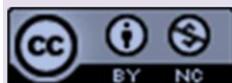
#### Article History

Received: 31.08.2020

Received in revised form: 07.10.2020

Accepted: 16.10.2020

Available online: 20.05.2021



This article is licensed under the terms of the Creative Commons Attribution-Non Commercial 4.0 International License.

#### INTRODUCTION

A bezoar is an indigestible accumulation of foreign material in the gastrointestinal tract. Because of the stomach's large capacity, gastric bezoars do not become symptomatic until they are substantial. The masses are classified according to their contents. Phytobezoars include fruit fibers or plants,

lactobezoars are composed of milk, trichobezoars are concretions of hair, and pharmacobezoars are composed of medications.<sup>1</sup> Trichobezoars are rare conditions that consist of hair bundles in the stomach or small bowel. A Trichobezoar is seen in Rapunzel Syndrome, where swallowed hair forms a mass in the stomach with strands extending like a tail into the small intestine.

Infants and children, particularly if mentally disturbed or abnormal, may have a habit of swallowing foreign material, which may lead to the formation of a bezoar in the gastrointestinal tract. Trichotillomania is a condition where the individual pulls out his/her hair from various body sites.

The site from which hair is most frequently pulled is the scalp, but hair may be removed from eyelashes, eyebrows, the pubic region, or other body part. Trichotillomania is usually associated with a condition called trichophagia in which the individual chews and swallows hair.

The condition could go unnoticed for a long time because, on most occasions, it is not witnessed and, even if witnessed, is not considered as pathological. In these cases, loss of hair is the first symptom or complaint. However, often patients are undiagnosed - until it is too late.

Of those with trichobezoars, only 50% give a history of trichophagia. 55% of all bezoars are Trichobezoars.<sup>2</sup> In sporadic cases, strands of hair from the hairball extend through the pylorus, a condition aptly described as "Rapunzel Syndrome".<sup>3</sup> We present a case of an 11-year-old girl who succumbed to this condition where the diagnosis was made at autopsy.

### CASE HISTORY

History revealed that the deceased lived with her uncle and aunt as they do not have children. Her parents had separated a few years ago. Even though she had preferred to be alone, her guardians or others had not noticed any abnormal behavior. However, they noticed loss of hair of the victim of about 6-8 months duration. They had sought medical treatment, including a dermatological opinion. Following treatment, the relatives had noticed an apparent improvement but said it recurred after some time.

She has also complained of loss of appetite and occasional abdominal pain of 3-4 months' duration and abdominal distention for one month. Four days before the fatal event, she had complained of constipation.

On the fatal day she had complained of severe abdominal pain with fever and vomiting. Soon after, she collapsed and was rushed to hospital where she was pronounced dead. An inquest was ordered.

An inquest was held where a post-mortem examination was ordered. There was no significant history of any illnesses, including psychiatric conditions. Autopsy performed the following day revealed an averagely built female with mild pallor and dehydration. There were no external injuries suggestive of accidental or intentional trauma. The length of head hair was 30cm, being somewhat sparse with areas of alopecia.

A collection of about 1000ml of yellow coloured purulent fluid was seen in the peritoneal cavity. A perforation was noted on the anterior wall of the stomach close to the lesser curvature in its mid-region (Figure 1). There was a sticky ball of human hair weighing 600g in the stomach (Figure 2). No

other abnormalities were seen at autopsy. A small tail of hair was seen extending into the 1st and 2nd part of the duodenum. No other abnormality was detected in the gastrointestinal system or any other system in the body. The cause of death was stated as peritonitis due to perforation of the stomach due to a trichobezoar in the stomach.

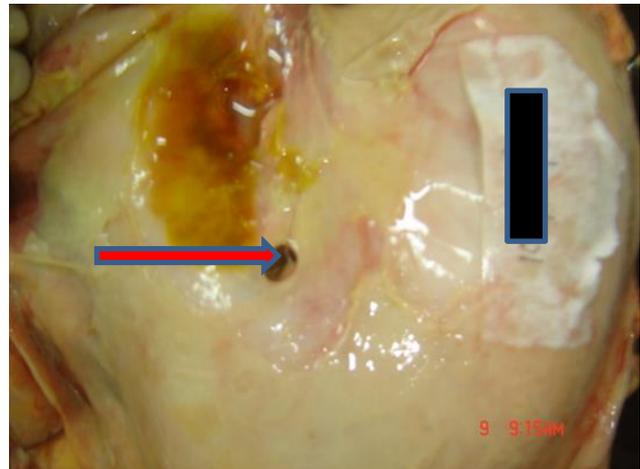


Fig 1: Site of perforation on the outer wall of the stomach (indicated with the arrow)



Fig 2: Trichobezoar in the stomach

### DISCUSSION

Bezoar is a collection of foreign material in the intestinal tract. There are many types of bezoars. Trichobezoar is a collection of hair, while Phytobezoar contains vegetable fiber. The term bezoar originates from either Arabic or Persian languages.<sup>4</sup> They are known to give rise to various symptoms and signs mainly related to the gastrointestinal tract.

An important question that needs some explanation is why the condition was not diagnosed until the patient was dead. The reasons are many. The condition which commonly leads to this complication is an illness classified and described under psychiatric disorders. In DSM-5, trichotillomania is classified as Obsessive-Compulsive and Related Disorders.<sup>5</sup> This condition is usually associated with another condition called trichophagia, which means ingestion of hair. Commonly patients present with the complaint of loss of hair. As in this case, they usually seek treatment from family physicians and dermatologists but are not referred to psychiatrists. It is known to be common in young females. When a young female presents with abdominal symptoms accompanied by alopecia and trichophagia, the possibility of trichobezoar should be considered.<sup>6</sup>

Trichobezoars are usually symptomless until they reach a large size. They may present with varied upper gastrointestinal symptoms. Large bezoars are often palpable. However, the asymptomatic and unsuspecting nature of this pathology is mainly responsible for delayed or missed diagnosis. A highly variable and non-specific pattern of alopecia and the presence of other differential diagnoses usually complicates the picture and makes the diagnosis more difficult.

In most instances, trichophagia would go unnoticed or undiagnosed, leading to chronic hair accumulation in the stomach. Human hair usually resists peristaltic movement. It adheres to the stomach wall and gradually grows into a sticky ball of hair.<sup>7,8,9</sup> This often leads to the development of chronic symptoms and signs, mainly of the gastrointestinal system. Complications include bowel obstruction, nausea and vomiting, the sensation of mass and fullness, loss of appetite, and in severe cases, even perforation and bleeding.<sup>10,11</sup> Valencia et al. stated the most common complication of trichobezoar to be either stomach or intestinal perforation.<sup>12</sup>

In this case, the diagnosis of trichobezoar was never considered despite the presence of characteristic symptoms and signs.

Gastric perforation is an infrequent complication of this condition and is said to occur due to high intra-gastric pressure resulting from the bezoar. Chronic stagnation of substances due to accumulating hair could also result in inflammation and infection of the stomach wall rendering it susceptible to perforation.<sup>13</sup> Pressure necrosis is the possible cause of gastric or intestinal perforation. Mortality

due to this condition is said to be rare but is associated with perforation of the gastrointestinal tract.

Various imaging modalities help in the detection of bezoars. Computerized tomography with contrast medium is useful in the diagnosis. Furthermore, CT scans could also be used to differentiate the bezoar from other pathologies such as intra- or extra-gastric neoplasms.<sup>14</sup>

Rapunzel syndrome was first described in 1968 by Vaughan et al., who used the term to describe large trichobezoar, found in two teenage girls.<sup>15</sup> Rapunzel syndrome is sporadic, with very few documented cases. The literature review did not reveal any reports of Rapunzel syndrome in Sri Lanka. Also, according to a study where 49 cases from 1968 - 2015 were reviewed, only 3 cases have resulted in a fatal outcome.<sup>16</sup> Awareness of this treatable condition is essential for clinicians to avoid a fatal outcome like this case.

This case also highlights the other medico-legal issues involved with it. Child abuse is a possible differential diagnosis that one should consider. However, in this case, there was no evidence of child abuse. Apart from alopecia, the child appeared to be healthy and normal. The absence of fresh or recent injuries also makes the possibility of child abuse unlikely.

From a preventive viewpoint, the question arises as to whether this case was medically mishandled. However, the nature of the condition and asymptomatic period before the condition gets aggravated is likely to misdirect the clinician, especially when such pathology is very rare. Though the condition was not diagnosed since this condition's incidence is rare and unrecorded in the local literature before this death, a charge of medical negligence is unlikely.

## CONCLUSIONS

Even though Rapunzel syndrome is an extremely uncommon variant of trichobezoar, awareness and early suspicion could prevent fatal complications in a young female with chronic hair loss and abdominal pain.

Compliance with Ethical Standards: No conflict of interest. Informed consent was obtained from parents/guardians for useful information for educational purposes with the maintenance of confidentiality of the identity.

## REFERENCES

- Iwamuro M, Okada H, Matsueda K, et al. Review of the diagnosis and management of gastrointestinal bezoars. *World J Gastrointest Endosc.* 2015;7 (4):336-345. doi:10.4253/wjge.v7.i4.336.
- Dhinakar M, Balkhair W. Rapunzel Syndrome: A Case Report. *Oman Med J.* 2010;25(4):e016. doi:10.5001/omj.2010.96
- Phillips MR, Zaheer S, Drugas GT. Gastric trichobezoar: case report and literature review. *Mayo Clin Proc.* 1998;73(7):653-656. doi:10.1016/S0025-6196(11)64889-1.
- Gupta Naik S, Naik S, Chaudhary AK, Jain P, Sharma A. Rapunzel Syndrome Reviewed and Redefined. *Dig Surg* 2007; 24:157-161.
- American Psychiatric Association. (2013). Diagnostic and statistical manual of mental disorders (5th ed.). Arlington, VA: Author.
- Sharma RD, Kotwal S, Chintamani, Bhatnagar D. Trichobezoar obstructing the terminal ileum. *Trop Doct.* 2002;32(2):99-100. doi:10.1177/004947550203200217
- Gorter RR, Kneepkens CM, Mattens EC, Aronson DC, Heij HA. Management of trichobezoar: case report and literature review. *Pediatr Surg Int.* 2010;26(5):457-463. doi:10.1007/s00383-010-2570-0
- Nuss MA, Carlisle D, Hall M, Yerneni SC, Kovach R. (2003). Trichotillomania: a review and case report. *Cutis*, 72: 191– 6. [PubMed]
- Kleiner O, Finaly R, Cohen Z. Giant gastric trichobezoar presenting as abdominal mass in a child. *Acta Paediatr.* 2002;91(11):1273-1274.
- Gorter, R. R., Kneepkens, C. M., Mattens, E. C., Aronson, D. C., & Heij, H. A. (2010). Management of trichobezoar: case report and literature review. *Pediatric surgery international*, 26(5), 457–463. <https://doi.org/10.1007/s00383-010-2570-0>
- Avisar, E., Goldberg, M. & Lernau, O. Bezoar-induced ulceration and perforation of the upper gastrointestinal tract in mentally retarded patients. *Pediatr Surg Int* 9, 279–280 (1994). <https://doi.org/10.1007/BF00832257>
- Santos Valenciano J, Nonose R, Bragattini Cruz R, Tiemi Sato D, Monteiro Fernandes F, Fabricio Nascimento E, et al. Tricholithobezoar causing gastric perforation. *Case Rep Gastroenterol.* 2012;6:26–32. doi: 10.1159/000336203000336203. [PMC Free Article] [PMC free article] [PubMed] [CrossRef] [Google Scholar]
- Wadlington WB, Rose M, Holcomb GW Jr. Complications of trichobezoars: a 30-year experience. *South Med J.* 1992;85(10):1020-1022. doi:10.1097/00007611-199210000-00024
- Kaushik NK, Sharma YP, Negi A, Jaswal A. Images- Gastric trichobezoar. *Indian J Radiol Imaging* 1999; 9:137-139.
- E.D. Vaughan Jr., J.L. Sawyers, H.W. Scott Jr., The Rapunzel syndrome. An unusual complication of intestinal bezoar, *Surgery* 63 (1968) 339–343.
- Kim, Soon Chul MD, PhD; Kim, Seong Hun MD, PhD; Kim, Sun Jun MD, PhD A Case Report, *Medicine*: May 2016 - Volume 95 - Issue 22 - p e3745 doi: 10.1097/MD.0000000000003745