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# Rapunzel Syndrome: A Case Report and Literature Review

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## Abstract

Rapunzel syndrome is an extremely rare clinical condition in children. Usually affects girls of adolescent age group with history of hair ingestion (trichophagia) and trichotillomania (hair-pulling). Patients present with vague abdominal pain and bowel obstruction caused by a hairball in the stomach, with its tail extending into duodenum and beyond. We report a case of 13-year-old girl with poor general condition, who presented with recurrent abdominal pain, vomiting and a palpable mass in the abdomen. She gave history of trichophagia and trichotillomania for more than two years. On exploration, a large trichobezoar with a tail was noted in the stomach, duodenum and proximal jejunum. The bezoar was removed. The girl had uneventful recovery. She received psychiatric treatment and improved.

## Introduction

Rapunzel syndrome is characterized by presence of hairballs or hair-like fibers in the stomach and intestine. These results from chewing and swallowing hair or any other indigestible materials (trichophagia) often associated with hair-pulling disorder (trichotillomania) in young girls [1-3]. The syndrome is named after the long-haired girl 'Rapunzel' in the fairy tale by the Brothers Grimm [4]. We present a case of Rapunzel syndrome in an adolescent girl, who needed surgical removal of the hairball and psychiatric treatment.

## Case Presentation

A 13-year old girl with poor general condition presented with recurrent abdominal pain with distension, vomiting and constipation. She had a palpable mass in the epigastric region. An ultrasonography showed an intragastric mass which extended to the duodenum and proximal jejunum. The patient gave history of hair-pulling and chewing for more than two years. A laparotomy and gastrotomy revealed a big hair-ball occupying almost the whole stomach (Figure 1). The tail of the hair-ball extended into the duodenum and proximal jejunum and was carefully pulled out in its entirety (Figure 2). The stomach was repaired and the abdomen closed. The patient had uneventful recovery and subsequently received psychiatric treatment. On follow up after 6 months, the girl had gained weight and recovered from psychiatric disorder.

## Discussion

Vaughan first reported a patient with Rapunzel syndrome in 1968 [5]. This was named so after a fairy tale written in 1812 by Brothers Grimm's about the German princess 'Rapunzel' who let her long golden hair down from her prison tower to permit her young prince to climb up to her window and rescue her [4]. This condition occurs most commonly in females under the age of 30, youngest being a six-month-old infant. Trichotillomania is an impulse control disorder characterized by the repeated urge to pull out scalp and body hair. Trichophagia is the compulsion to eat or chew on hair. These patients have underlying psychological disorders, family discord, childhood neglect or abuse or mental retardation [6]. In the present case, the mother of the patient revealed that the girl was an introvert and average in her study.

Trichobezoars forms when ingested hair strands and fibers are retained in the gastric mucosa, peristalsis causes this to become enmeshed until a ball is formed. The ball gradually becomes large enough to cause gastric atony. Trichobezoars are typically black regardless of the color of the hair ingested because of enzymatic oxidation of gastric acid on the hair fibers and often emits an unpleasant odor due to decomposition and fermentation of fats [7]. If bezoars are left without treatment, the mortality rate can reach up to 30%.

In Rapunzel syndrome, the gastric trichobezoar has a long extension of hair into the duodenum or even to the large bowel [8]. It has been reported to cause jaundice and pancreatitis as a result of obstruction of the ampulla of Vater by hair. As well as pancreatitis and malabsorption complications

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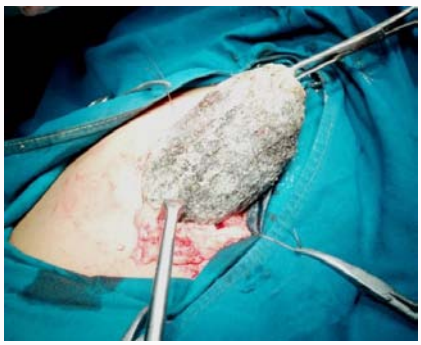

**Figure 1:** The hairball being removed from the stomach.

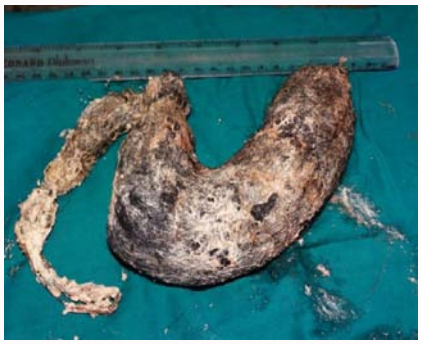

**Figure 2:** The trichobezoar occupying the whole stomach and duodenum and proximal jejunum.

like protein-losing enteropathy and iron deficiency and megaloblastic anemia [9]. Poor nutritional status in the present case was because of lack of appetite and irregular bowel habit.

The most common symptom in these cases is vague epigastric discomfort. Other symptoms include nausea, vomiting, anorexia, early satiety, and weight loss. If a bezoar reaches large size and is present for a prolonged period, it may cause pressure necrosis and ulcers, which leads to bleeding or obstruction [10,11]. Once in the small bowel, bezoars most commonly result in obstruction.

Investigation in such cases includes imaging, such as, plain film, Ultrasound (USG), upper gastrointestinal series, and CT scan. USG may demonstrate a band of increased echogenicity caused by the intermixed hair, air and food in the trichobezoar, which is diagnostic. Detection rate of trichobezoars by USG has been reported to be around 88% [12]. Upper gastrointestinal series can demonstrate a filling defect in the stomach; however, this procedure is not recommended as it may cause obstruction or perforation. CT scan demonstrates heterogeneous masses containing trapped air bubbles or homogenous mottled appearance in the region of stomach or intestine [12]. Upper gastrointestinal endoscopy may be helpful to diagnose trichobezoars, but it cannot confirm small bowel extension [13]. In our case, the diagnosis was made with the help of an USG, and so no other imaging was necessary.

Treatment of Rapunzel syndrome comprises of removal of the trichobezoar by various means combined with behavioral therapy and psychiatric treatment of the patient. Endoscopic removal of small trichobezoars have been reported, but overall success rate is less than 5% [14,15]. Laparoscopic removal of small to moderate sized bezoars can be done, but large bezoar (>20 cm) requires removal by laparotomy. Other methods of treatment, such as, enzymatic dissolution (by papain syrup, pancreatic lipase, and cellulose), Nd-YAG laser and extracorporeal shock wave fragmentation have been reported [16]. Long-term surgical follow-up with upper GI endoscopy or USG abdomen [17] and psychiatric follow-up with psychotherapy and cognitive behavioral therapy are critical for prevention of recurrence [18].

## References

1. Sah DE, Koo J, Price VH. Trichotillomania. Dermatol Ther. 2008;21(1):13-21.
2. Ventura DE, Herbella FA, Schettini ST, Delmonte C. Rapunzel syndrome with a fatal outcome in a neglected child. J Pediatr Surg. 2005;40(10):1665-7.
3. Mateju E, Duchanová S, Kovac P, Moravanský N, Spitz DJ. Fatal case of Rapunzel syndrome in neglected child. Forensic Sci Int. 2009:190(1-3):e5-7.
4. Grimm Brothers: Rapunzel. Translated by Godwin-Jones R. Richmond, Virginia Commonwealth University Department of Foreign Languages. 1994-99.
5. Vaughan ED, Sawyers JL, Scott Jr HW. The Rapunzel syndrome. An unusual complication of intestinal bezoar. Surgery. 1968;63(2):339-43.
6. Grant JE, Odlauq BL. Clinical characteristics of trichotillomania with trichophagia. Compr Psychiatry. 2008;49(6):579-84.
7. Barzilai M, Peled N, Soudack M, Siplovich L. [Trichobezoars]. Harefuah. 1998;135(3-4):97-101,167.
8. Kaspar A, Deeg KH, Schmidt K, Meister R. [Rapunzel syndrome, an rare form of intestinal trichobezoars]. Klin Padiatr. 1999;211(5):420-2.
9. Sharma V, Sahi RP, Misra NC. Gastro- Intestinal Bezoars. J Indian Med Assoc. 1991;89(12):338-9.
10. Jiledar Singh G, Mitra SK. Gastric perforation secondary to recurrent trichobezoar. Indian J Pediatr. 1996;63(5):689-91.
11. Klipfel AA, Kessler E, Schein M. Rapunzel syndrome causing gastric emphysema and small bowel obstruction. Surgery. 2003;133(1):120-1.
12. Ripolles T, Garcia-Aguayo J, Martinez MJ, Gil P. Gastrointestinal Bezoars: Sonographic and CT characteristics. AJR Am J Roentgenol. 2001;177:65-9.
13. Coulter R, Antony MT, Bhuta P, Memon MA. Large gastric trichobezoar in a normal healthy woman: Case Report and review of pertinent literature. South Med J. 2005;98(10):1042-4.
14. Konuma H, Fu K, Morimoto T, Shimizu T, Izumi Y, Shiyanagi S, et al. Endoscopic retrieval of a gastric trichobezoar. World J Gastrointest Endosc. 2011;3(1):20-2.
15. Gorter RR, Kneepkens CM, Mattens EC. Management of trichobezoar: Case report and literature review. Pediatr Surg Int. 2010;26(5):457-63.
16. Nirasawa Y, Mori T, Ito Y, Tanaka H, Seki N, Atomi Y. Laparoscopic removal of a large gastric trichobezoar. J Pediatr Surg. 1998;33(4):663-5.
17. Fallon SC, Slater BJ, Larimer EL, Brandt ML, Lopez ME. The surgical management of Rapunzel syndrome: a case series and literature review. J Pediatr Surg. 2013;48(4):830-4.
18. Miltenberger RG, Rapp JT, Long ES. Habit reversal treatment manual for trichotilomania. 2001;171-95.