

## Case Report

# Rapunzel's giant hairball: a report of Rapunzel syndrome in a young man

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**Received:** 12 June 2024

**Accepted:** 10 July 2024

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### ABSTRACT

Rapunzel syndrome, a rare form of trichobezoar, is a condition in which an accumulation of hair forms in the stomach and extends to the small intestine. Here we describe a case of a 17-year-old male presenting with gastric outlet obstruction secondary to Rapunzel syndrome, requiring laparotomy and gastrotomy.

**Keywords:** Laparotomy, Rapunzel syndrome, Trichobezoar, Trichophagia

## INTRODUCTION

Bezoar refers to an accumulation of undigested material in the stomach. A trichobezoar is an unusual condition which involves the accumulation of hair in the stomach and is generally seen in young women with a background of trichotillomania (the urge to pull one's hair) and trichophagia (the urge to eat one's hair). Small trichobezoars can generally be removed endoscopically. Rapunzel syndrome is the syndrome described when the trichobezoar is so large that it extends down into the small bowel.<sup>1</sup> This is a very rare condition which is almost exclusively seen in young women.<sup>2</sup>

## CASE REPORT

A previously well 17-year-old male presented to his GP with complaints of intermittent abdominal pain for the past 18 months, vomiting and dry retching, which had worsened over the last 6 months. This was associated with 15 kg of unintentional weight loss. The GP arranged blood tests which showed profound iron deficiency anaemia, thrombocytosis, and hypoalbuminaemia, and he was sent for a CT Abdomen and Pelvis. The CT revealed

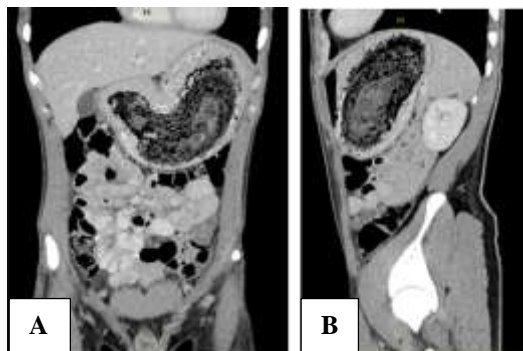
a large, partially calcified mass in the abdomen which filled the stomach and extended to the duodenum (D3).



**Figure 1: Axial CT image demonstrating large, calcified trichobezoar in the stomach.**

The GP referred the patient to the emergency department for review. On review, he appeared well, had an afro hair style and a slim build but did not appear overtly malnourished. On history, he revealed that he had been

eating his hair since he was 9 years old. His parents were unaware of this, and he never previously sought medical attention. On examination, he had a non-distended, non-tender abdomen, with a palpable mass in the epigastrium. He was admitted under gastroenterology. The following day he underwent gastroscopy which revealed a gigantic gastric trichobezoar.



**Figure 2 (A and B): Coronal and sagittal CT images.**

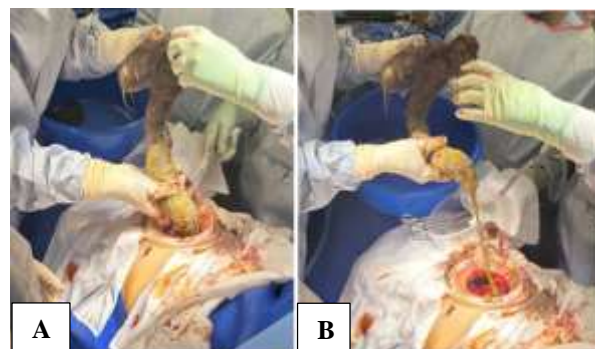
In addition, a few small nonbleeding, superficial ulcers were seen (Forrest Class III). Removal was attempted but was unsuccessful due to its size. He was also referred to psychiatry, who established that his hair ingestion was a maladaptive response to stress but did not give him any formal diagnoses. An outpatient psychology review was suggested for counselling and pharmacological management was not required. He was referred for a second endoscopy with a specialist interventional team, with the use of an over-tube to assist in removal. During this procedure they were able to pass beyond the mass and found that it extended to the proximal jejunum. Due to its massive size endoscopic removal was unsuccessful, so he was referred to the surgical team.



**Figure 3 (A and B): Endoscopic images demonstrating trichobezoar in stomach and extension into jejunum.**

Upon reviewing the CT scans, a laparoscopic approach was deemed infeasible due to its size and calcified core. A mini midline laparotomy and gastrotomy was performed. A foul-smelling enormous trichobezoar was immediately evident. Sponges were used to pack other surrounding organs away and to minimize any spillage or contamination. With traction, it was removed, en bloc

from the stomach. The giant trichobezoar was 26 cmx18 cmx8 cm and kept the shape of the stomach and duodenum. A nasogastric tube (NGT) was inserted and guided past incision under vision. The stomach was closed with double layer closure.



**Figure 4 (A and B): Intra-operative images of removal of trichobezoar through laparotomy and gastrotomy, demonstrating the long “tail” extending to jejunum.**

The patient was kept fasted with a NGT in situ on free drainage and Q4hourly aspirates. A leak test with oral contrast was performed on Day 6 post-operatively, which did not demonstrate contrast extravasation. His NGT was removed with diet progression whilst being carefully monitored for re-feeding syndrome. He was discharged home on post-operative Day 10 and recovered well following discharge.



**Figure 5: Retrieved specimen- massive trichobezoar 15 cm ruler placed above for reference.**

## DISCUSSION

The clinical presentation of trichobezoars is generally vague, so there is often delay to diagnosis and treatment. The condition commonly presents in adolescents, and symptoms are non-specific, often including chronic abdominal pain, vomiting, decreased oral intake and nutritional deficiencies. Trichobezoar most commonly presents in young women and adolescents with a history of trichotillomania and trichophagia, who may also have concurrent psychological or behavioural conditions.<sup>1</sup> On examination, an epigastric mass may be noted, or

alopecia from excessive hair pulling. Although associated with trichophagia, only 1% of patients with trichophagia go on to develop a trichobezoar.<sup>5,6</sup> Rapunzel syndrome is a term originally coined by Vaughn et al and generally refers to a trichobezoar that extends to the small intestine, and occasionally as far as the right colon. Only around 7-10% of patients with trichobezoar described in the literature are male and Rapunzel syndrome is even more uncommon in males, with only 5% of described cases occurring in male patients.<sup>4,8</sup> Workup of patients with suspected trichobezoar can be difficult as often patients are children or adolescents, and excessive radiation exposure is avoided. However, if there is significant clinical suspicion for trichobezoar, plain Xray, Ultrasound or, ideally cross-sectional imaging such as CT or MRI scanning should be performed. The gold standard for diagnosis of trichobezoar is upper GI endoscopy and may also have a therapeutic role.<sup>1,9</sup> Potential complications of Rapunzel syndrome, as seen in our case, include anaemia (due to malabsorption or bleeding), and gastric outlet obstruction. Other described complications include bowel perforation, intussusception, pancreaticobiliary obstruction and pancreatitis.<sup>4,10,11</sup> The literature describes both endoscopic and surgical management of trichobezoar. In retrospect, attempts at endoscopic removal were likely futile based on the CT images, and first line operative management would have shortened his hospital stay. Both laparoscopic and open removal have been described. A laparoscopic approach was described by Jan et al, in which the trichobezoar was removed piecemeal via temporary gastrotomy and was able to be removed completely in this fashion.<sup>12</sup> This is a novel approach, which we considered, however given the calcification seen on CT and the sheer size of the trichobezoar, the decision was made to proceed with laparotomy, as piecemeal removal, if possible, at all, would greatly increase operative time. Additionally, laparoscopically it would be difficult to assess if the whole trichobezoar was removed, particularly as there was known extension into the jejunum. Psychiatric input is essential in the management of patients with trichobezoar. Patients benefit from cognitive behavioural therapy to decrease their habitual trichophagia and prevent trichobezoar recurrence.<sup>2</sup>

## CONCLUSION

Rapunzel syndrome is a rare form of trichobezoar. Generally, patients with Rapunzel syndrome require surgical management consisting of laparotomy and gastrotomy. Although rare, this condition should be considered in young patients presenting with vague abdominal pain, vomiting or nutritional deficiency, and cross-sectional imaging and specialist opinion should be promptly sought to avoid development of complications.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

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**Cite this article as:** Gabsi K, Cheng E, Yu Y. Rapunzel's Giant Hairball: A report of Rapunzel Syndrome in a young man. *Int Surg J* 2024;11:1355-7.