Case Report

Rapunzel syndrome in surgical emergency managing dual trichobezoars in a patient with cerebral palsy

Abhishek Saini, S Balamurugan

From Senior Resident, Department of General Surgery, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India

ABSTRACT

Rapunzel syndrome, a rare form of trichobezoar, may lead to significant bowel obstruction. The patient often presents with features of bowel obstruction, such as vomiting, abdominal pain, and distention. We report the case of a 13-year-old female with cerebral palsy who arrived at the emergency department with symptoms indicative of gastric outlet obstruction. Diagnostic imaging and laparotomy revealed two large trichobezoars, causing concurrent obstruction at the gastric outlet and in the small bowel. Given the association with psychiatric conditions such as trichophagia, post-surgical psychiatric evaluation was arranged to address the underlying behavior and minimize recurrence risk. This case highlights the critical role of an integrated diagnostic and therapeutic approach for effective management in these patients.

Key words: Intestinal obstruction, Laparotomy, Rapunzel syndrome, Trichobezoar

Trichobezoars are rare gastric masses typically resulting from the ingestion of hair, often associated with trichophagia and trichotillomania [1]. In severe cases, these hair masses extend beyond the pylorus, resulting in a rare manifestation known as Rapunzel syndrome. This condition primarily affects young females, particularly adolescents aged 10–19 years, and can lead to acute gastric outlet or intestinal obstruction [2]. This term, introduced by Vaughan *et al*. [3], draws its name from the Brothers Grimm's fairy tale character Rapunzel, known for her exceptionally long hair. Trichobezoar resulting in Rapunzel syndrome is an exceptionally uncommon condition, with fewer than 50 cases documented in medical literature to date [4]. Management usually involves surgical intervention, often followed by psychiatric counseling to prevent recurrence.

This case report discusses a unique presentation of Rapunzel syndrome in a young patient with cerebral palsy, focusing on the diagnostic, surgical, and psychological aspects essential for comprehensive treatment. Despite the rarity of Rapunzel syndrome, its potential for severe complications, such as gastric outlet obstruction and intestinal perforation, underscores the importance of case documentation to enrich clinical understanding. Reporting this case contributes to the growing body of literature, facilitating timely diagnosis and multidisciplinary management of this rare condition.

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CASE REPORT

A 13-year-old female with a history of cerebral palsy presented to the emergency department with acute abdominal pain, abdominal distension, nausea, and 2 days of non-bilious vomiting. She had also experienced constipation for 4 days. Her parents reported a history of trichophagia.

On general examination, the patient appeared well-built but showed signs of dehydration. Tachycardia was noted, although blood pressure remained within normal limits, and there was no fever at the time of presentation. Scalp examination revealed areas of patchy hair loss. Abdominal examination demonstrated marked upper abdominal distension on inspection. Palpation revealed a large, firm, immobile mass in the epigastric region, extending to the umbilical and right lumbar areas. Auscultation of the abdomen revealed an absence of bowel sounds.

Initial laboratory tests showed anemia (hemoglobin 8.9 g/dL). An erect abdominal X-ray displayed multiple air-fluid levels, suggesting obstruction. A computed tomography (CT) scan of the abdomen identified a large, heterogeneous mass measuring 18 × 12 cm within the stomach and a second mass of 7 × 5 cm located in the proximal jejunum, with the collapse of the distal bowel and substantial free fluid present in the abdominal cavity (Fig. 1).

The patient underwent an emergency laparotomy under general anesthesia due to acute intestinal obstruction. Upon exploration, intraoperative findings revealed a large trichobezoar measuring 20 × 8 × 5 cm occupying the stomach and a second trichobezoar

Correspondence to: Abhishek Saini, Department of General Surgery, JIPMER, Puducherry, India. E-mail: abhishek5500@gmail.com

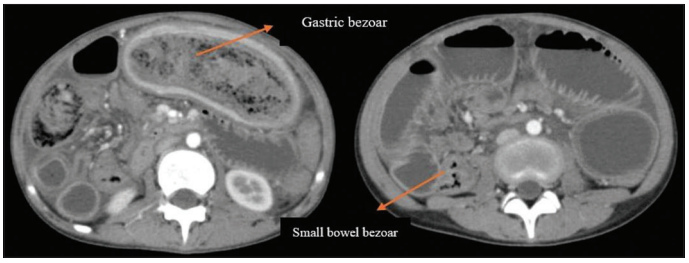

Figure 1: Contrast-enhanced computed tomography imaging demonstrating gastric and small bowel bezoar

measuring 7 × 4 × 3 cm located in the proximal jejunum (Figs. 2 and 3). The bowel distal to the second trichobezoar was found to be collapsed. The gastric trichobezoar was removed through a gastrotomy incision made along the anterior wall of the stomach (Fig. 2). The jejunal trichobezoar was extracted through an enterotomy performed at the affected segment of the jejunum (Fig. 3). Both surgical sites were meticulously repaired with primary closures to ensure structural integrity and prevent leakage. In addition, the abdominal cavity was irrigated thoroughly to remove free fluid and prevent potential post-operative complications such as infection or adhesions. The patient's post-operative course was uneventful, and she was discharged with scheduled psychiatric follow-up to address the underlying trichophagia and prevent recurrence.

DISCUSSION

Rapunzel syndrome, first described by Vaughan *et al*. in 1968, is an unusual form of trichobezoar where the hair mass extends from the stomach into the intestines, creating a cast-like obstruction [1,3]. This rare entity primarily presents with symptoms such as abdominal pain, palpable mass, nausea, and vomiting. Patients, often young females, may have a history of psychiatric conditions such as trichotillomania and trichophagia, leading to the formation of hair-based bezoars in the stomach [2]. In cases like the one described, the bezoar not only obstructs the gastric outlet but also extends into the small intestine, necessitating surgical intervention. Hair strands can get trapped in the stomach's mucosal lining, where the peristalsis causes them to intertwine and form a ball. Eventually, this ball becomes too large, leading to gastric atony [5]. According to the patient's parents, she had a childhood habit of playing with hair, often picking up strands, chewing on them, and likely swallowing them, which contributed to the condition.

In pediatric cases, especially those complicated by underlying conditions such as cerebral palsy, the presentation may be atypical, requiring high diagnostic suspicion and prompt management. Imaging modalities, including ultrasound and CT scans, are critical for identifying bezoars, with the characteristic "mottled" appearance confirming the diagnosis [2]. Endoscopic retrieval of trichobezoars may be attempted; however, large or complicated cases like this one often demand laparotomy for effective removal.

Compared to other cases in the literature, our patient had cerebral palsy, which added a layer of complexity to diagnosis and management. While most cases reported healthy adolescent

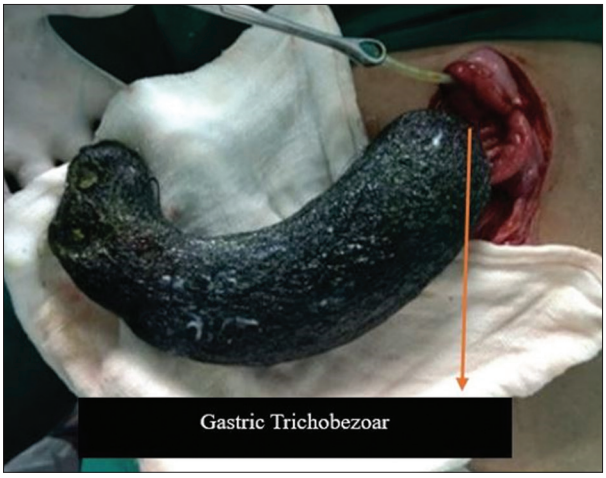

Figure 2: Gastric trichobezoar extracted through a gastrostomy incision

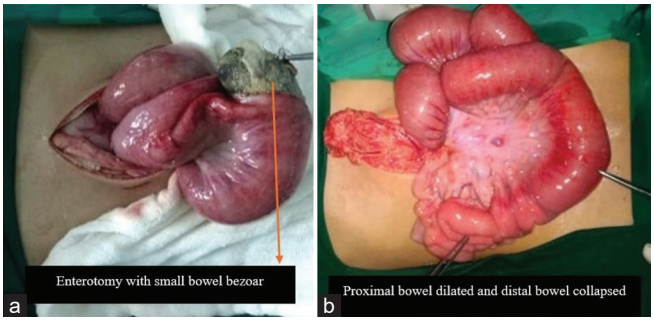

Figure 3: (a) Enterotomy over proximal jejunum demonstrating a separate trichobezoar; (b) Demonstrating proximal dilated bowel and distal bowel collapsed

females, the presence of an underlying neurological disorder in this patient introduced additional challenges in clinical evaluation, as symptoms such as abdominal distension and vomiting could be attributed to other gastrointestinal issues common in cerebral palsy. Moreover, the occurrence of two bezoars instead of a single mass deviated from the typical presentations, making the surgical procedure more intricate.

Another distinction was the co-presentation of proximal jejunal obstruction alongside gastric outlet obstruction, as highlighted by CT imaging. Most cases describe an extension of trichobezoars into the small intestine without causing a separate obstruction. The dual obstruction in this patient necessitated meticulous planning during laparotomy to ensure complete removal and prevent further complications.

Rapunzel syndrome, primarily managed through surgical intervention, has limited success with medical and endoscopic approaches, especially in larger or complex trichobezoars. Enzyme therapy and endoscopic removal using forceps, baskets, or lithotripsy have been trialed but are often ineffective for sizeable bezoars. For smaller trichobezoars, endoscopy can sometimes facilitate removal, although recurrence remains a risk [6-9]. Laparotomy is the preferred approach for comprehensive extraction, minimizing the risk of obstruction [7].

Psychiatric assessment is vital post-surgery, as recurrence is common without addressing the underlying behavioral disorder.

Studies suggest that nearly half of the patients with trichobezoars experience recurrence, underscoring the importance of multidisciplinary management. Treatment strategies, therefore, include not only surgical excision of the bezoar but also long-term psychiatric follow-up to prevent recurrence, addressing underlying conditions such as anxiety, depression, or trichotillomania. This integrative approach can improve outcomes and reduce the risk of future obstructions, as psychiatric therapy has proven beneficial in minimizing trichophagia and trichotillomania tendencies [8,10].

CONCLUSION

In summary, Rapunzel syndrome presents unique diagnostic and therapeutic challenges. This case highlights the importance of surgical intervention, complemented by psychiatric support to manage both the acute and long-term aspects of the syndrome. Proper recognition and comprehensive treatment are essential, as delays in diagnosis can lead to complications such as gastric perforation, malnutrition, or recurrent intestinal obstructions.

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