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RAPUNZEL SYNDROME - A TALE HARD TO SWALLOW AND HARDER TO DIAGNOSE: A CASE REPORT

General Surgery

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ABSTRACT

Bezoars are concretions of indigestible material that accumulate within the gastrointestinal tract, most commonly in the stomach. Among the various types, trichobezoars-composed primarily of ingested hair-account for approximately 20% of all bezoar cases [1]. In rare and severe presentations, the trichobezoar extends beyond the pylorus into the small intestine, a condition referred to as Rapunzel syndrome [2]. A 17-year-old female presented with progressive abdominal distension, abdominal pain, nausea, constipation, and bilateral pedal oedema for one month. Laboratory investigations revealed mildly deranged liver enzymes and elevated ascitic fluid adenosine deaminase (ADA), raising suspicion of infective pathology. Ultrasonography and contrast-enhanced computed tomography (CT) demonstrated gross ascites with features of subacute intestinal obstruction but failed to identify a definitive bezoar. Exploratory laparotomy revealed a giant gastric trichobezoar with a long intestinal extension, consistent with Rapunzel syndrome. Complete removal was achieved through gastrotomy. The patient had an uneventful postoperative recovery and was discharged in stable condition. Rapunzel syndrome should be considered in adolescent females presenting with unexplained abdominal distension and intestinal obstruction, even when imaging and laboratory findings are inconclusive. Timely surgical intervention remains both diagnostic and therapeutic.

KEYWORDS

Rapunzel syndrome; Trichobezoar; Intestinal obstruction; Ascites; Adolescent; Gastrotomy

INTRODUCTION

Bezoars are concretions of indigestible material that accumulate within the gastrointestinal tract, most commonly in the stomach. Among the various types, trichobezoars—composed primarily of ingested hair-account for approximately 20% of all bezoar cases [1]. In rare and severe presentations, the trichobezoar extends beyond the pylorus into the small intestine, a condition referred to as **Rapunzel syndrome** [2].

Trichobezoars predominantly affect young females and are often associated with psychiatric conditions such as trichotillomania and trichophagia, although these behaviors may be denied or remain unrecognized [3,4]. Clinical manifestations are usually vague and nonspecific, leading to diagnostic delays and potential complications. We report a rare case of Rapunzel syndrome presenting with ascites and subacute intestinal obstruction, posing a significant diagnostic challenge.

CASE STUDY

Clinical History and Examination

A 17-year-old female presented with abdominal pain, progressive abdominal distension, nausea, and constipation for approximately one month. The pain was insidious in onset, gradually progressive, mild to moderate in intensity, non-radiating, and not associated with jaundice, gastrointestinal bleeding, loose stools, or clay-coloured stools. She also had bilateral pedal oedema.

There was no history of hypertension, diabetes mellitus, tuberculosis, asthma, previous abdominal surgery, blood transfusion, or drug allergy.

Investigations

Ultrasonography of the abdomen revealed thickened, oedematous bowel loops with altered luminal echotexture. Contrast-enhanced CT (CECT) of the abdomen and pelvis demonstrated gross loculated ascites with smooth peritoneal thickening and internal septations, centrally displaced small bowel loops, dilated jejunal and ileal loops with air-fluid levels, and a small-bowel feces sign. Mild circumferential mural thickening of distal ileal loops was noted. No definitive radiological evidence of a trichobezoar was identified preoperatively.

Management And Operative Findings

In view of persistent symptoms and radiological features of intestinal obstruction, the patient was scheduled for exploratory laparotomy under general anaesthesia. Intraoperatively, a giant gastric trichobezoar composed of hair and undigested food material was identified and removed via a gastrotomy and jejunotomy. The mass measured approximately **22 × 7 × 8 cm** and had a long tail extending into the duodenum and jejunum, confirming the diagnosis of Rapunzel syndrome (Fig.1). The stomach and intestinal walls were intact, with no evidence of ulceration or perforation.

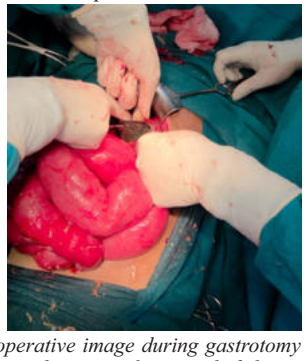

Figure 1: *Intraoperative image during gastrotomy showing a large brown intraluminal mass of entangled hair, representing a trichobezoar*

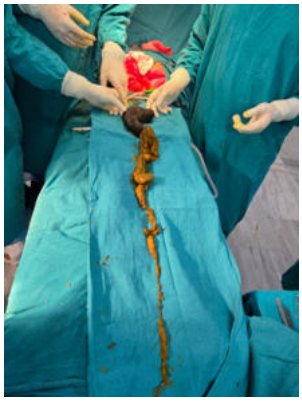

Figure2: *Extracted specimen showing large trichobezoar*

approximately 120x7x1 cm

The extracted specimen was an elongated, fusiform, tapering mass resembling a thread-like structure, measuring approximately **120 cm in length**, with progressive reduction in girth from **7 cm proximally to 1 cm distally** (Fig. 2)

Postoperative Course

The postoperative period was uneventful. The patient was managed with intravenous fluids, antibiotics, and analgesics. Oral feeding was gradually resumed, and she was discharged in stable condition on postoperative day seven days with advice for psychiatric evaluation and follow-up.

DISCUSSION

Over time, swallowed hair accumulates within the stomach and entangles with food particles, forming a dense intragastric mass. Due to the smooth and slippery nature of hair, gastric peristalsis fails to expel the mass, allowing progressive enlargement [5,6]. When the mass extends into the small intestine, the condition is termed Rapunzel syndrome, representing a rare but more severe manifestation.

Clinical presentations are typically vague and nonspecific, including abdominal pain, nausea, vomiting, early satiety, and occasionally a palpable abdominal mass. Diagnostic modalities such as abdominal ultrasonography, computed tomography, and upper gastrointestinal endoscopy aid in confirming the diagnosis, although imaging may occasionally be inconclusive, as seen in the present case [7].

This case is notable for **Misleading biochemical markers** (elevated ADA, LDH), **Radiological features mimicking infective or inflammatory pathology** and **Absence of definitive CT diagnosis**

Elevated ascitic ADA levels have been reported to cause diagnostic confusion, particularly in regions endemic for tuberculosis. However, Rapunzel syndrome should remain a differential in young females presenting with bowel obstruction and ascites, especially when imaging is inconclusive. Surgical exploration remains both diagnostic and therapeutic.

Most cases are diagnosed in adolescent females, reflecting the higher prevalence of trichotillomania and trichophagia in this demographic [8,9]. The most effective non-invasive treatment option for selected cases is endoscopic fragmentation with aspiration; however, large bezoars with intestinal extension usually require surgical removal [9].

Management requires not only definitive removal of the bezoar but also psychiatric evaluation and long-term follow-up to prevent recurrence [10].

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Management requires not only definitive removal of the bezoar but also psychiatric evaluation and long-term follow-up to prevent recurrence [10].

CONCLUSIONS

Rapunzel syndrome is a rare but important differential diagnosis in adolescent females presenting with unexplained abdominal distension, ascites, and intestinal obstruction. This case highlights the limitations of imaging and laboratory investigations and underscores the importance of clinical judgment and timely surgical intervention.

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