Successful laparoscopic extraction of trichobezoar due to Rapunzel syndrome: first reported case in Kuwait

Dr. Ahmad E. Al-Mulla MD , Dr. Ali Altabeekh MD, Dr. Ahmad Al-Jafar MD and Dr. Shaimaa Dashti MD
Department of Surgery Farwaniya Hospital
Ministry of Health Kuwait

Abstract:

Rapunzel syndrome is a rare condition typically found in young female patients with mental or psychiatric disorders. It manifests as a large report, we discuss a successful laparoscopic extraction of a trichobezoar in a 12-year-old girl with acute abdominal pain due to small bowel obstruction. We describe our technique compared with others' techniques and include a literature review on this topic.

Introduction:

Bezoar is defined as an indigestible mass in the gastrointestinal tract, especially in the stomach. The second most common bezoar is the trichobezoar, which occurs in young female psychiatric patients. Trichobezoars are usually confined to the stomach. In extremely rare cases, they extend beyond the pylorus and into the small intestine, which is known as Rapunzel syndrome (RS). This rare case report describes the unusual presentation of abdominal pain in a 12-year-old girl that was managed laparoscopically.

Case presentation:

A 12-year-old girl presented to our emergency department with a five-day history of severe colicky abdominal pain associated with vomiting, nausea and anorexia. No previous surgical and medical history. Clinical examination, with typical vital signs, epigastric and right upper quadrant pain (RUQ). Laboratory tests: elevated amylase (141U), lipase (138 U/L) and urine amylase (1,776 U/L). The rest of the blood test was unremarkable. Ct-scan abdomen and pelvis revealed minimal pelvic fluid with a foreign body at the stomach extending to the pylorus (Figure 1). An endoscopic attempt to remove the foreign body was unsuccessful due to the size of the bezoar and the high possibility of aspiration and respiratory tract obstruction.

Surgical procedure:

- Preformed laparoscopically with three working ports.
- Gastrostomy was performed and complete extraction of bezoar from the greater curvature of the stomach via 5- cm gastric incision (Figure 2).
- A 15 cm long bezoar was extracted via endo-bag without spillage nor remaining particles intra-abdominal (Figure 3).

Post-operative:

- Patient was discharged after receiving antibiotics and resuming normal bowel and diet.
- She was referred for psychiatric consultation.

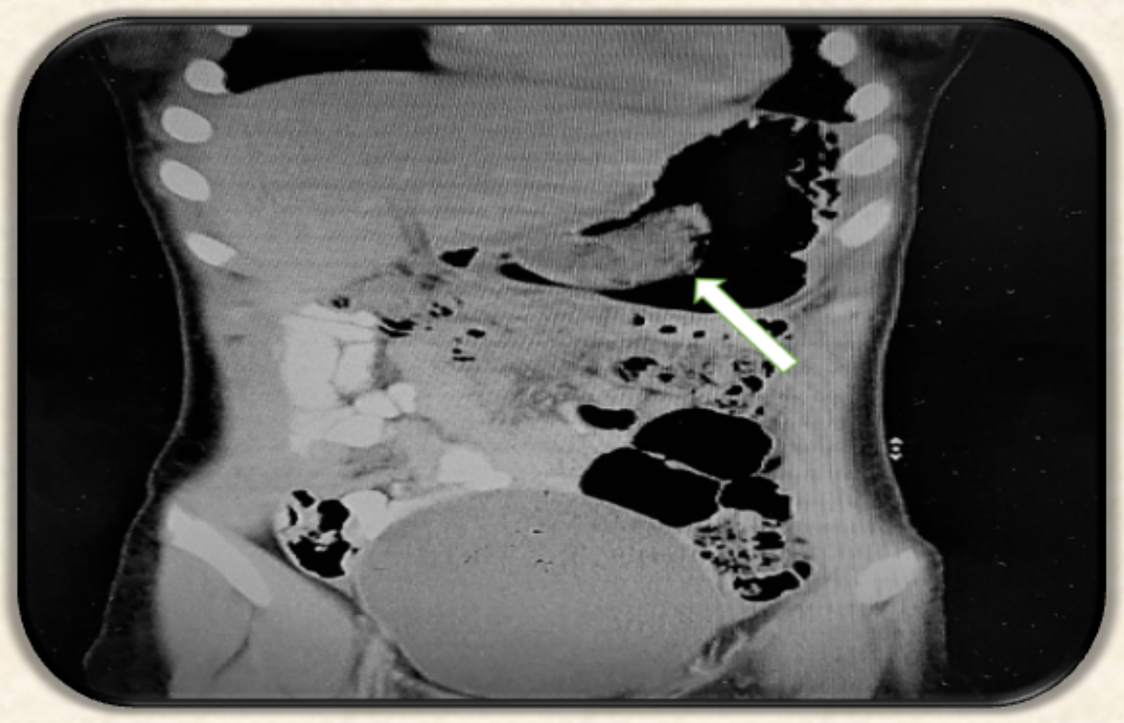

Figure 1 : CT-scan abdomen showing the bezoar at the pylorus

Conclusion:

RS is an extensive form of trichobezoar, it can present a variety of complications. Imaging is an essential tool to diagnose the disorder because it presents with vague symptoms. *Trichobezoar* must be considered in the differential diagnosis of a young female with pancreatitis or vague small bowel abdominal symptoms. Management is primarily by open surgery, but it can be performed laparoscopically (dependent on the patient's clinical characteristics).

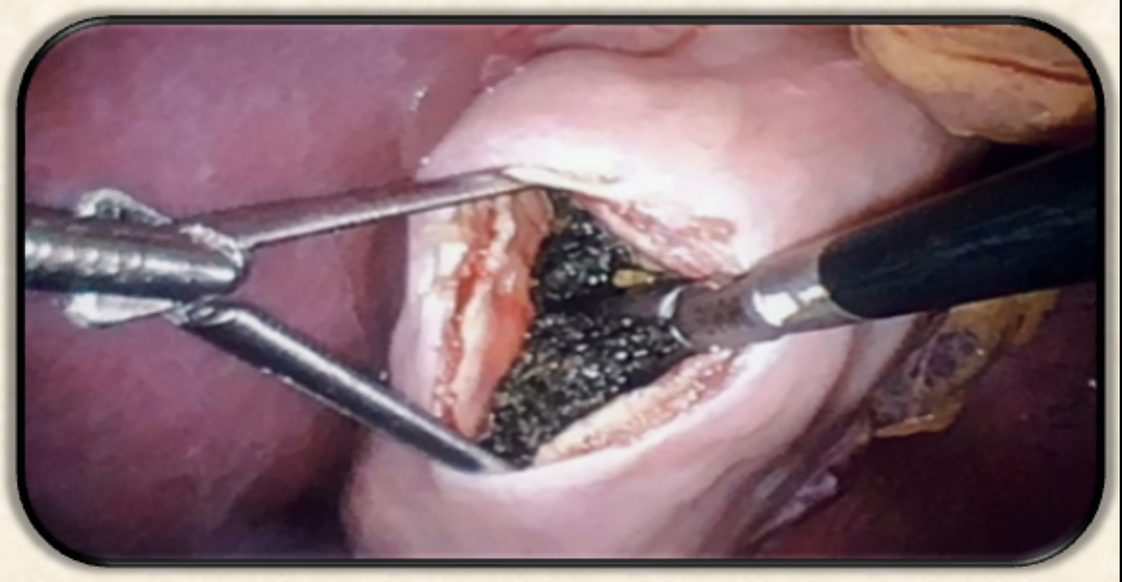

Figure 2: A 5-cm laparoscopic gastrostomy and extraction of a bezoar.

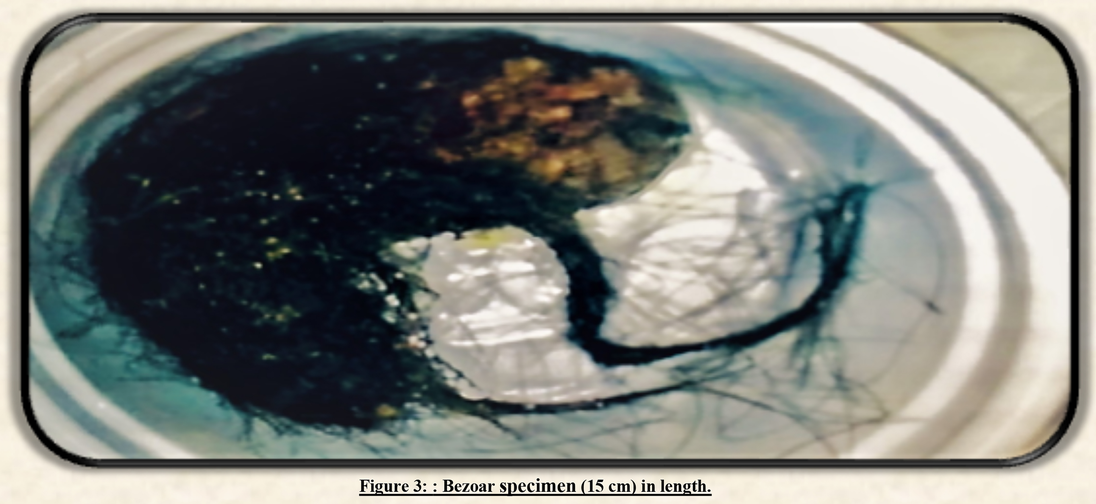

Figure 3: : Bezoar specimen (15 cm) in length.

Discussion

Bezoars can be classified into four main types: phytobezoars, trichobezoars, pharmacobezoars and lactobezoars. Trichotillomania and trichophagia are common in young women aged 13–20 years who have intellectual disabilities or psychiatric disorders. Trichotillomania is an impulse control disorder characterized by the repetitive urge to pull out one's hair, leading to hair loss. Many of these patients also ingest their hair and animal hair as well as rug, synthetic and natural fibres. The slippery nature of the hair traps it within the gastric folds, leading to bezoar formation . RS is a unique manifestation of an advanced gastric trichobezoar, first described in literature by Vaughn et al. in 1968. RS has three standard features: 1) trichobezoar in the stomach, 2) long hair strands that extend from the stomach to the small intestine and sometimes the large bowel and 3) gastrointestinal symptoms. It is typically asymptomatic until the bezoar reaches a substantial size, which leads to abdominal discomfort (37%), weight loss and anorexia (38%), later developing into nausea or postprandial vomiting (38%) and abdominal cramps. Physical examination often shows a palpable mobile abdominal mass (70%) and loss of scalp and eyebrow hair as well as eyelashes. Long-standing gastric bezoars lead to adverse complications, such as obstruction, perforation, bleeding, intussusception, peritonitis, ulceration, obstructive jaundice, weight loss, pancreatitis, and mortality. In our case, the patient presented with acute abdominal pain with a CT scan suggestive of small bowel obstruction. Elevation of lipase and amylase was due to migration of the bezoar to the ampulla of Vater, causing spasms and congestion of the sphincter of Oddi, resulting in bile regurgitation and trypsin activation once the pressure in the bowel was higher than in the pancreatic duct. Diagnosis of trichobezoar can be made by ultrasound, CT scan, barium meal and endoscopy.

RS treatment aims to extract the trichobezoar endoscopically, laparoscopically or via open surgery. Upper endoscopy is less invasive and more cost-effective; however, it is not recommended in these cases due to its lower success rate and potential for airway obstruction and respiratory arrest. Most cases are primarily managed with open surgery, which was described in a retrospective study of seven cases of gastric bezoars; all patients required exploratory laparotomy. The first laparoscopic extraction of a bezoar was conducted in 1998; only a few cases that were successfully managed laparoscopically are mentioned in the literature.

There are *three techniques* are described in the literature. First, Don et al. described passing a 10-mm port into the gastric wall to create an intra-gastric port, followed by removing the specimen with a gastroscopy. Second, Kanetala et al. described a similar approach using two gastric ports to cut the bezoar into small fragments and removing them via gastroscopy; both approaches are time-consuming. The third technique, described by Shami et al., was used in our case. In our case we performed a gastrotomy, extracting the specimen with a grasper and placing it in a Endo-bag then removing it through the 10-mm camera port . We did not experience any spillage or postoperative infection. A laparoscopic approach is also an option; however, it can be time-consuming and technically challenging cases. There is as of yet no gold standard procedure due to the rarity of the disease; however, it should be performed with attention to patient safety and outcomes.