

Stomach trichobezoar (rapunzel syndrome) with iatrogenic intussusception

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SUMMARY

We present a rare case of stomach trichobezoar complicated with iatrogenic intussusception noted intra-operatively after failed attempt of endoscopic removal in a 13-year-old girl. At presentation, she had gastric outlet obstruction with anaemia. Endoscopy established the diagnosis of trichobezoar. Surgical removal was warranted after failed endoscopic removal. Her postoperative course was uneventful. Detailed history and careful examination disclosed trichotillomania and associated trichophagia. Psychiatric referral was sought with the intention to prevent future recurrence.

KEY WORDS:

Intussusception; Rapunzel syndrome; Trichobezoar; Trichophagia; Trichotillomania

INTRODUCTION

A bezoar is an accumulation of indigestible exogenous matter in the stomach or intestine. Many different types of bezoars have been described, including phytobezoar, pharmacobezoar, trichobezoar and lactobezoar.

Gastric trichobezoar consists of accumulation of hair, usually patient's own, in the stomach. It can become very large, often with a "tail" extending into the duodenum (Rapunzel syndrome), which may cause gastric or small bowel obstruction, jaundice or even acute pancreatitis or peritonitis.¹

Diagnosis of gastric trichobezoar may be aided by radiological investigations together with a high index of suspicion.² Surgical removal was required in the presence of gastrointestinal obstruction, with careful exploration for additional bezoars^{1,3}. Though this condition was rare, identification of trichotillomania and trichophagia was important to prevent future recurrence¹.

CASE REPORT

A 13-year-old Indian girl presented with epigastric pain for seven days associated with vomiting for one day. History revealed symptomatic anaemia with loss of appetite for three months. At presentation, clinical examination found pallor with mild epigastric tenderness but without peritonitis.

Laboratory investigations revealed hypochromic, microcytic anaemia with normal amylase and liver function test. Abdominal radiographs showed no evidence of bowel obstruction. With these features, she was initially diagnosed with anaemia, probably secondary to peptic ulcer disease by the attending physician. Due to her unrelieved, persistent symptoms of abdominal pain and vomiting, she was later referred to the surgical team for assessment. Further examination of the abdomen revealed a vague epigastric mass with no jaundice. She has localised epigastric tenderness with no signs of peritonitis. Detailed history taking from her mother disclosed past history of pica (habitual eating of particles from the floor including rubber bands) and trichotillomania (act of hair pulling). However, her mother did not notice her swallowing her own hair (trichophagia). In addition, she has an identical twin sibling, also had pica (eating charcoal), but remained well.

In view of her persistent obstructive symptom with the possibility of a stomach bezoar, an upper endoscopy (OGDS, oesophagogastroduodenoscopy) was performed to assess her upper gastrointestinal tract. At endoscopy, a large trichobezoar was identified, with its body extended beyond the stomach into the duodenum (Figure 1A). Attempted endoscopic removal failed. She underwent exploratory laparotomy with a large trichobezoar located along the greater curvature of the stomach (Figure 1B), removed via an anterior gastrotomy (Figure 1C). The trichobezoar extended beyond the pylorus into the duodenum, with a third bezoar concretion located at 120 cm from the ileocecal valve (Figure 1D). The gastroduodenal bezoars were noted to have a string embedded in it upon removal. The string was snapped during removal of the gastroduodenal bezoars. Further exploration identified an iatrogenic ileo-ileal intussusception (Figure 1E). The intussusception was reduced, with an enterotomy performed to retrieve the bezoar from the ileum (Figure 1F). The complete removal of all trichobezoars (Figure 2) was confirmed by full exploratory laparotomy. Her postoperative course was uneventful. Oral feeding was commenced three days after surgery and intravenous antibiotics were administered for three days. The total duration of her admission was six days. Retrospective examination revealed alopecia areata of her frontal forehead, consistent with trichotillomania. She was referred for psychiatric assessment before discharge. She remains well 9 years after surgical intervention with regular psychiatric follow-ups.

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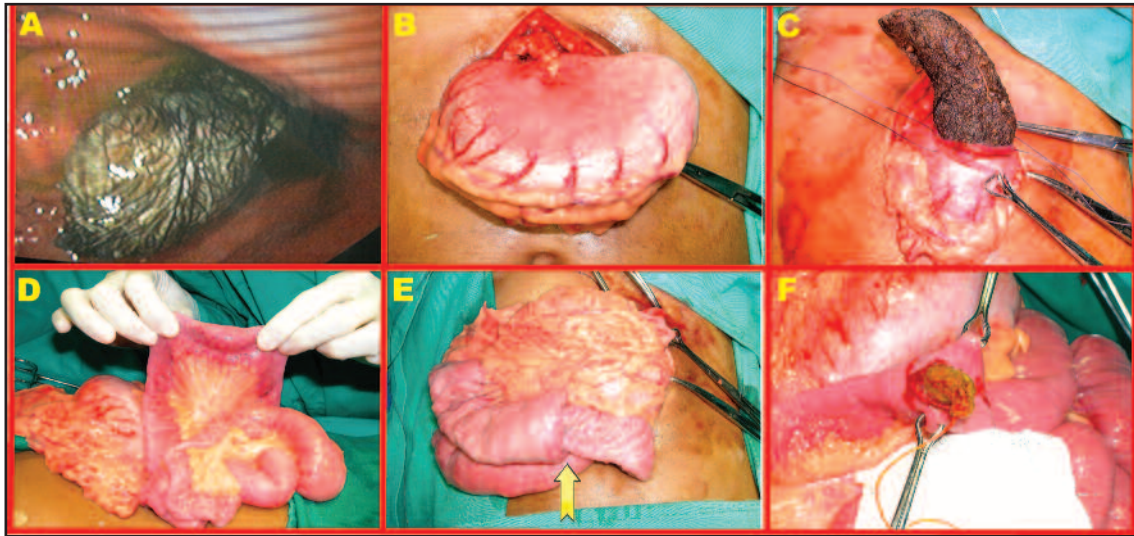


Fig. 1: (A) Endoscopic finding of a trichobezoar extended beyond the pylorus, with attempted endoscopic removal failed. (B) At laparotomy, stomach was grossly distended by a "sausage-shaped" trichobezoar along the greater curvature of the stomach. (C) Gastric trichobezoar delivered via anterior gastrotomy. (D) A third bezoar concretion was found in the ileum. (E) Exploratory laparotomy found an ileo-ileal intussusception (arrow). (F) Enterotomy performed for removal of the 3rd intestinal bezoar concretion after reduction of ileo-ileal intussusception.



Fig. 2: Complete trichobezoars removed, consisted of: (1) a huge stomach trichobezoar (1st bezoar), (2) a large duodenal trichobezoar (2nd bezoar, bile-stained) with a snapped string embedded and communicated with the 3rd bezoar, (3) a small intestinal trichobezoar (3rd bezoar).

DISCUSSION

Gastric trichobezoar is an unusual cause of gastric outlet obstruction, bleeding and perforation in the Paediatric

Emergency Department. It most commonly occurs in teenage girls with concomitant, frequently undiagnosed psychiatric disorder known as trichotillomania. Only one third of these patients have trichophagia, and just 1% of these individuals ingest enough hair to require surgical intervention. The clinical presentation of trichobezoar is usually delayed, after ingestion of large amount of hairs over many years. Trichobezoar occurs most commonly in young females under the age of 30 years, with the youngest reported case affecting a 6-month-old infant. Trichobezoar is principally located in the stomach, but can secondarily extend into the duodenum, causing the so-called Rapunzel syndrome,¹ named after the long-haired heroine of Grimms' fairy tales. Rarely, a trichobezoar confines to the small bowel.³ The presence of simultaneous gastric and small intestinal bezoars has been reported.³

Alopecia produced by intentional hair pulling may be the only clue to diagnosis, but the child usually denies eating or swallowing hair.¹ Observation and history from family members are essential to identify these behavioural disorders.

Radiological investigations together with high index of suspicion are usually able to identify trichobezoar accurately due to its characteristic anatomical abnormality.² Plain abdominal radiograph is difficult to interpret unless the concretion of hairs has other associated foreign body with air trapping as well, appearing as a mottled, soft tissue opacity in the shape of the distended stomach. Contrast radiograph is helpful and diagnostic to delineate the anatomical abnormality created by the trichobezoar and has been reported.¹ Computed scintigraphy is uniquely characteristic and diagnostic in trichobezoar with the effects of air trapping within the concretion of the trichobezoar.²

Upper endoscopy has added benefits compared with other modality of investigation. It is very helpful in differentiating a trichobezoar from a phytobezoar, as therapy for each is

different. In contrast to phytobezoars, trichobezoars are resistant to enzymatic or chemical degradation and therefore removal by endoscopic fragmentation³ is required, or for larger trichobezoars, laparoscopy or a formal laparotomy is necessary,^{1,3} as in our case.

Early diagnosis of trichotillomania with associated trichophagia and prevention of bezoar formation should be of primary importance, as recurrence is common without treatment of the underlying psychiatric disorders.¹

Our case emphasizes the role of endoscopic assessment in the diagnosis of trichobezoars and the importance of complete exploration of the abdomen at the time of removal of an obstructing gastric bezoar,³ in view of, a possible second or third bezoars, which can be easily missed and may cause further obstruction, if they are not identified. The occurrence of ileo-ileal intussusception in our case is perhaps iatrogenic, secondary to the overzealous, failed attempt of endoscopic removal, prevented by the long and embedded, communicating string, holding the “baby” bezoar to the “mother” gastroduodenal bezoar or it is possibly related to the open retrieval of the entire gastroduodenal bezoars without knowing that it was connected to the small intestinal bezoar via a string. However, the small intestinal “baby” bezoar may well be the pathological lead point for the intussusception to occur. More importantly, psychiatric

referral for the underlying trichotillomania and trichophagia prevented the future, disastrous occurrence of trichobezoars in this patient, perhaps in her twin sister too.

In conclusion, when a female child presents with epigastric pain, the findings of an epigastric mass with anaemia should prompt the clinicians to sought for trichophagia in the history and to look out for trichotillomania in physical examination, and the possibility of trichobezoar has to be considered.

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