

Rapunzel Syndrome and Atypical Pancreatitis Acute Abdomen: A Case Report

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Abstract

Trichobezoar is a condition characterized by the accumulation of undigested hair and or other foreign bodies in the stomach. The reported occurrence is uncommon in the Middle East and usually affects female adolescents and young children. Rapunzel syndrome, which almost exclusively affects young women, is a rare form of trichobezoar in the stomach that spreads to the intestine and is associated with psychiatric disorders such as trichotillomania and trichophagia. This report describes a rare case of giant trichobezoar complicated with acute pancreatitis in a young patient which was ultimately treated with removal of trichobezoar by surgical gastrotomy.

Keywords: Trichobezoar; Trichotillomania; Trichophagia; Laparotomy; Acute Pancreatitis

Introduction

A trichobezoar is an accumulation of undigested hair in the stomach. The term “trichobezoar” is a combination of the words “tricho”, meaning “hair”, and “bezoar”, meaning “undigested foreign body that accumulates in the gastrointestinal tract, which can lead to significant health problems”. The condition is rare occurring in less than 1% in the general population [1,2]. Females are more commonly affected (90%) than males, with 80% of female cases occurring primarily in children and female adolescents [3].

Human hairs resist digestion and tend to accumulate between the mucosal crypts of the stomach, eventually transforming to a mass of “hairball”. A trichobezoar can migrate from the stomach to the duodenum [4-7]. Trichobezoar is usually associated with other psychiatric disorders such as depression, anorexia, obsessive-compulsive disorders [8-10] and has elements suggestive of trichophagia and trichotillomania. Rapunzel Syndrome was a term coined in 1968 for trichobezoars extending from the stomach to the small intestine.

The signs and symptoms of trichobezoar are caused either by gastric outlet obstruction or its complications and include anorexia, bloating, early satiety, weight loss, vomiting immediately after meals, acute epigastric pain and patchy hair loss on the scalp. Possible complications of trichobezoar include intestinal obstruction [11] and gastric perforation with peritonitis, pancreatitis, erosion of the mucosa of the small intestine and ulceration.

We present the case of a 16-year-old female patient who presented to our Urgent Care Centre with abdominal pain and blood investigations suggestive of acute pancreatitis. Imaging was suggestive of a trichobezoar occupying the full gastric cavity which on further questioning became apparent as a result due to trichotillomania and trichophagia.

Case Presentation

A 16-year-old female student presented to our Urgent Care Centre with generalized colicky abdominal pain, nausea and vomiting for a day. She also complained that the episodes of nausea and vomiting had increased for the last one week and also that she was feeling bloated with a mass like feel in the upper abdomen, which was gradually increasing for the last one year. Further gentle history suggested that she had a history of trichotillomania (eating nails, hair, plastic beads, plastic caps and bracelets), which she had recently stopped. She denied any previous medical or surgical history.

General examination revealed a conscious patient with normal vitals and absence of dehydration, pallor and/or jaundice. Abdominal examination demonstrated mild tenderness in the epigastric aspect and a hard fibrinous mass with distinct borders extending from the left upper abdomen, epigastrium to the umbilical region.

Laboratory tests were within normal ranges except for an elevated serum amylase, 291.07 U/L and serum lipase, 604.79 U/L.

An abdominal radiograph showed a distended stomach with soft tissue with a mottled gas pattern and an air rim surrounding the gastric contents (Figure 1).



Figure 1: Plain abdominal xray.

An abdominal ultrasound revealed a large lesion in the epigastrium, suggesting a distended stomach with 'dirty shadowing', suggestive of the presence of mixed contents with numerous air pockets.

A CT scan (Computed Tomography Scan) showed an overly distended stomach filled with heterogeneous contents with a swirled pattern. A large inhomogeneous mass with a concentric pattern of air foci was observed almost completely occupying the gastric lumen and extending into the first part of duodenum (Figure 2).



Figure 2: CT abdomen coronal.

A slight circumferential thickening of the stomach wall was present. Subtle heterogeneous parenchymal hypo-enhancement was observed in the tail of the pancreas. Mild free fluid was noted around the tail of the pancreas and in the left para-colic gutter, suggestive of pancreatitis (Figure 3).

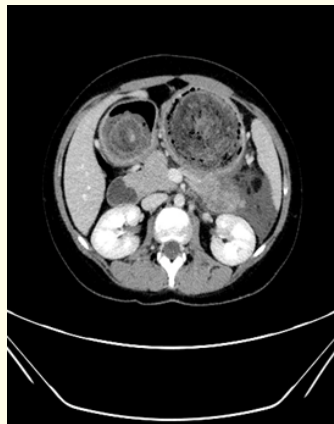


Figure 3: CT abdomen transverse.

Upper GI endoscopy confirmed the presence of trichobezoar occupying the entire gastric cavity.

The patient was therefore taken to the operating theatre for exploratory laparotomy due to the enormous size effectively ruling out laparoscopic removal. An anterior gastrotomy (7 cm long) and en bloc removal of the bezoar was performed (Figure 4 and 5) and the stomach closed in 2 layers after placing a nasogastric tube to aid feeding and healing. The patient had peripheral parenteral nutrition for the first week followed by soft diet and discharged after psychiatric evaluation and appropriate surgical outpatient follow up with patient and family education.



Figure 4

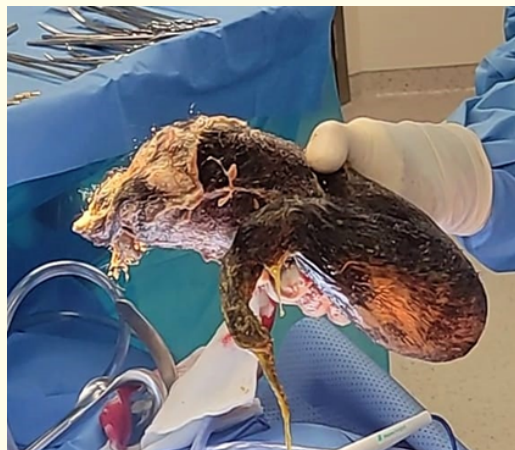


Figure 5

Discussion

The term “bezoar” has Arabic and/or Persian roots. According to their composition, gastric bezoars are classified into Phytobezoars (Composed of vegetable fibres), Trichobezoars (Composed of hair or hair-like fibres), Diospyrobezoars (Composed of persimmons), and Pharmacobezoars (from pills). The most common type of gastric bezoar is trichobezoar, which is the accumulation of undigested hair in the gastric cavity that over a period of time transforms itself to a massive “hairball” [12-15]. It was first discovered in 1779.

Approximately 90% of those affected by Trichobezoar are young females between the ages of 13 and 20 years old. The patient’s past medical history is usually suggestive of trichotillomania and trichophagia which coexist with psychiatric disorders. The chronic mental illness trichotillomania involves an irresistible urge to pull hair and trichophagia is compulsive eating of hair/hair like fibres [8]. Depression, anxiety, dysmorphia and other psychiatric illness aid trichophagia and bezoar.

Patients suffering from a gastric trichobezoar typically experience nonspecific gastric abdominal symptoms, including abdominal pain, nausea, vomiting, diarrhoea, constipation, indigestion, weight loss, digestive bleeding, and patchy hair loss on the scalp, as well as in most cases, patients suffer from malabsorption-related complications, including protein-losing enteropathy, iron deficiency anaemia, and megaloblastic anaemia [16-18]. If left untreated for a long time, it may lead to complications such as obstructive jaundice, small bowel obstructions, mucosal erosion and ulceration, perforation, peritonitis, and acute pancreatitis. Our patient displayed vomiting, abdominal pain, nausea, vomiting and a palpable abdominal mass as the most prominent clinical signs and symptoms.

Radiographically trichobezoars are visualized as hollow viscus masses on abdominal ultrasound but may be clinically suspected by x-rays. Furthermore, an abdominal CT scan may detect trichobezoars and reveal the presence of a mass. But upper gastrointestinal endoscopy, on the other hand, remains the gold standard for diagnosis and can also be therapeutic, depending on the size of the trichobezoar present [19,20].

The management of a trichobezoar is determined by its size, consistency, and localization, as well as its complications. Smaller sizes can be removed endoscopically using a basket or direct suction. In cases involving a larger size, surgery is the preferred method of treatment as it allows for examination of the entire gastrointestinal tract, has a higher success rate, and has a lower incidence of complications [21]. Dissolution therapy with Coca-Cola has been suggested in systematic review [22] with a 90% success rate as first-line treatment along with other endoscopic measures for phytobezoars.

We chose laparotomy in our case due to the size of the trichobezoar, which occupied the entire stomach extending to duodenum, also allowing us to explore the gastric cavity for any ulcerations and/or abnormalities. It is advisable to remove the bezoar en-bloc (including its tail) and to make sure there are no remnants in the duodenum, otherwise, the patient is at risk for postoperative small bowel obstruction. Delayed gastric emptying is common in those patients due to loss of gastric tone from the long-standing distention. A low threshold for parenteral nutrition is advisable. During the patient's admission, psychiatric consultation was done for evaluation of trichotillomania and to avoid recurrences. Adequate psychiatric counselling and adherence to medication is suggested for successful recovery. We consider that a multidisciplinary approach with involvement of surgeons, paediatricians, psychiatrists, and gastroenterologists is vital for this case.

Conclusion

Trichobezoar is an underdiagnosed and rare disease that should be addressed in adolescents with trichotillomania and trichophagia, especially in females. Clinicians need to be aware of the risk factors that contribute to the development of trichobezoar, as well as the delicate clinical signs, in order to detect these diseases early and prevent gastrointestinal problems. Endoscopy can help with a definitive diagnosis and, in certain cases, fragmentation and removal of the trichobezoar. Regardless, surgery remains the gold standard for the treatment of this condition. Psychiatric therapy is indicated as part of the treatment regimen for trichobezoar patients to prevent a recurrence.

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