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CASE REPORT

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Rapunzel Syndrome: Rare 'Tale' of a Broken 'Tail'

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Summary

Background: Rapunzel syndrome is a rare and unique form of trichobezoar, in which a cast of hairs is formed in the stomach with its 'tail' extending up to varying lengths into the small bowel. Almost all cases described in literature are of 'classic' and more common form of Rapunzel. Sometimes however, the tail of bezoar is broken into smaller fragments. There is not much literature available on this rarer subset of Rapunzel syndrome.

Case Report: In this report we present the ultrasound (USG) and CT findings of a case of Rapunzel syndrome in which the tail had broken into five separate fragments. The patient presented clinically with intestinal obstruction due to the impaction of the distal- most fragment in the ileum.

Conclusions: Our case highlights the fact that although USG features may be suggestive, a careful evaluation of CT images is essential for a confidant preoperative diagnosis of Rapunzel syndrome. In cases of this syndrome with a broken tail, CT is essential for precise count and localization of the separated fragments to ensure their complete removal at the time of surgery. We also propose to name the fragments of the broken tail as 'bezoarlets'. This word aptly describes the tail fragments as it suggests their origin from the larger gastric bezoar and the suffix 'lets' conveys they are smaller in size.

MeSH Keywords: Bezoars • Intestinal Obstruction • Tomography, Spiral Computed • Trichotillomania • Ultrasonography

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Background

A bezoar is a firmly packed clump of non-digested material that gets accumulated in the gastrointestinal tract (mostly stomach). They are classified according to the material constituting the core, like trichobezoar (hair), phytobezoars (fibres of plant origin), lactobezoars (milk or curds), lithobezoars (rock-like material) etc. [1].

Trichobezoar is the most common type of bezoar. It is composed of hair and is mostly found in the stomach. It is commonly associated with underlying psychiatric disorders like trichotillomania and trichophagia. It mostly occurs in adolescent females, possibly due to their long hair.

Rapunzel syndrome is an unusual and a rare kind of trichobezoar in which hair extend from the stomach into the intestines up to varying lengths. Our case was even more unusual as the tail of the trichobezoar had broken into multiple separate fragments (five) leading to intestinal obstruction.

Case Report

A 19-year-old female presented in the emergency department of our hospital with complaints of abdominal pain, vomiting and constipation for the last 10 days. She also gave a history of decreased appetite and weight loss for the past few months. On abdominal examination there was a 6×6-cm hard to firm, mobile lump palpable in the epigastric region. Her vitals were stable at the time of presentation and routine laboratory tests were normal apart from mild leucocytosis.

The abdominal USG revealed presence of a curvilinear echogenic intragastric mass with posterior acoustic shadow (PAS). Similar, albeit smaller echogenic masses with prominent PAS were also seen in segments of dilated small gut

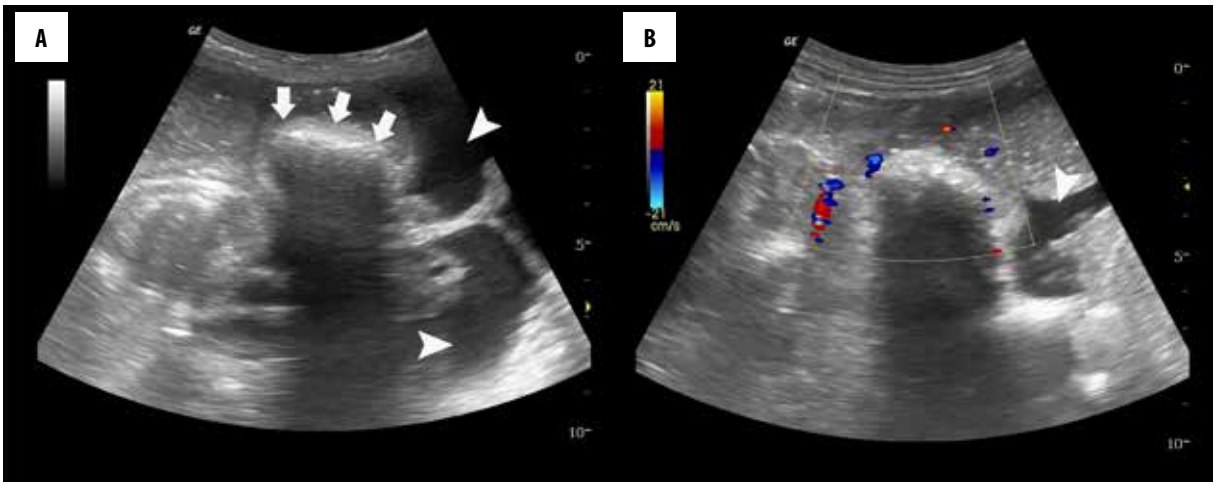


Figure 1. Transverse ultrasound image (**A**) showing a small bowel bezoarlet as curvilinear hyperechoic arc with posterior acoustic shadowing in a dilated jejunal loop (arrows) and the Doppler image (**B**) showing the lack of internal vascularity. Free fluid can also be seen (arrowheads).

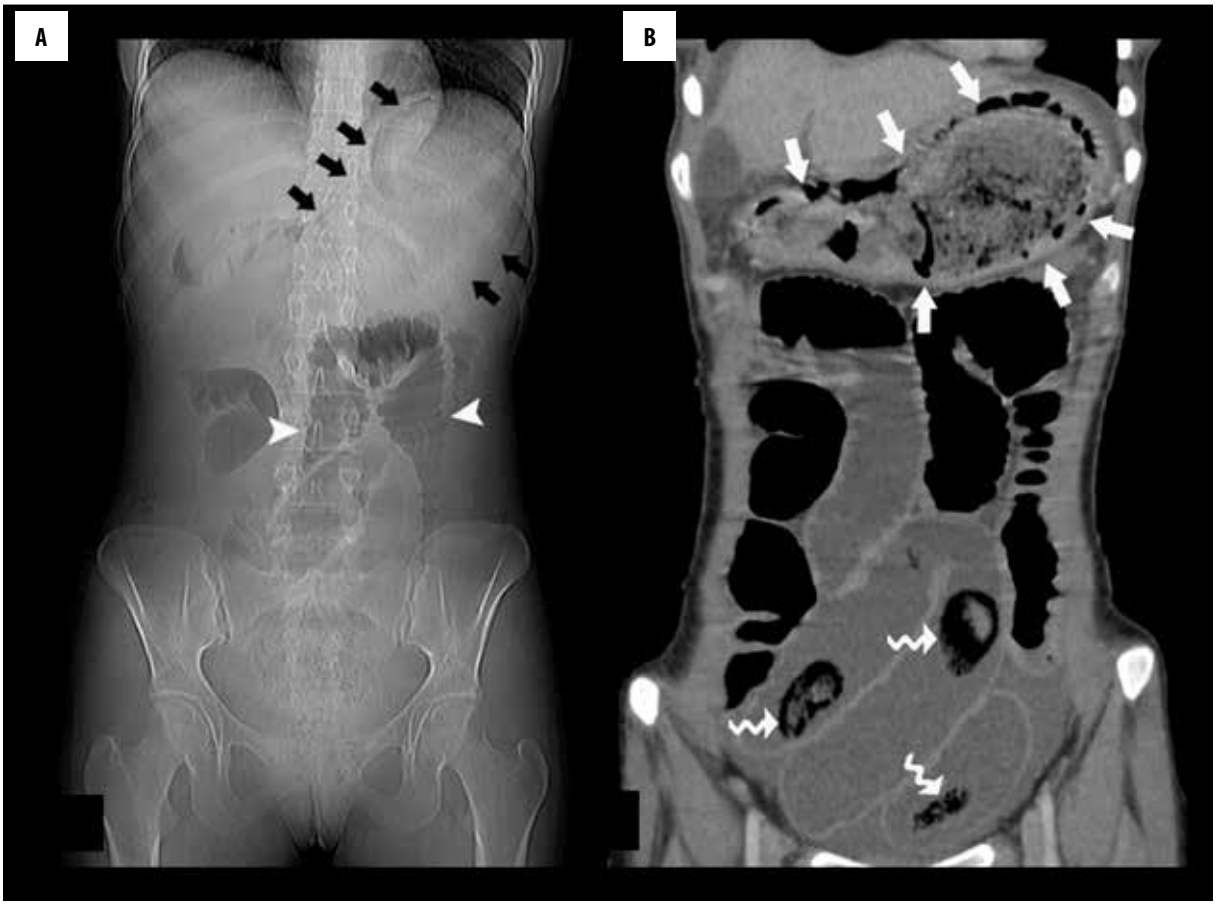


Figure 2. CT scout view of the abdomen (**A**) showing the gastric bezoar as a localized area of mottled lucencies with a peripheral crescent of air (arrows) and dilated air- filled small bowel loops in the mid abdomen (arrowheads). Coronal CT MPR image (**B**) showing the gastric bezoar (arrows) and multiple small bowel bezoarlets (zig-zag arrows)

loops (Figure 1A). Doppler USG did not reveal any abnormal internal vascularity (Figure 1B). Mild free fluid was present in the peritoneal cavity. When asked specifically, the patient admitted to the habit of eating her own hair (trichophagia). The ultrasonographic findings, coupled with the history of trichophagia led us to a suspicion of intragastric and small bowel trichobezoars with intestinal obstruction. CT of the abdomen was done subsequently to confirm the diagnosis. The CT scout view of the abdomen showed mottled radiolucencies in the stomach with a crescent of air in the periphery along with dilated jejunal loops (Figure 2A). The axial plain and contrast images showed an inhomogeneous attenuation hypodense non-enhancing mass 16×11×8 cm in size, occupying almost the

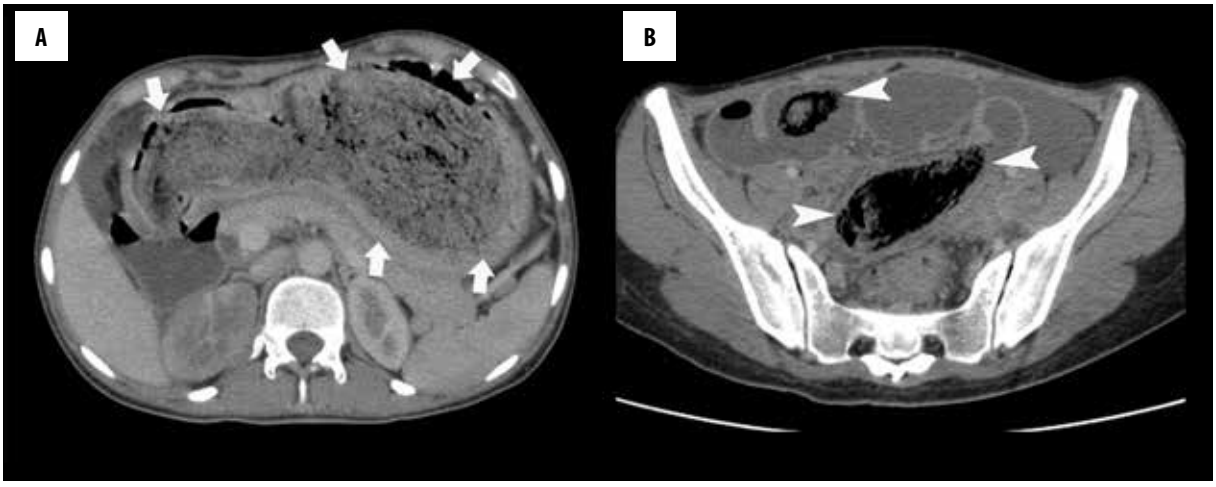


Figure 3. Axial contrast- enhanced CT image (**A**) showing the gastric bezoar with air in the interstices and peripheral air crescent (arrows). Axial contrast- enhanced CT image at the level of the pelvis (**B**) showing the separate small bowel bezoarlets (arrowheads).



Figure 4. Intraoperative photograph showing the gastric bezoar along the small bowel bezoarlets.

entire stomach with pockets of air in the interstices (Figure 3A). The small bowel loops were dilated with presence of multiple (five) similar but smaller hypodense masses in the jejunum and ileum (Figure 2B, 3B). The largest among the small bowel masses measuring 8.5×3.3 cm was impacted in the distal ileum leading to intestinal obstruction. A mild amount of free fluid was noted in the abdomen and pelvis.

Patient subsequently underwent exploratory laparotomy with gastrostomy and enterotomy, under general anaesthesia. A large trichobezoar was extracted from the stomach and the smaller bezoars were milked and extracted from the ileal enterotomy site (Figure 4). The patient recovered uneventfully and was discharged after a week.

Discussion

Trichobezoar was reported for the first time in 1779 by Baudamant. The condition most commonly occurs in young females with an underlying psychiatric disorder [2,7]. The ingested hair get entrapped within the musosal folds of the stomach as their slippery nature resists propulsion [3,4]. Peristalsis coupled with continued ingestion of hair results in formation of a cast which takes the shape of the stomach. Decay and fermentation of trapped fats in the interstices results in a rancid smell [5].

Rapunzel syndrome is a bizarre presentation of trichobezoar which is named after a princess with long tresses in a fairy tale written by Grimm brothers in 1812. The syndrome was first described by Vaughan in 1968 [6]. The criteria for defining Rapunzel syndrome has been used variably by different investigators. Naik et al. [4] studied 27 cases of Rapunzel syndrome and identified three common features: 1) trichobezoar with tail; 2) extension of the tail at least to the jejunum, and 3) symptoms of obstruction.

Expectedly, most of the cases of Rapunzel syndrome are reported from areas where females traditionally keep long hair. The patients may remain symptom- free for years. When symptomatic, the commonest presenting signs are abdominal pain, vomiting or obstruction. Less commonly the patient may present with weight loss, anorexia, hematemesis or intussusceptions [7].

A history of trichotillomania or halitosis and patchy alopecia may provide clues for diagnosis.

Plain abdominal radiograph may show mottled radiolucencies in the stomach along with features of intestinal obstruction like dilated bowel loops. USG may reveal intraluminal mass(es) with a hyperechoic curvilinear surface and casting strong acoustic shadow along with features of associated intestinal obstruction if present. In Rapunzel syndrome CT is the preferred imaging modality for diagnosis and shows a hypodense intragastric lesion with a mesh- like pattern containing air. While the mesh itself shows relatively little oral contrast, the margins may show a prominent rim of contrast [8,9]. The tail component of the trichobezoar is seen as a similar rounded area of mottled hypodensity in the small bowel [10]. MRI is not the preferred modality in this syndrome because the low signal intensity of bezoar can be confused with air.

Our case showed typical radiological findings described in literature. In addition, the tail component of the trichobezoar had broken up into multiple fragments of varying

sizes in the small intestine. The largest of those fragments had got impacted in the distal ileum and was the cause of intestinal obstruction. Though there is substantial literature on the 'classic' Rapunzel syndrome i.e. trichobezoar with a continuous tail extending for varying lengths into the small gut, the description of cases with a broken or fragmented tail is lacking. Pubmed search of literature yielded only one case with findings similar to our case [11]. In this case authors described the occurrence of simultaneous trichobezoars in the stomach and distal ileum. Our case is probably the first one to describe the USG, CT and surgical findings in a Rapunzel syndrome with a tail broken into multiple separate pieces resulting in intestinal obstruction. We also propose to name the fragments of the broken tail as 'bezoarlets'. This word is suitable for describing the tail fragments as it suggests origin from the larger gastric bezoar and the suffix 'lets' conveys that they are smaller in size.

The management and treatment of Rapunzel syndrome entails surgical removal of the mass/masses. In addition a psychiatric assessment and a long- term follow-up are advocated with parental or spouse counselling as an integral part of treatment to prevent recurrence.

Conclusions

Rapunzel syndrome is a rare form of trichobezoar with a tail of variable length extending at least into the jejunum. While the USG can help in raising a suspicion, definitive diagnosis will require a CT. Fragmentation of the tail into multiple smaller bezoarlets can occur and careful evaluation of CT images is essential to precisely localize each bezoarlet so that they may be extracted at the time of surgery.

Conflict of interest

The authors declare they have no conflict of interest.

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