

Rapunzel Syndrome with Small Bowel Malrotation

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INTRODUCTION

A bezoar refers to a mass of indigestible foreign material found in the gastrointestinal tract, mainly stomach. The second most common bezoar is the trichobezoar, which usually occurs in the young psychiatric female patients with history of trichotillomania and trichophagia.¹

Malrotation is an abnormal position of the duodenojejunal junction bowel within the peritoneal cavity and usually involves both the small and the large bowel. Malrotation is accompanied by abnormal bowel fixation by mesenteric bands or absence of fixation of portions of the bowel, leading to increased risks of bowel obstruction, acute or chronic volvulus, and bowel necrosis. The term malrotation applies to a wide range of intestinal anomalies, from a readily apparent omphalocele in newborns to asymptomatic nonrotation of the large and small bowel in adults.²

This is a case report of a 18 -year-old female patient came with complaints of nausea, vomiting on and off, constipation and fullness of abdomen. Ultrasound, X-ray abdomen erect, and Computed Tomography scan were done, which suggested trichobezoar and small bowel malrotation.

PRESENTATION OF CASE

An 18-year-old female presented to the Emergency Department with nausea, vomiting on and off, constipation and fullness of abdomen since 3 months and fever since one month. She also had loss of appetite and weight loss for 4 months. On abdominal examination well defined mass was felt in left hypochondrium of approx. 8 X 6 cm. It was firm in consistency and mobile on palpation. Her cardiac and respiratory systems were normal.

- Routine investigations were done, revealed Haemoglobin (Hb) of 9.9 g/dl and TLC of 15,000
- Patient was referred to the Radiology Department for further evaluation

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X Ray Abdomen Erect

Shows Distended J Shaped Stomach with Mottled Gas Pattern without Attachment to the Stomach Wall and Air-Fluid Level in the Second Part of Duodenum.³

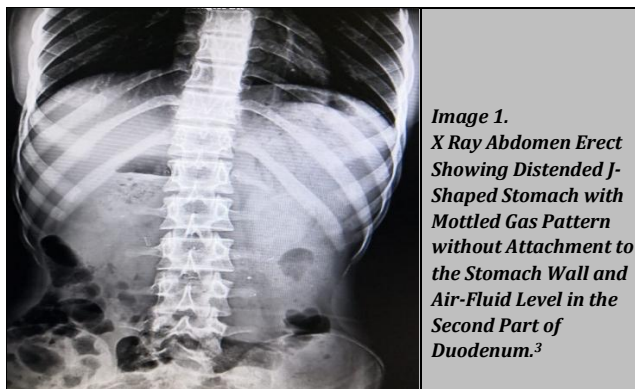


Image 1. X Ray Abdomen Erect Showing Distended J-Shaped Stomach with Mottled Gas Pattern without Attachment to the Stomach Wall and Air-Fluid Level in the Second Part of Duodenum.³

Ultrasonogram (USG) Abdomen

- An echogenic mobile shadow in the stomach with dense posterior acoustic shadow (image 2).
- Multiple enlarged lymphadenopathy of size up to 1 cm (in SAD).
- Circumferential wall thickening of stomach, jejunal and ileal loops with mildly dilated small bowel loops
- Moderate amount of free fluid.

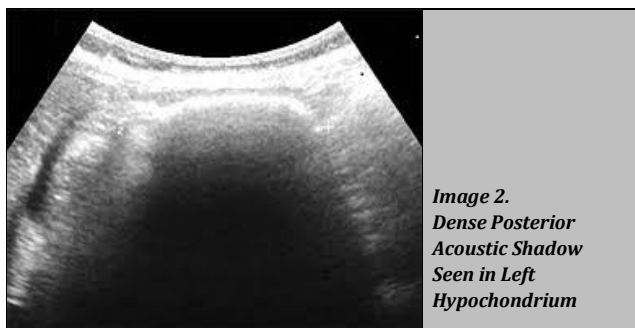
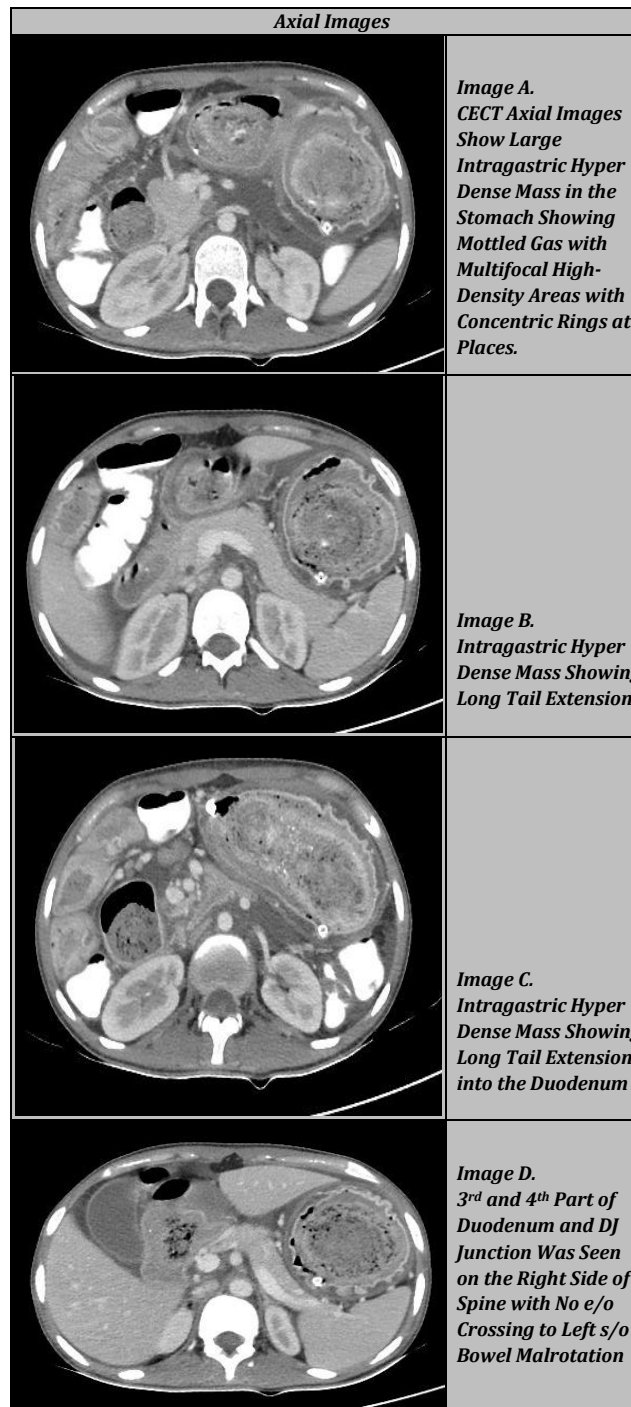


Image 2. Dense Posterior Acoustic Shadow Seen in Left Hypochondrium

Computed Tomography (CECT) Abdomen

- Axial Images (A and B) show markedly distended stomach and duodenum with large intragastric hyper dense mass showing mottled gas with multifocal high-density areas with concentric rings at places. The lesion was markedly distending the gastric lumen and showing its mass effect on Ryle's tube by displacing it along lesser curvature. It was forming a cast of stomach of size 6.5 X 6.8 X 22 cm. A long tail like extension of it was seen into the duodenum as well as proximal jejunum causing their luminal distension. Few similar smaller hyper dense lesions were also seen in distal small bowel. There associated diffuse circumferential gastric and proximal small bowel wall thickening⁴
- Axial Image (C and D) 3rd and 4th part of duodenum and DJ junction was seen on right side of spine with no e/o crossing to left s/o bowel malrotation. However colon is normal in position with normal IC junction. SMA and SMV relation was maintained

- Multiple enlarged lymph nodes were seen in mesentery in right upper quadrant up to 11 mm in SAD Other abdominal organs were normal. Based on imaging, she was diagnosed with trichobezoar.



Axial Images

Image A. CECT Axial Images Show Large Intra gastric Hyper Dense Mass in the Stomach Showing Mottled Gas with Multifocal High-Density Areas with Concentric Rings at Places.

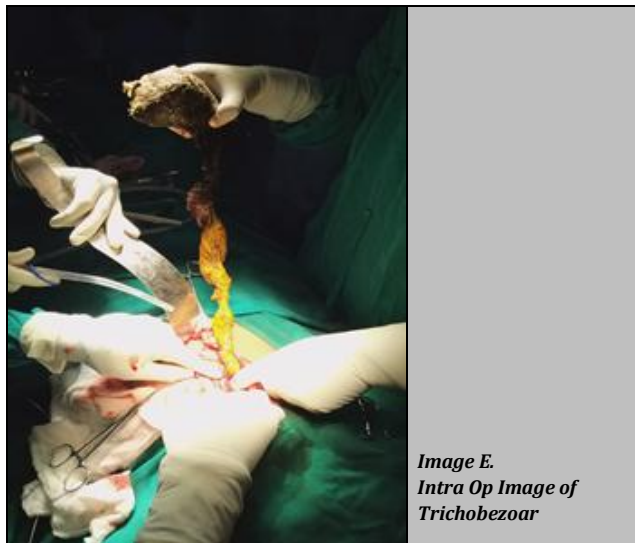
Image B. Intra gastric Hyper Dense Mass Showing Long Tail Extension

Image C. Intra gastric Hyper Dense Mass Showing Long Tail Extension into the Duodenum

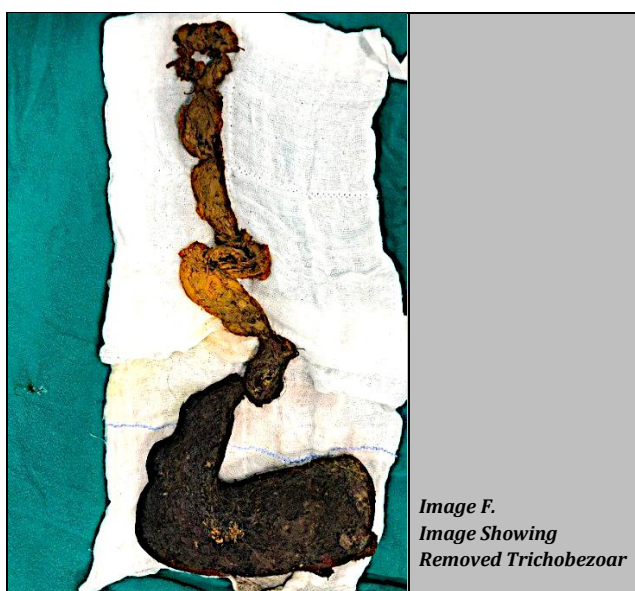
Image D. 3rd and 4th Part of Duodenum and DJ Junction Was Seen on the Right Side of Spine with No e/o Crossing to Left s/o Bowel Malrotation

DISCUSSION OF MANAGEMENT

Patient underwent exploratory laparotomy which revealed trichobezoar (images E and F). Postoperatively the patient was put on regular psychiatric therapy and endoscopic follow up.



*Image E.
Intra Op Image of
Trichobezoar*



*Image F.
Image Showing
Removed Trichobezoar*

DISCUSSION

Bezoars are foreign material in the GIT, mainly the stomach. Bezoars composed of hair are called trichobezoars. Most patients with trichobezoars suffer from psychiatric disorders including trichotillomania (pulling out of their own hair) and trichophagia (eating of hair). Young women between 13 and 20 years of age with psychiatric disorders are most commonly affected.

It has been estimated that only 1% of patients with trichophagia develop a trichobezoar.^{5,6} Hair strands escapes peristaltic propulsion, and are therefore retained in the folds of the stomach. As more hair accumulates, peristalsis causes it to be enmeshed into a ball. This further leads to gastric atony.

The ball of hair becomes matted together and assumes the shape of the stomach.^{7,8} Trichobezoar putrid smell is due to decomposition and fermentation of fats and has a glistening shiny surface due to the mucous covering it.⁹ In most cases, the bezoar is confined within the stomach. If it extends into the jejunum, ileum or even further it is called as Rapunzel syndrome.

CONCLUSIONS

Usually, there are no symptoms until, it reaches substantial size. The most common features are abdominal pain, abdominal mass, gastrointestinal obstruction, weight loss and poor appetite. Complications include ulcers, perforation, peritonitis, intussusceptions, malnutrition, rarely obstructive jaundice, pancreatitis and fatalities.

Diagnostic procedures are commonly initiated owing to acute abdominal symptoms. Given the density of hair and mucus, identification on plain radiography can be difficult. Identifying the borders of the stomach, detecting gastric distension, mottled gas pattern, surrounding lucency and multiple air-fluid levels can be helpful in the diagnosis.

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