A RARE CASE OF RAPUNZEL SYNDROME VARIANT

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ABSTRACT

BACKGROUND

We are presenting a rare case of simultaneous occurrence of gastric and ileal trichobezoar presenting with intestinal obstruction.

Case Report- A 17-year-old female presented with intestinal obstruction due to multiple trichobezoars, gastric and ileal, which were removed successfully by exploratory laparotomy. Gastric and ileal trichobezoar presenting as intestinal obstruction makes this case rare and interesting. In this patient view of the clinical history of trichophagia, radiological imaging studies showed features suggestive of intestinal obstruction due to trichobezoar. Exploratory laparotomy done revealed simultaneous gastric and ileal trichobezoar which were successfully removed.

CONCLUSION

These cases emphasize the importance of careful clinical history, role of radiological studies including USG, CT abdomen, complete intraoperative evaluation of intestine at the time of dealing with a gastric and ileal trichobezoar in diagnosis of trichobezoar.

KEYWORDS

Trichobezoar, Rapunzel.


BACKGROUND

“Rapunzel” was the name of the maiden in the “Grimm brothers” fairy tale in 1812, whose long hair flowed out of her prison tower allowing her prince to rescue her. Because of the resemblance of the tail of trichobezoar extending into the small intestine to the hair of Rapunzel, this condition is given the name “Rapunzel Syndrome.” The term Bezoar refers to a tightly packed mass of fruit, vegetable matter, hair, or other material that formed in gastrointestinal tract. The term Bezoar derives from the Arabic word Badzehr which means antidote.(1) Trichobezoar is a Greek word, Trich means hair. (2) Trichobezoar consists of hair with other fibre and usually occur in young women (90%), including those with psychiatric difficulty. The incidence of small bowel obstruction due to bezoars including food boluses is 0.3-6%. (3) Trichobezoars are often associated with trichophagia (hair swallowing). Trichotillomania may be unintentionally done and is part of the DSM IV psychiatric classification of impulse control disorders. (4,5) Trichobezoars most commonly occur in adolescent females. (6) Acute intestinal obstruction during pregnancy has a reported incidence of 1 in 1500. (7)

Case Report

A 17-year-old female was admitted in GRH Madurai with chief complaints of abdominal pain, vomiting & distension for the past 5 days. One month prior to admission, she developed mild, dull, aching, constant upper abdominal pain with any food-pain relation, flatulence or retrosternal burning sensation. Soon she noticed mild slowly increasing upper abdominal swelling, gradual loss of appetite and occasional vomiting. There was no jaundice, haematemesis, fever, breathlessness, palpitation, cough, oliguria, and haematuria. Her diet was mixed, and her bowel movements were normal. Her past and family history were unremarkable. She belonged to poor socioeconomic status. She was adequately immunized, unmarried with normal menstrual history. Clinically, an anxious looking with normal vital parameters, BMI-17 kg/m2, mild anemia. There was no jaundice, lymphadenopathy, or raised JVP.

Abdominal examination revealed epigastric and left hypochondrium fullness which on palpation showed diffuse tenderness more on right iliac fossa and epigastrium. A mass of size 15x12 cm over the epigastrium which moves with respiration and its lower margin being palpable with round edges & surface over the swelling, smooth, firm in consistency extending medially up to midline & laterally up to left costal margin 2 cm below the midclavicular line. No other abnormal intra-abdominal mass, no succussion splash, no free fluid, no bruit, normal bowel sounds. PR examination normal.

Other Systemic Examination – Normal.

A provisional diagnosis of subacute intestinal obstruction with epigastric mass was made and the patient was further evaluated.

Laboratory Investigations - Hb-9 g%, TLC- 8500/cu. mm, DLC P-77%, L-20%, E-02%, M-01%; RBC-hypochromic, microcytic. Platelets-2.5 lakhs/cu. mm. Urine examination normal, LFT-normal, blood urea 25 mg%, Sr. Creatinine 0.8. Sr. electrolytes-normal. Total serum protein 3.7 g%, Sr. albumin 1.2 g%, Sr. globulin- 2.5 g%. Plasma glucose 109 mg%.
Imaging Studies

Figure 1. X-ray Abdomen– Multiple Air Fluid Level

Figure 2. CECT Abdomen: Large Hypodense Ball noted in the Stomach

Figure 3. Intraoperative Finding showing a Dilated Stomach

Figure 4. Gastrotomy being Done

Case Report

CECT Abdomen
Large hypodense ball noted in the stomach suggestive of gastric trichobezoar and small intestinal obstruction due to ileal trichobezoar.

UGI Scopy
Stomach– huge trichobezoar in the entire stomach extending into the duodenum obstructing the gastric outlet, duodenum not entered.

Final Diagnosis– Subacute Intestinal Obstruction with Gastric & Ileal Trichobezoar
Patient was planned for elective explorative laparotomy, underwent elective gastroscopy. A large trichobezoar, occupying fundus, body, greater curvature, lesser curvature. Ileostomy was done at around 20 cm from the ileocelecal junction and retrieval of the trichobezoars was done. Both specimen weighed about 720 grams. Patient made uneventful recovery and was followed for the past one year and there was no recurrence.

Operative Images
DISCUSSION
Clinical manifestations of trichobezoar are nonspecific abdominal pain, nausea, constipation but trichobezoars can lead to serious complications like bowel obstruction, haemorrhage or perforation\(^{(8,9)}\). Although 1 in 2000 children suffer from trichotillomania, only half of the patients give history of trichophagia and just 1% of these individuals eat enough hair to accumulate trichobezoars that require surgical intervention\(^{(10)}\). Small bowel bezoars are managed surgically if intestinal obstruction supervenes. At laparotomy, attempts can be made to advance the bezoars into colon manually. If these efforts are unsuccessful, enterotomy and extraction are necessary. One must guard against infrequent occurrence (4-17%) of multiple bezoars by examining the stomach and the entire small bowel at laparotomy. Preoperative endoscopy is important in cases of small bowel obstruction as a result of bezoars in order to recognise unsuspected gastric or duodenal bezoars and extract or fragment these if possible, as they may be readily missed upon attempted palpation specially when there has been previous Gastric surgery.\(^{(11)}\) In up to 10% of
patients of trichotillomania trichophagia; one third of patients of trichophagia develop trichobezoar.\(^5\) Plain abdominal radiographs and barium meal study are helpful in confirming gastric trichobezoar and excluding calcified masses which are confusing in ultrasound abdomen.\(^12\) Trichobezoars are intestinal. The term Rapunzel syndrome was given to Trichobezoar extending continuously through the entire length of small intestine as a tail and was described by Vaughan et al in 1968.\(^13\) Small bowel obstruction from trichobezoar is very uncommon because trichobezoars usually do not migrate into small bowel. The small Intestinal bezoars has been rarely reported.\(^14,15,16\) Bezoars may be removed either laparoscopically or by open laparotomy.\(^17\) Due to high success rate, relative low complexity and ability to carefully examine the entire gastrointestinal tract for satellites in a short period of time, laparotomy is still considered the treatment of choice. Endoscopic fragmentation may be attempted, but often fails.

**CONCLUSIONS**

Rapunzel syndrome, a gastric trichobezoar with intestinal extension is quite uncommon and should be considered strongly in a young patient with abdominal pain, and nontender abdominal mass with history of trichotillomania and trichophagia. Surgical removal is the gold standard treatment of choice. Psychiatric treatment prevents its recurrence.

**REFERENCES**