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HUNGRY FOR HAIR - RAPUNZEL SYNDROME: A CASE REPORT

General Surgery

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ABSTRACT

Rapunzel syndrome is a rare condition where bezoar made of ingested hair is found in stomach or intestine. Incidence is <18% (In patients with trichotillomania). Small bezoars can be removed endoscopically whereas large ones require surgery. The medical & psychiatric treatment of Trichotillomania should not be underestimated & early diagnosis & treatment play's a major role in saving patient's life. We describe a case of 14yr old girl with Trichophagia and Trichotillomania who presented to our surgery op with history of pain abdomen and vomiting. Clinical examination & Investigations showed the presence of a Trichobezoar causing Gastric Outlet Obstruction requiring immediate surgical intervention.

KEYWORDS

Rapunzel Syndrome, Trichobezoar, Trichotillomania, Trichophagia, Gastric outlet obstruction (GOO)

INTRODUCTION

Bezoars are concretions of vegetable fibers that accumulate in the GIT. In humans, the most common type of bezoar is the trichobezoar, which is made of hair. Other types - Phytobezoars, lactobezoars.[1]

The name Rapunzel Syndrome comes from the Fairy tale by Grimm brothers.[1][2][4] Trichobezoars are not associated with alterations in gastrointestinal motility but with underlying psychiatric disorders, and mostly present in adolescents. Human hair is resistant to digestion and peristalsis due to its smooth surface resulting in its accumulation within the stomach. It leads to impaction of hair along with mucus and food within stomach resulting in the formation of trichobezoar.[3] Rapunzel syndrome is a rare form of trichobezoar extending into the small intestine, hence the name.[1][2][3].

It was reported that abuse, pica, mental disorders, depression, anorexia nervosa, or ocd might represent potential comorbidities.[3] The clinical picture is usually scarce in the early stages resulting in delay of diagnosis.

If not recognized, trichobezoar continues to grow in weight and size occupying complete stomach increasing risk for gastric mucosal erosions or ulcerations, and even perforation.[3][5] Other complications reported are protein-losing enteropathy, intussusception, obstructive jaundice, pancreatitis, or even death in cases of unrecognized bezoars or delayed diagnosis.[3][6] Various options for treatment are medical and enzymatic degradation, endoscopic, laparoscopic, or laparotomic removal .[3][5] The majority of these cases presented between 13 and 20 years of age.[1] This is a rare condition with less than 64 cases reported till date. We report a case of 14-year-old girl.

Case Report:

A 14-year-old girl presented to Owaisi Hospital & Research Centre with complaints of pain in epigastrium since 4 months, colicky type, associated with non-bilious vomitings 2-3 episodes/day since 2-3 weeks. Her parents described that the she had a habit of plucking and swallowing hair since 4 years. She had no evident psychiatric illness and had no prior surgical history. No other complaints were reported.

On Examination:

Our patient was fully conscious, dehydrated and pale with no signs of icterus. She was hemodynamically stable. On inspection there was an obvious asymmetrical abdominal distension with centrally inverted umbilicus. On palpation abdomen was soft, with no signs of guarding/rigidity and an immobile hard palpable mass was felt in the epigastric and left hypochondriac region measuring approximately 15*10cms. The mass had ill-defined edges and smooth surface and was not tender, compressible or pulsatile. On auscultation bowel sounds were normally heard.

Investigations:

Hb: 9.3gm/dl, RBC: 4.6 million/cumm, WBC: 7200/cumm, Platelets: 3.9 lakhs/cumm. Other laboratory findings, including electrolytes, acid balance, LFT, RFT were within normal limits.

Radiological Findings:

USG abdomen revealed a large complex mass occupying entire stomach with few visible thick gastric rugae. It was casting a huge semilunar echo density with distal shadowing obscuring underlying details. [fig-1,2]

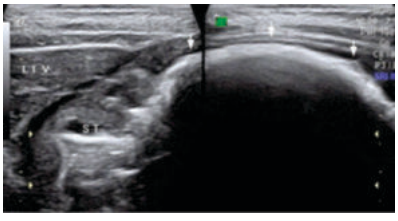

Fig-1: Huge Semilunar crescentic echo density seen with distal shadowing.

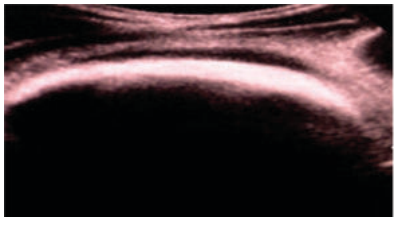

Fig-2: Large semilunar echo density with posterior acoustic shadowing.

Ct Abdomen:

shows an intragastric well-circumscribed mass consisting of 'mottled gas pattern' due to the presence of entrapped air and food debris. Normal stomach wall can be traced completely separate from the lesion; wall thickening was noted. [Fig-3,4,]

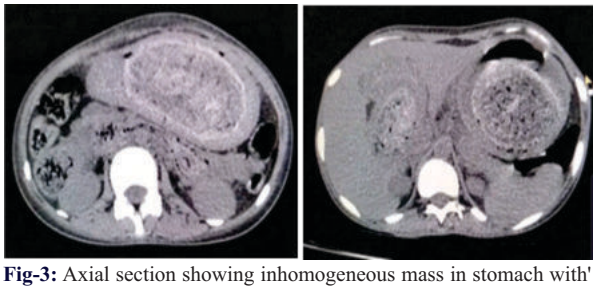

Fig-3: Axial section showing inhomogeneous mass in stomach with' mottled gas pattern' or 'compressed concentric rings' pattern.

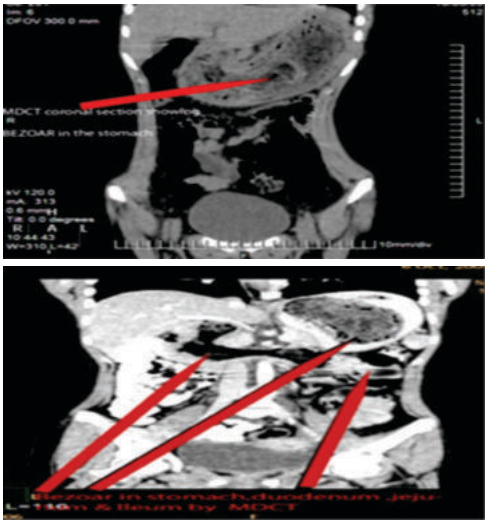

Fig-4: Coronal sections showing extent of mass till small intestine.

UGIE: Large trichobezoar noted extending beyond pylorus causing GOO. Scope could not be passed beyond duodenum. [Fig-5]

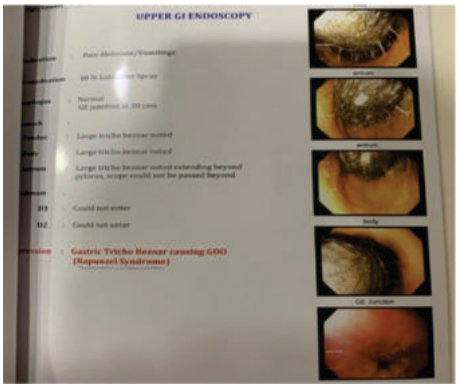

Fig-5: UGIE showing a trichobezoar causing GOO.

Surgery:
Exploratory Laparotomy + Gastrotomy + Removal of trichobezoar under GA.

Findings:
A gastric fundus shaped, slate grey tissue mass measuring 21*20*5cms broad at the top and tapering towards the tail, extending from fundus to second part of duodenum. [Fig-6]

Fig-6: Trichobezoar taken the shape of the stomach and intestine-Rapunzel Syndrome.

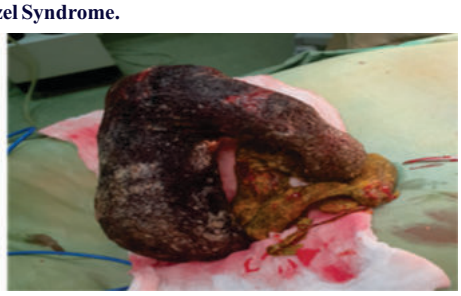

Fig-7: Trichobezoar-specimen.

Histopathological Examination:
Sections from the specimen showed the presence of hair shafts, hair roots and presence of gastric and duodenal mucous glands thus confirmed the diagnosis of gastric trichobezoar.

DISCUSSION:
Trichotillomania & trichophagia were reported following the psychiatric consultation after noticing that the patient had some bald patches on her scalp resulting from the habit of pulling out her hair and secretly swallowing it.[7]

The characteristic symptoms of trichobezoars are abdominal pain, nausea, vomiting progressing to gastric outlet obstruction and peritonitis.[8] Obstruction of the upper digestive tract is the most common clinical manifestation of this disorder.[9] UGIE is the gold standard investigation for confirming trichobezoar.

Treatment options include the use of chemical substances in the stomach to dissolve the material, and mechanical fragmentation. However, in large trichobezoars, these methods are not successful. [6][10][12]

Although rare, it may present as emergency and requires proper preparation by surgeons. In our case the trichobezoar was very big weighing approximately 1.5kgs, therefore endoscopic removal was not possible. Laparotomy was successful in most cases. Reports on the use of video laparoscopy are encouraging. However, sample sizes have so far been small and hence, the role of this technique in the treatment of the Rapunzel syndrome remains to be defined.[11][12] The literature provides no evidence of superiority of endoscopy or laparoscopy over laparotomy. The lack of invasiveness of these techniques doesn't seem to outweigh the disadvantages and complexity of these procedures.

CONCLUSION:
The medical and psychiatric sequelae of trichotillomania shouldn't be underestimated, early diagnosis and treatment plays a role in saving patient's life.[7][12] Although laparotomy is the main modality of treatment, it should be supplemented with pharmacotherapy and behavioral assessment to prevent recurrence.

In our case, we have treated the patient as a whole rather than just treating the symptoms, trichobezoar was removed, malnutrition, iron deficiency anemia, hypoalbuminemia was also corrected. The patient didn't have any post-operative complications, and was referred to psychiatrist for the management of trichotillomania, trichophagia and counselling in order to prevent recurrence.

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