

A RARE CASE OF RAPUNZEL SYNDROME CAUSING SMALL BOWEL OBSTRUCTION

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ABSTRACT

Rapunzel syndrome is a rare form of bezoar in which trichobezoar is seen mostly in sufferers with records of psychiatric disorders, trichotillomania (dependency of hair pulling) and trichophagia (morbid addiction of chewing the hair), consequently developing gastric bezoars. The fundamental signs are vomiting and epigastric ache. In this case report, we describe this syndrome in a younger female.

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INTRODUCTION

A bezoar is a mass, ingested material that gets trapped in the gastrointestinal tract. Bezoars can be made of up vegetable or fruit fibres (phytobezoars), milk curds (lactobezoars) or any indigestible material that is ingested. Trichobezoar is the most common type of bezoar and is made up of human hair and found in the stomach. [1]

This syndrome is named after the woman with the long tresses within the fairy tale. The Rapunzel syndrome became first said within the literature by Vaughan *et al.* in 1968 [1] and so named because of the length of the hair and the uniqueness of the situation that are characteristics both of the fairy tale and of the clinical cases described inside the report published. There are numerous different styles of presentation of this syndrome; but, in popular, it entails the presence of a gastric trichobezoar with a protracted tail extending beyond the duodenum, as found in the case pronounced right here. The component chargeable for this syndrome is the compulsion of patients to drag out their hair and swallow it, approaches referred to as trichotillomania and trichophagia, problems that have an effect on young ladies with or without recognised psychiatric issues. [1]

Case report

A young aged female with psychiatric symptoms presented with acute abdominal pain and vomiting in the emergency department. There were similar episodes in the past medical history of the patient.

The patient was slightly pale, this being the only abnormality found at physical examination. She had a history of slightly retarded neural development. The patient's mother admitted that she had a habit of pulling out her hair and secretly swallowing it.

Ultrasound of the patient was done and revealed dilated stomach and small bowel loops showing echogenic masses with intense internal shadowing. CT abdomen of the same patient revealed an intragastric well-circumscribed in homogenous mass consisting of 'mottled gas pattern' due to the presence of entrapped air and food debris. Similar masses were seen in the terminal ileum causing small bowel obstruction. The distal large gutis collapsed. No contrast enhancement was seen within visualized masses. Findings were suggestive of trichobezoars causing small bowel obstruction. The patient was taken for surgery to relieve a small bowel obstruction. On surgery, trichobezoars were found out lodged in the antrum region of the stomach and distal ileum. A rare case of Rapunzel syndrome causing small bowel obstruction was reported.



Fig 1 Ultrasound showing characteristic echogenic content with dense internal shadowing.

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Fig 2 CT images showing dilated stomach with trichobezoars within the lumen

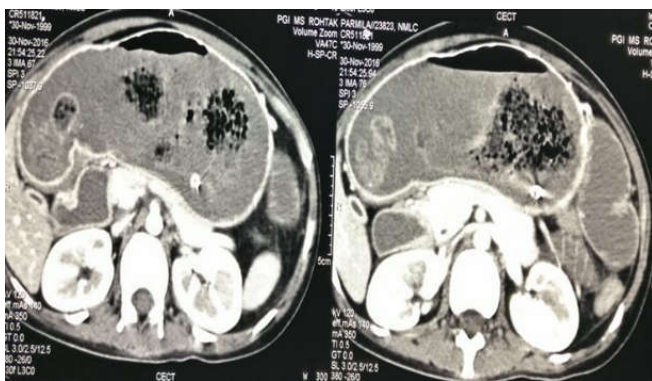


Fig 3 CT scan of the same patient shows intragastric and ileal contents showing Characteristic mottled appearance with small bowel obstruction.

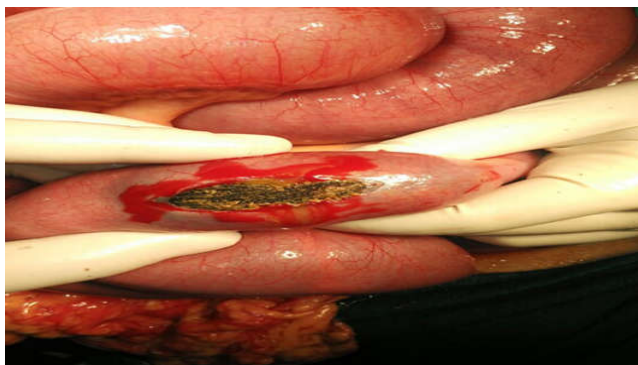


Fig 4 Of the patient reveals dilated small bowel loops with internal hair filled lumen.



Fig 5 Reveals hair ball i.e. trichobezoar removed from bowel.

DISCUSSION

Trichobezoar, a hair ball within the proximal gastrointestinal tract, is an extraordinary condition nearly exclusively seen in younger girls. [1][2] Human hair is resistant to digestion as well as peristalsis. Therefore it accumulates among the mucosal folds of the belly. Over a length, continuous ingestion of hair leads to the impaction of hair together with mucus and meals, causing the formation of a trichobezoar. In most

instances the trichobezoar is limited inside the stomach. In a few cases, but, the trichobezoar extends via the pylorus into jejunum, ileum or even colon. This situation is called Rapunzel syndrome [3] [4]. Its part can break off and pass into the small intestine, causing intestinal obstruction [5] [6]. The frequent place of bezoars within the belly is due to the holdup by way of the stomach which allows entangling new hair into the already formed solid. The bezoar takes on a glistening vivid floor from the mucus that covers it. Decomposition and fermentation of fats deliver the bezoar and affect the person's breath. [6] The acidic contents of the belly denature the hair protein and supply the bezoar its black color.[7]The causes of a bezoar can be due to the presence of indigestible fabric inside the lumen, gastric dysmotility (such as preceding surgery which includes vagotomy and partial gastrectomy, and so on.). Bezoars are found mainly in young ladies and so named because of the substances which are swallowed and produce them, which include hair (trichobezoar), phytobezoar (vegetable fibers), diospyrobezoar (persimmon fibers), or pharmacobezoar (tablets/semi-liquid masses of drugs). [8]

Gastric bezoars must be differentiated from the pictures produced via retained food particles within the belly. The marked acoustic shadowing in the back of the echogenic band produced by means of a bezoar is different from "grimy" shadowing generated via ingested meals and fuel inside stomach on ultrasonography. [9] On ultrasound, the featured appearance persists irrespective of the angulation of the transducer, the alteration of the location of the patient, or the management of clean water. This excludes the pancreatic pseudocyst, a splenic or renal mass, a noncalcified gastric tumour, a gastric duplication cyst, or a gastric outlet obstruction. [10]On CT, a cellular intraluminal mass along with "compressed concentric jewelry" is seen. There is a combined density sample because of the presence of entrapped air and meals particles. Similarly to the function look as described above, the free mobility of the mass in the belly lumen may be visible in supine and prone views. CT helps in the better configuration of the bezoar and appropriately identify its location. Further, CT helps in better characterization and helps to differentiate from other pathologies that would be difficult to find out on simple radiography or ultrasound. [11]

The treatment of huge bezoars and concretions is largely surgical. Gastric bezoars can be retrieved endoscopically or via gastrotomyendorsed bezoar extraction by way of multiple enterotomies in cases of Rapunzel syndrome. [12]Its far obligatory to do a radical exploration of the rest of the small gut and the stomach to see bezoars in the rest of the gut and stomach. Other methods like modified needle-knife (bezotome) and a modified lithotripter (bezotripter) have been successfully used. The affected person's psychological problems additionally need to be treated. [13]

CONCLUSION

It is very important to make an early diagnosis of Rapunzel syndrome to avoid unnecessary investigations and confusion with another alternative diagnosis.

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