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CASE REPORT

Rapunzel Syndrome: The not-so-fairy tale of a long tail

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Abstract:

Background: Rapunzel Syndrome associated with Trichobezoar secondary to a psychiatric illness Trichotillomania, is often a rare diagnosis of Gastrointestinal symptoms, occurring predominantly in young & adolescent females.

Clinical Description: We present here a case of such disease in a 12-year-old female who presented to us with recurrent vomiting & pain abdomen for the last 2 months.

Management & Outcome: On thorough examination & investigations, she was suspected to be a case of trichobezoar on ultrasound abdomen, which was confirmed by upper GI endoscopy. Mass was surgically removed from the patient's abdomen with a long tail of Rapunzel.

Conclusion: Although trichobezoar is a rare condition, it must be considered in patients with trichotillophagia & abdominal symptoms. Symptomatic trichobezoars are often so large that they are palpable on simple clinical examination as in this case. USG may be inconclusive and endoscopy is confirmative. Undiagnosed or untreated trichobezoar continues to grow in size & weight, resulting in devastating complications, such as gastric ulcers, perforation, or even death.

Keywords: Trichobezoar; Trichotillomania; trichotillophagia; Rapunzel Syndrome

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Introduction

Indigestible accumulation of foreign material in the gastrointestinal tract is called 'bezoar'. The collection of fibers or plants is called 'Phytobezoar', collection of milk is 'Lactobezoars', collection of hairs is termed as 'Trichobezoar' & collection of medication is 'Pharmacobezor' [1]. Trichobezoar is more common in young girls & results due to psychiatric conditions like 'trichotillomania' (hair pulling) & 'trichophagia' (hair swallowing). A rare & unusual presentation of trichobezoar is when hairs extend into the intestines from the stomach & this condition is called as Rapunzel Syndrome. Removal by using an endoscope may be possible for small trichobezoars while large ones need surgical intervention [2]. Here, we report a case of Rapunzel Syndrome in which the mass was removed surgically.

Case Report

A 12-year-old female child weighing 27 kg presented to Out Patient Department of our hospital with complaints of pain abdomen for the last 2 to 3 months, which was insidious in onset, mainly in the epigastric region, with no radiation, mild to moderate in intensity, no diurnal variation, occasionally colicky in nature with no aggravating or relieving factors. She also had a history of vomiting for the last 2 to 3 months, mostly after ingestion of food, approximately 3-5 episodes per day, non–bilious in nature, projectile and mainly containing food particles. There was no history of fever, headache, constipation, or abdominal distension.

General examination revealed pallor. On per abdomen examination, there was an intra-abdominal mass palpable in the epigastric region extending up to the left hypochondrium which was approximately 5x5 cm in size, round in shape, with smooth surface, well-defined margins with normal skin overlying the mass, firm in consistency, non-tender, moving with respiration, non-pulsatile, non-fluctuant, non-reducible or compressible, non-trans-illuminant & non-pitting on pressure. There was no hepatosplenomegaly. Initially, a differential diagnosis of malignancy was thought of.

The patient was admitted to Pediatric Intensive Care Unit and a workup was done, which revealed Hb- 9.2 gm/dL %; TLC- 9,420/µL; DLC- $N_{67}L_{22}$; PLT- 2.79 lacs/µL; MCV-78.5 fl; MCH- 23.2 pg; RDW–14.5%; Reticulocyte Count – 2.6%; S. Creatinine- 0.66 mg/dL; B. Urea – 30 mg/dL; Na – 130 mEq/L; K- 3.3 mEq/L; OT/PT – 47/24 Units/L; Total Bilirubin- 0.28 mg/dL.

Ultrasound abdomen revealed an echogenic lesion with posterior acoustic shadowing of size approximately 68 x 62mm in the left hypochondriac region, with a possibility of Trichobezoar.

History was reviewed and a history of Trichotillomania and Trichophagia for the last 2 years was elicited due to which parents trimmed her hair.

Trichobezoar of about 10 x 15 cm was confirmed on Upper GI endoscopy and surgery was planned after clearing the pre-anesthetic checkup.

A midline incision was given over the epigastric region, planes were dissected, the anterior stomach wall was opened, trichobezoar was identified & was extracted along with the tail of Rapunzel of about 50 cm, extending up to jejunum (Figure 1). The stomach was then closed &

skin sutures were finally placed thus completing the surgery. The patient was subsequently kept nil per oral, along with intravenous fluids and antibiotics. She recovered well. Feeds were started on the 7th day and the patient was discharged on post-operative day 10. A psychiatry reference was done subsequently.

Discussion

Accumulation of exogenous matter in the stomach or intestine, is called a bezoar which is predominantly composed of food & fiber. The term 'bezoar' is derived from the Arabic word 'badzehr' which means antidote [1]. Females with underlying personality disorders or neurologically impaired persons are more prone to develop bezoar. Apart from this, patients who have undergone abdominal surgery are also at higher risk of developing bezoars, as it decreases the size of the stomach, interferes with the passage of gastric contents and leads to reduced secretion of gastric acid [3]. Diabetes mellitus, autoimmune diseases, peptic ulcer disease, Crohn's disease, carcinoma of the GI tract, hypothyroidism & excessive fiber intake are some of the other predisposing factors for bezoar formation [4]. The peak age of onset of symptoms is 2nd decade of life [5].

Trichotillomania, a psychiatric condition, is most frequently associated with this condition and the most severe form is known as Rapunzel syndrome (hair bezoar extending beyond the stomach to the small intestine).[6] It is named after a tale written in 1812 by the Brothers Grimm, about a long-haired young maiden, Rapunzel, who lowered her hair to the ground from a castle in order to permit her young prince to climb up [7]. Rapunzel Syndrome was described for the first time in 1968 by Vaughan et el., and it is almost exclusively seen in young females [8].

Phytobezoars are composed of a combination of plant & animal material. Lactobezoars were previously found most often in premature infants & can be attributed to the high casein or calcium content of some premature formulas. Swallowed chewing gums can occasionally lead to a bezoar.

Symptoms of gastric outlet or partial intestinal obstruction, including vomiting, anorexia, & weight loss are the main manifestations of trichobezoar. Complaints of abdominal pain, distension and severe halitosis, with physical examination demonstrating patchy baldness & a firm mass in the left upper quadrant are characteristics of trichobezoar [9]. Patients occasionally have iron deficiency anemia, hypoproteinemia, or steatorrhea caused by associated chronic gastritis.

An abdominal plain film suggests the presence of bezoar, which can be confirmed by ultrasound or CT examination which show a nonhomogeneous, non-enhancing mass within the lumen of the stomach or intestine [4].

Bezoars in the stomach can usually be removed endoscopically if they are of small size & confined to the stomach only with no satellite masses. Lactobezoars usually resolve when feeding is withheld for 24-48 hrs. Coca-Cola has been used as a dissolution therapy for gastric phytobezoar & has been shown to be effective when used with endoscopy [10].

Sunflower seed bezoars are reported to cause rectal pain & constipation as a result of seed shells being associated with fecal impaction [11]. Endoscopic removal is indicated, as these bezoars are refractory

to enema or lavage management. Trichobezoars almost always require surgical removal as in our case.

Conclusion

Although trichobezoar is a rare condition, it must be considered in patients with trichotillophagia & abdominal symptoms. Symptomatic trichobezoars are often so large that they are palpable on simple clinical examination as in this case. USG may be inconclusive and endoscopy is confirmative. Undiagnosed or untreated trichobezoar continues to grow in size & weight, resulting in devastating complications, such as gastric ulcers, perforation, or even death.

Conflicts of interest

The authors declares that they do not have conflict of interest.

Author contibution

All authors have equal contributions to patient care, establishing a clinical diagnosis, planning investigations, management and follow-up. Dr. Deepak Mittal was the chief Operating Pediatric surgeon. All have contributed to manuscript preparation.

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