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A RARE CASE REPORT OF RAPUNZEL SYNDROME

Surgery

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ABSTRACT

Trichobezoars are masses, which are commonly encountered in patients after stomach surgery or in those with psychiatric problems, formed by the accumulation of intraluminal nondigestible substances that can lead to obstruction of the stomach and the small intestine. The anatomical changes in the gastrointestinal tract are known to cause bezoar formation. In the absence of an anatomical change, psychiatric disorders such as trichotillomania may lead to the formation of trichobezoars in the stomach. The so-called Rapunzel syndrome is the extension of the bezoars down to the duodenum and the jejunum, which is a rare condition. In this paper, a 15-year-old patient with trichotillomania is reported, who was admitted to our clinic with nausea, vomiting, and fatigue complaints, in whom a giant trichobezoar was identified, which completely filled the stomach and duodenum, and jejunum without causing obstruction.

KEYWORDS

Rapunzel syndrome, trichobezoar, trichotillomania,bezoars

TRICHOBEZOAR

Rapunzel syndrome is an extremely rare condition seen in adolescents or young females with psychiatric disorders consisting of a gastric trichobezoar with an extension within the small bowel. The delays in diagnosis are common since in its early stages, it is usually asymptomatic. We report the case of a 15-year-old girl admitted in our clinic for abdominal pain, anorexia, and weight loss. The clinical exam pointed, a palpable mass in the epigastric area, and abdominal tenderness at palpation. The abdominal ultrasound showed a impression of clumping a small bowel loops in the periumblical region with surrounding edema. The upper digestive endoscopy revealed a large elongated ball of hair occupying majority of stomach lumen extending downwards into D1 and D2 confirming the diagnosis of Rapunzel syndrome. The giant trichobezoar, measuring about 110 cm, was successfully removed through laparotomy. Although rare, Rapunzel syndrome must never be forgotten as a differential diagnosis for digestive symptoms since its early detection hinders the occurrence of further complications.

INTRODUCTION

The word “bezoar” originates from the Arabic word “bedzehr” or the Persian “padzhar,” which means “protecting against a poison,” since historically, bezoars from animal guts were use ful asanti dotestopoisons. A bezoar is an indigestible accumulation of foreign materials in the gastrointestinal tract. Because of the stomach's large capacity, gastric bezoars do not become symptomatic until they are very large. The masses are classified according to their contents. Phytobezoars include fruit fibers or plants, lactobezoars are composed of milk, trichobezoars are concretions of hair, and pharmacobezoars are composed of medications. Trichobezoars are rare conditions that consist of hair bundles in the stomach or small bowel. In young women, trichobezoars are associated with psychiatric disorders such as trichotillomania (hair pulling) and trichophagia (hair swallowing). The Rapunzel syndrome is a rare type of trichobezoar that extends into the small intestine. Some types of bezoars, including small trichobezoars, can be removed after endoscopic fragmentation. However, very large trichobezoars, such as those in a patient with Rapunzel syndrome, are resistant to endoscopic fragmentation due to the dense hair mass. Therefore, these masses must be surgically removed, despite the large scars that result. Here, we report the case of Rapunzel syndrome with a large trichobezoar. Rapunzel syndrome was described for the first time in 1968 by Vaughan et al., and it is almost exclusively seen in young females. Multiple definitions emerged for this syndrome, but overall, it represents a trichobezoar with a tail extending in the small bowel . The name of this syndrome comes from a fairy tale about a 12-year-old princess who was locked in a tower without stairs or doors and managed to escape with the help of Rapunzel's long tresses (8). Young females diagnosed with trichobezoars or Rapunzel syndrome are usually associated with a psychiatric disorder, and it was reported that abuse, pica, mental disorders, depression, anorexia nervosa, or obsessive–compulsive disorder might represent potential comorbidities.

Case Report

We report the case of a 15-year-old female admitted in our hospital for pain abdomen and vomiting

Clinical Findings

The clinical exam at the time of admission pointed out diffuse some hair loss and palpable mass in the epigastric area, and abdominal tenderness at palpation.

Diagnostic Focus and Assessment

Based on the presenting concerns and the clinical findings, we raised the suspicion of trichotillomania and trichophagia resulting in a possible trichobezoar. Initially, the patient denied to have ingested her hair, but after a thorough history, she admitted to have this habit, but rarely. **The abdominal ultrasound** a mass formed impression of clumping a small bowel loops in the periumblical region. The **upper digestive endoscopy** revealed a large elongated ball of hair occupying majority of stomach lumen extending downwards into D1 and D2 confirming the diagnosis of Rapunzel syndrome. **CECT** reveal well circumscribed inhomogenous non enhancing lesion with mottled gas pattern in the overdistended stomach separate from the enhancing gastric wall. The lesion is extending from body of stomach into the dilated duodenum and proximal/mid jejunal region.

Management

The size of this mass did not allow us to remove it endoscopically, and the trichobezoar was surgically removed by median supraumbilical laparotomy with gastrotomy (fig-3 and 4). This was about 110 cm long mass and was extended upto the ileum.The good part of this managenment is that no further incisions were required and it was retrived from a single incision without breakage.

DISCUSSION

Trichobezoars, the most common bezoars in humans, usually occur in preadolescent or adolescent girls with psychiatric comorbidities or developmental delay. Similarly, our case describes a preadolescent girl previously diagnosed with developmental delay. Gorter et al. described four cases of trichobezoars all in girls aged between 7 and 15 years, but Rapunzel syndrome was identified only in one case .Nevertheless, trichobezoars are also possible in males as reported by Hal et al. describing to the best of our knowledge the youngest patient diagnosed with trichobezoar, a 3-year- old boy. . In our case, it was impossible to attempt endoscopic removal due to the large size of the trichobezoar occupying almost the entire gastric cavity.

Laparoscopy is a relatively better alternative compared with endoscopy, but it is usually converted into open laparotomy .Nevertheless, the successful rate mightincrease as a result of the combination between laparoscopic fragmentation of trichobezoar and endoscopic removal of the fragments .Despite its potential complications, laparotomy was reported to be 100% effective for trichobezoar removal especially in the setting of Rapunzel syndrome.

Our patient also underwent a laparotomy, and the giant trichobezoar along with its tail were successfully removed without any break and via a single incision. The long-term prognosis of these patients depends completely on preventing recurrences. Therefore, parental counseling, behavioral therapy for controlling trichotillomania and trichophagia, as well as psychiatric and psychological support and close follow-up are essential in these cases. Our patient was also sent for psychiatric counselling

CONCLUSION

Rapunzel syndrome is an extremely rare cause of digestive symptoms, such as abdominal pain, anorexia, or weight loss in children. Delays in diagnosis are relatively common taking into account that it is rarely considered in the differential diagnosis of children with digestive symptoms. Increased awareness regarding risk factors for trichotillomania and trichophagia, such as developmental delay and anemia as a cause of pica, is crucial for the early diagnosis. Laparotomy remains the treatment of choice in the case of large bezoars. Thus rare, Rapunzel syndrome must never be forgotten since its early detection is very important.

Figures.

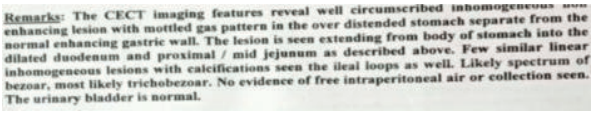
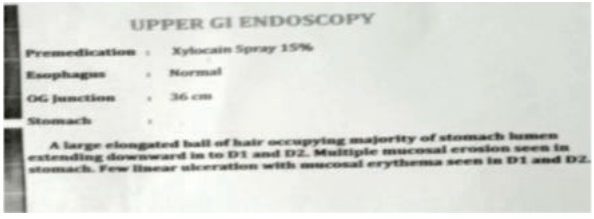

Remarks: The CECT imaging features reveal well circumscribed inhomogeneous [illegible] enhancing lesion with mottled gas pattern in the over distended stomach separate from the normal enhancing gastric wall. The lesion is seen extending from body of stomach into the dilated duodenum and proximal / mid jejunum as described above. Few similar linear inhomogeneous lesions with calcifications seen the ileal loops as well. Likely spectrum of bezoar, most likely trichobezoar. No evidence of free intraperitoneal air or collection seen. The urinary bladder is normal.

Figure 1-CT report


UPPER GI ENDOSCOPY

Premedication : Xylocain Spray 15%

Esophagus : Normal

OG Junction : 36 cm

Stomach :

A large elongated ball of hair occupying majority of stomach lumen extending downward in to D1 and D2. Multiple mucosal erosion seen in stomach. Few linear ulceration with mucosal erythema seen in D1 and D2.

Figure 2 -UGIE REPORT

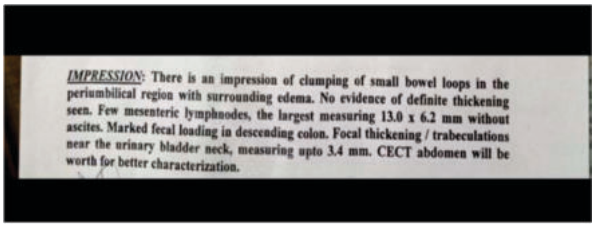

IMPRESSION: There is an impression of clumping of small bowel loops in the periumbilical region with surrounding edema. No evidence of definite thickening seen. Few mesenteric lymphnodes, the largest measuring 13.0 x 6.2 mm without ascites. Marked fecal loading in descending colon. Focal thickening / trabeculations near the urinary bladder neck, measuring upto 3.4 mm. CECT abdomen will be worth for better characterization.

Figure 3-usg report

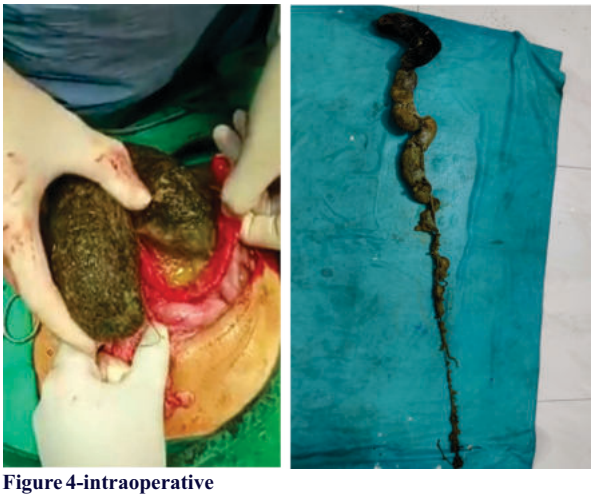


Figure 4-intraoperative

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