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clinical practice

The Rapunzel syndrome. Report of a case

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SUMMARY: **The Rapunzel syndrome. Report of a case.**

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Trichobezoar is a rare pathology in which swallowed hairs accumulate in the stomach.

An unusual form of bezoar extending from the stomach to the small intestine or beyond has been described as Rapunzel syndrome.

Trichobezoars typically cause abdominal pain and nausea, but can also present as an asymptomatic abdominal mass, progressing to abdominal obstruction and perforation. Trichobezoar with Rapunzel syndrome is an uncommon diagnosis. It is predominantly found in emotionally disturbed or mentally retarded young people.

The diagnosis may be suspected in young females with abdominal pain, epigastric mass and malnutrition, who have a history of trichophagia.

The Authors present a case of successful laparotomy removal of a giant gastro-duodenal trichobezoar in a 9-year-old girl with a history of trichotillophagia. Physical examination revealed diffuse abdominal pain and an epigastric mass. Psychodynamic aspects, clinical manifestations, diagnosis and therapautic strategies are discussed.

KEY WORDS: Trichobezoar - Trichophagy - Rapunzel syndrome - Trichotillomania.

Introduction

Trichobezoars are concretions of swallowed hair in the digestive tract.

Trichobezoars, unlike other bezoars, are not associated with alterations in gastrointestinal motility but with underlying psychiatric disorders, and most commonly present in adolescents and during the second decade of life. Rapunzel syndrome is an unusual and rare form of trichobezoar extending into the small intestine.

This condition is more common in women, especially adolescent girls (90%). Trichobezoars are associated with trichophagia as a result of pica – an eating disorder manifested by an appetite for nonnutritive substances and often associated with mental alteration – and coexistent psychiatric disturbances. The insidious development of the trichophytobezoar accounts for the delayed presentation and large size at the time of diagnosis (1).

The condition is usually associated with the mentally retarded or with young children and may be caused by a variety of conditions, including anxiety, depression, and family stress.

Here we describe a nine-year old girl with huge gastric trichobezoar causing poorly localized abdominal pain, nausea and 2 days of postprandial emesis.

Case report

A 9-year-old female child presented to the emergency room with a 1.5 week history of poorly localized abdominal pain, nausea and 2 days of postprandial emesis.

The mother commented on early satiety and chronically decreased appetite.

There were no changes in her bowel habits.

The child had no history of diarrhea, night sweat, cough, urinary symptoms, and allergy or drug ingestion.

She is in primary school with an average performance and she belongs to a middle social class family. The mother reported that the child was suffering several years with a too severe teacher. She was observed to ingest her own hair up to about 1 year ago.

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On physical examination, her weight and height are at 50th centile for age.

Abdominal examination revealed a hard, non-tender, non compressible and non mobile mass of approximately 20 cm x 10 cm in the epigastric region extending to the right hypochondrium. Other systemic examination was unremarkable. Her scalp showed no signs of alopecia.

Vital signs were normal. Laboratory work-up were normal. Her random blood sugar, renal values, liver function tests, and serum amylase were within the normal range.

Ultrasound of abdomen showed a well defined hyperechoic mass of 20 cm in diameter with acoustic shadow in the epigastric region (Fig. 1). Upper endoscopy disclosed the nature of the mass as a large hair ball occupying the whole stomach. The size of the mass precluded endoscopic removal (Fig. 2).

Therefore the patient underwent laparotomy (Fig. 3). An upper midline incision was performed.

The giant mass was found to be a trichobezoar with a tapering tail extending into the duodenum, and was a perfect cast of the stomach, pylorus and duodenal bulb (Fig. 4). The mass was removed through an anterior gastrotomy. The gastrotomy was closed in two layers and the abdomen was closed without drainage.

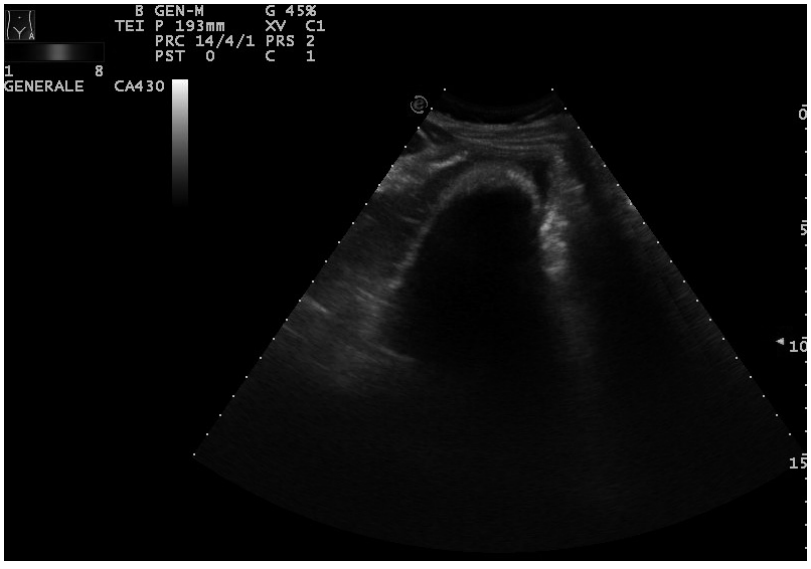

Fig. 1 - Ultrasound of tricobezoar in situ.

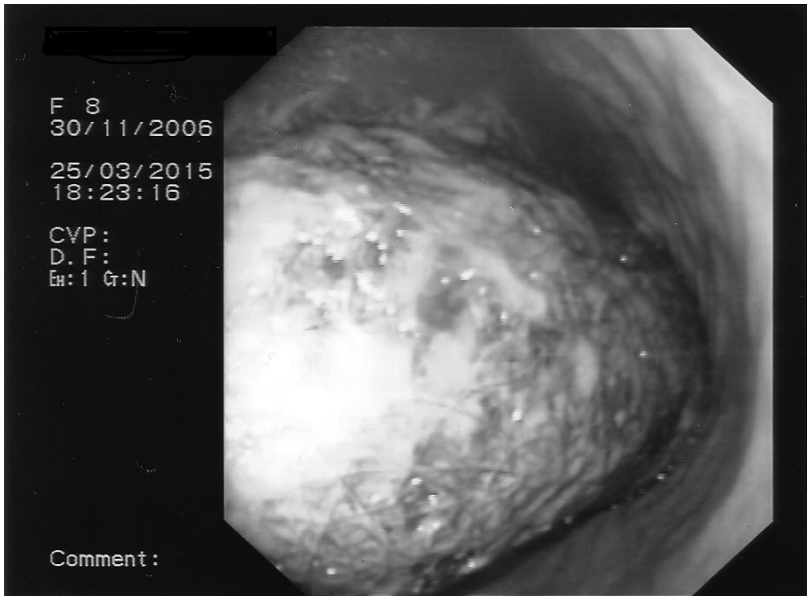

Fig. 2 - Tricobezoar during gastroscopy.

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Fig. 3 - Intraoperative image.

Fig. 4 - Tricobezoar after the extraction.

After surgical intervention, the patient recovered well, and her hospital stay was uneventful. The patient was discharged 5 days later

A psychiatric evaluation diagnosed an adjustment disorder. She was discharged for psychiatric follow-up. She also received a course of psychotherapeutic treatment to avoid recurrence.

Discussion

Bezoars are concretions of foreign material in the gastrointestinal tract, mainly the stomach. Trichobezoars composed of hair conglomerates form a smooth mass that peristatic contractions do not expulse, with a reported occurrence of 0.012%. Although the stomach is the most common location, bezoars have also been found in the duodenum, jejunum, ileum, colon, appendix and Meckel´s diverticulum (2).

Trichobezoars most commonly present in the second decade of life. They account for 12% of bezoars. Up to 90% of the all trichobezoars occur in girls younger than 20 years old. Males are rarely affected (3).

Rapunzel syndrome is a rare form of trichobezoar; some Authors define it as a gastric trichobezoar with a tail extending up to the ileocaecal junction; others de-

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scribe it as a simple trichobezoar with a long tail, which may extend up to the jejunum or beyond; and some others still define it as a bezoar of any size which causes intestinal obstruction (4).

Trichobezoars can become large and form as a cast of the stomach. They may enter into the proximal duodenum as occurred in our case, to present with symptoms of gastric outlet or partial intestinal obstruction.

Trichobezoars result from compulsive pulling out of hair (trichotillomania) and then swallowing the hair (trichophagia). Patients frequently have accompanying comorbid mood and anxiety disorders that can be caused by a variety of condition, including anxiety, depression, and family stress and require comprehensive psychiatric or psychologic evaluation for obsessive compulsive syndrome. It has been estimated that only 1% of patients with trichophagia develop a trichobezoar.

The cause why hair is collected in the stomach is not fully understood. Due to its indigestibility and resiliency, it becomes entrapped within the mucosal folds where it gets enmeshed, and acquires more hairs and then a larger size.

As more hair accumulates, peristalsis causes it to be enmeshed into a ball. The ball of hair becomes even more matted together and assumes the shape of the stomach, usually as a single solid mass.

The acid content of the stomach denatures the hair protein and give the bezoar its black color.

Clinical manifestations vary, depending on the location and size of the tricobezoar, from asymptomatic patients to acute abdomen.

The patient with a gastric trichobezoar usually presents with vague and nonspecific symptoms, including abdominal pain (70%), nausea and vomiting (64%), digestive bleeding (61%), epigastric discomfort, early satiety, dyspepsia, weight loss (38%), diarrhea or constipation (32%) and halitosis. A bezoar may also lead to mechanical obstruction, gastric perforation, gastrointestinal bleeding, anaemia and ulcer formation.

Patients may occasionally have iron deficiency anemia, hypoproteinemia or steatorrhea caused by associated chronic gastritis, protein losing enteropathy and pancreatitis.

Less common manifestations and complications include acute appendicitis and intussusception. Occasionally bezoars are giant-sized, as in our case and may mimic an intraabdominal mass clinically. Anaemia and hypoalbuminaemia associated with chronic gastritis usually go unnoticed until the case is brought to light by the onset of severe complications such as hemorrhage, obstruction or perforation. Perforation and peritonitis are largely responsible for an attendant mortality of about 30%. Physical examination may demonstrate patchy baldness and a firm mass in the right upper quadrant. Clinical suspicion should be high for trichobezoar in women with psychiatric problems presenting with abdominal pain, while diagnosis in a healthy patient requires a high index of suspicion, as it can present with nonspecific symptomatology (5-7).

If suspected, trichobezoars can be diagnosed with imaging and endoscopic techniques. Imaging includes study with contrast medium, ultrasonography, CT scan. CT scan has a high accuracy rate, the accuracy of US in such cases is not so high.

Endoscopy provides a diagnosis and it is a possible mean of therapeutic disruption and removal of the mass (8).

The management and treatment of bezoar need to encompass removal of the mass and prevention of recurrence by addressing the underlying physical or emotional cause. Trichobezoars are resistant to enzymatic dissolution. Depending on its consistency, size and location, bezoar removal may occur via endoscopy or surgery.

Operative removal is usually indicated for large trichobezoar. Gastroscopic removal carries some risks such as perforation or intestinal obstruction.

Various other methods like extracorporeal shock wave lithotripsy, intragastric administration of enzymes (pancreatic lipase, cellulose), and medications (metoclopramide, acetylcysteine) demonstrate varying success. Few recurrences are reported after the initial removal of bezoars.

Nevertheless, treatment failure or large bezoars require surgical treatment.

Surgery is also indicated when a very large or solid bezoar causes perforation or hemorrhage, or in case of Rapunzel syndrome, when there is significant extension of the bezoar.

Surgical removal is accomplished by gastrotomy or enterotomy. Traditionally, a gastric trichobezoar was removed by gastrotomy through an upper midline laparotomy. Since the advent of minimally invasive surgery, surgeons now use laparoscopic techniques for small to moderate-size bezoars.

Laparoscopic and minilaparotomy approaches are widely recognized minimally invasive operations, with laparoscopy being the preferred procedure because of its decreased invasiveness. In cases of giant trichobezoar, the laparoscopic approach could be problematic. Minilaparotomy could also be an alternative to laparoscopy in cases of moderately large trichobezoars in hospitals with moderate experience in advanced laparoscopic surgery (9, 10).

The patient's long-term prognosis is excellent if behavioral therapy is used to control trichophagia, and psychological/psychiatric follow-up is maintained.

Psychological assessment and support is very important in the evaluation of a child with bezoar. The-

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rapy of hair pulling includes behavior modification and family or individual counseling. Until the underlying causes for the condition are cleared up, the only solution is to keep the hair too short for pulling.

Conclusion

A diagnosis of gastric bezoar should be suspected in any child with symptoms of gastric outlet obstruction, and surgical removal is usually indicated for large trichobezoar.

Trichobezoar has to be considered in the differential diagnosis of abdominal pain and a non-tender abdominal mass even in young children, and a history of pica should always be obtained. Small trichobezoars may be extracted by endoscopic fragmentation, enzymatic therapy or combinations of these approaches. Bezoars like Rapunzel syndrome, on the other hand, need surgical removal.

Counseling by a psychiatrist is an important part of management to prevent recurrence. In young girls with psychiatric history who present with abdominal pain, vomiting or abdominal mass, bezoars/Rapunzel syndrome should be considered in the differential diagnosis.

To avoid recurrences, each patient should have a psychiatric evaluiation and follow-up.

References

1. Zamir D, Goldblum C, Linova L. Phytobezoars and trichobezoars: A 10-year experience. J Clin Gastroenterol. 2004;38:873-876.
2. Meier CM, Furtwaengler R. Trichophagia: Rapunzel syndrome in a 7-year-old girl. J Pediatr. 2015 Feb;166(2):497.
3. Lynch KA, Feola PG, Guanther E. Gastric trichobezoar: an important cause of abdominal pain presenting to the paediatric emergency department. Pediatr Emerg Care. 2003;19:343-7.
4. Kaspar A, Deeg KH, Schmidt K, Meister R. Rapunzel syndrome, an rare form of intestinal trichobezoars. Klin Padiatr. 1999;211:420-422.
5. Athanasiou A, Michalinos A, Moris D, Spartalis E, Dimitrokallis N, Kaminiotis V, Oikonomou D, Griniatsos J, Felekouras E. Rapunzel syndrome: a rare presentation with giant gastric ulcer. Case Rep Med. 2014;2014:267319.
6. Islek A, Sayar E, Yılmaz A, Boneval C, Artan R. A rare outcome of iron deficiency and pica: Rapunzel syndrome in a 5-year-old child iron deficiency and pica. Turk J Gastroenterol. 2014 Feb;25(1):100-2.
7. Frey A, Mckee M, King R, Martín A. Hair apparent: Rapunzel syndrome. Am J Psychiatry. 2005;162:242-8.
8. Belsky J, Whitcomb V, Zimmerman E, Stankovic C. Rapunzel syndrome: diagnosis via radiograph and history. Pediatr Emerg Care. 2014 May;30(5):352-3.
9. Shulutko AM, Agadzhanov VG, Kazaryan AM. Minilaparotomy removal of giant gastric trichobezoar in a female teenager. Medscape J Med. 2008;10:220-3.
10. Vepakomma D, Alladi A. Complete laparoscopic removal of a gastric trichobezoar. J Minim Access Surg. 2014 Jul;10(3):154-6.

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