Rapunzel syndrome: A rare cause of intestinal obstruction. A case report

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Case Report

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BACKGROUND. Rapunzel syndrome is a rare form of gastric trichobezoar with post-pyloric extension associated with intestinal obstruction. It is a suspected diagnosis in female and young patients with a palpable epigastric tumor associated with intestinal obstruction.
We report a case of a 16-year-old female patient with an 8-year history of trichotillomania and trichophagia, who presented with intestinal occlusion and tomographic confirmation of Rapunzel's syndrome with gastric bezoar fragmentation and migration of the fragment to the distal ileum; managed with gastrotomy and enterotomy achieving satisfactory extraction of both trichobezoars.

KEY WORDS: Rapunzel syndrome, trichobezoar, intestinal obstruction.

Introduction

Bezoars are concretions of indigestible material within the digestive tract, made up of materials such as: plant fibers (phytobezoars), drugs (pharmacobezoars), milk conglomerates (lactobezoars, common in premature newborns) or, as in this case, hair (trichobezoars).[1,2,3]

The term bezoar derives from the words "badzher" (Arabic), "panzheir" (Turkish), "padzhar" (Persian) and "beluzzar" (Hebrew) translated as: "antidote", since formerly bezoars extracted from animals were used as a remedy for various diseases.[3,4] In 1779 Baudamant published the first case of bezoars in humans, and Schonbern in 1883 performed the first surgical intervention to remove a bezoar.[4]

This pathology is related to allotryophagia or pica, which consists of the inclination to ingest certain substances that cannot be assimilated by the body.[8] Because the ingested material is resistant to the action of hydrochloric acid, with chronic consumption bezoars develop. Other associated factors are inadequate chewing, a diet rich in fiber, previous gastric surgery, alterations in pyloric function,[6] diabetes mellitus, psychomotor retardation,[18] and psychiatric conditions such as trichotillomania associated with trichophagia.[2,3]

Trichotillomania was described in 1889 by the French psychiatrist Hallopeau,[11] composed of "tricho" (hair), "linden" (pulling) and "mania" (abnormal affect); It is currently included in the DSM V as an obsessive-compulsive disorder defined as: "hair-pulling disorder", with an incidence that ranges from 0.6-1.6%, the onset being frequent between 5-13 years of age.[9] On the other hand, Trichophagia (associated with trichotillomania in 30%) comes from "tricho" (hair) and "phages" (eating) consists of the act of ingesting hair, without necessarily being the patient, so not all patients with Trichotillomania suffer from trichophagia, not all patients with trichophagia develop trichobezoars and trichobezoars are not necessarily associated with areas of alopecia, and there may be alternative sources for trichophagia such as animals, dolls, brushes and carpets.[10, 11, 39]

In the genesis of trichobezoars (incidence of (0.06-4%)[13] the ingested hair remains attached to the gastric folds due to its poor surface tension,[6] with the consequent accumulation of hair, mucus and food fragments. The size is variable depending on the evolution time finding reports of bezoars up to 3.2kg. Trichobezoar is invariably black regardless of the color of ingested hair due to denaturation of proteins by hydrochloric acid, shiny due to retained mucus, and fetid due to the decomposition of food waste retained.[5]

Within the spectrum of trichobezoars there is a potentially fatal entity called "Rapunzel syndrome", a term coined by Vaughan in 1968.[3,4,6,12] Rapunzel syndrome is a rare entity with few reports, for which there is no formal consensus on the diagnostic criteria. Naik et al in 2007 proposed the diagnostic triad of: 1) trichobezoar with tail, 2) extension of the tail at least to the jejunum and 3) obstructive symptoms.[6] However, most authors agree that it is a gastric trichobezoar with postpyloric extension that presents in the form of intestinal obstruction.[17] (without being essential its arrival in the jejunum), with the risk of fragmentation and displacement to the small intestine and this is the most accepted definition of Rapunzel syndrome.[2,13]

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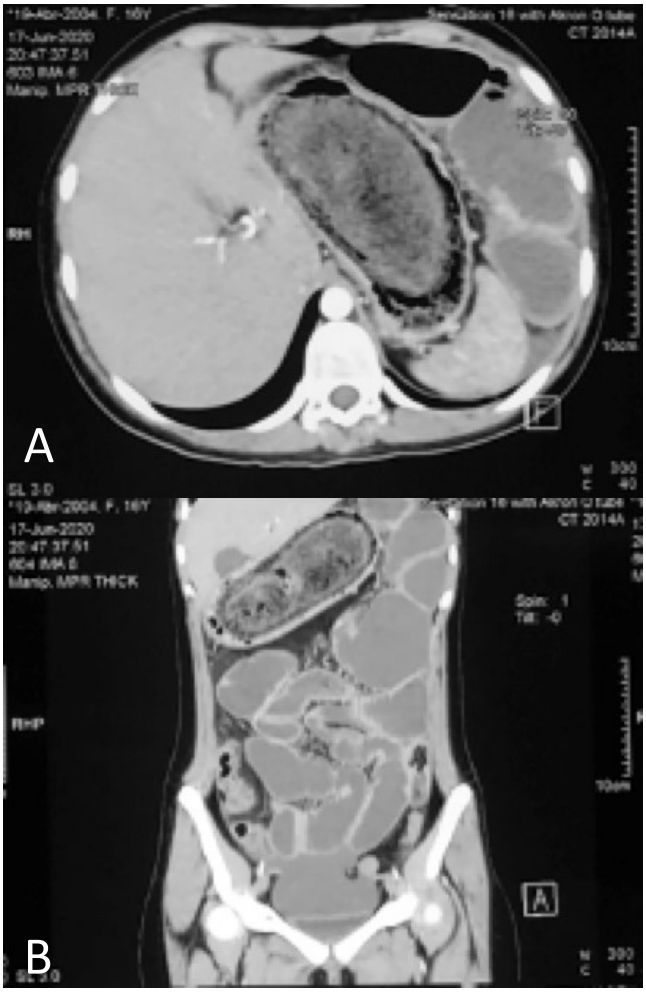

Figure 1. A. Trichobezoar occupying gastric chamber and intestinal loop dilation. B. Trichobezoar shaping gastric chamber.

Trichobezoars predominate in the female sex ~ 90%[3,4] of the cases and in those under 30 years of age in 80% of the cases, with a maximum incidence of 10-20 years.

The clinical picture associated with this entity depends on the size of the bezoar and the appearance or not of complications, being able to find early satiety, nausea, postprandial emesis (65%),[3] hematemesis (61%),[5] epigastric pain (70%),[3] palpable tumor (70%),[5] weight loss (38%),[4] malnutrition and iron deficiency anemia.[3,20] Among the complications are intestinal occlusion,[3] gastric ulcer (caused by constant abrasion of the mucosa, incidence of 10%),[3] bleeding, perforation (10%), invagination (1.8%) and even appendicitis,[19] cholangitis (<0.9%) and pancreatitis.[2,3,6,15]

The diagnosis is established with clinical suspicion, upper gastrointestinal series, endoscopy or tomography, (of choice to determine the extension or fragmentation of the bezoar).[2,3,15]

In 1959, Don et al proposed the enzymatic treatment for small bezoars that consists of the intake of papain found in the carnic macerator "Adolph's Meat Tenderizer" at a dose of 15cc in 150-300cc of water every 6 to 8 hours (success 37 %).[2,4] Currently there are cellulose preparations (83% success)[4] and acetylcysteine (50% success);[4] After these therapies, gastric lavage or endoscopic fragmentation (85% success)[4] is necessary, which usually requires reoperation in 9% of cases due to recurrence secondary to residual bezoars.[3] If there is a failure in this treatment, it is a large bezoar or there are data of intestinal occlusion, the need for surgical removal of the bezoar with intestinal exploration to rule out additional bezoars due to fragmentation is indisputable.[1,2,5]

Psychiatric management of trichophagia is essential since these bezoars tend to recur in up to 20% of cases.[2,5]

Case report

The case of a 16-year-old female patient with a history of trichotillomania and trichophagia of 8 years of evolution is presented. He began suffering five days prior to his arrival at our unit when he presented epigastric abdominal pain, colic type, intensity 10/10 on a visual analog pain scale, associated with postprandial emesis on more than 10 occasions and constipation, adding one day prior to his arrival. In this hospital, abdominal distension, obstipation and intolerance to the oral route.

Upon her arrival, she is assessed by the general surgery service who finds the patient underhydrated, with mucotegumentary paleness and large areas of alopecia in the occipital region; On abdominal examination, distension, hyperesthesia and hyperbaralgesia in the epigastrium, generalized pain, firm palpable tumor in deep planes of approximately 30x15cm, epigastric dullness and bloating in the rest of the abdominal regions.

Routine laboratory studies are performed, finding leukocytosis associated with neutrophilia, hypoalbuminemia, and mild anemia.

Under the diagnosis of intestinal occlusion and considering the history of trichotillomania with trichophagia, Rapunzel syndrome is considered as the main diagnostic suspicion, an abdominal tomography was performed confirming said diagnosis (**Figures 1A and 1B**).

Upon confirmation, the patient was transferred to an exploratory laparotomy, finding dilation of the intestinal loops (**Figure 2A**), as well as the entire gastric chamber occupied by a trichobezoar with extension to the second portion of the duodenum.

An exploration of the rest of the digestive tract was performed, finding a second bezoar in the ileum at 90cm from the ileocecal valve with dilatation of the intestinal loops prior to the bezoar.

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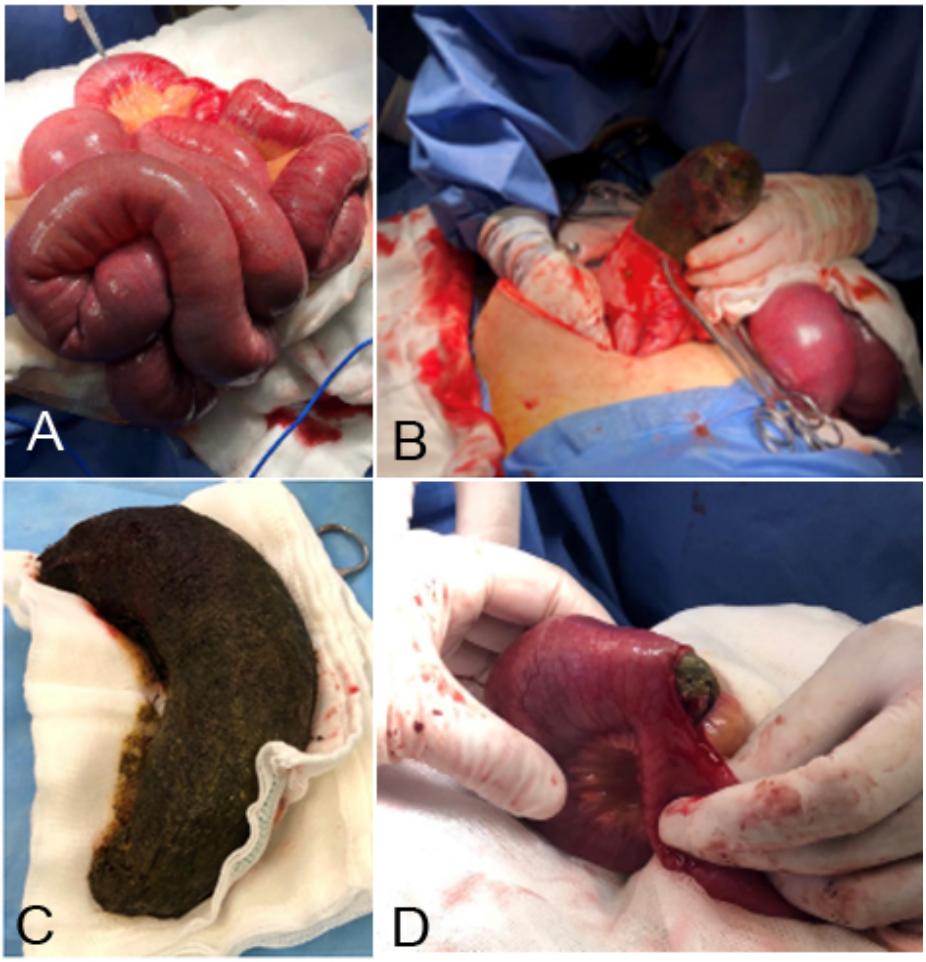

Figure 2. A. Dilation of intestinal loops. B. Gastrotomy and extraction of gastric trichobezoar. C. Gastric trichobezoar. D. Enterotomy and trichobezoar in the distal ileum.

The gastric bezoar is extracted by gastrotomy on the anterior side of the stomach, following the path of its major axis, obtaining a 30x5x15cm trichobezoar molded to the gastric chamber; Gastric wall closure is performed in two planes (**Figure 2B and 2C**).

An enterotomy was performed in the distal ileum, finding a second 10x4cm bezoar, closing it in two planes (**Figure 2D**).

The patient underwent a satisfactory evolution during her hospitalization and was discharged with reference to the Pediatric Psychiatry service.

Discussion

Trichobezoars are usually found in the stomach, but when they cross the pylorus and involve the duodenum, jejunum, ileum, ileocecal valve or even the colon, conditioning a picture of intestinal obstruction, they are called "Rapunzel syndrome". In our case, the trichobezoar compromised the stomach and duodenum up to its descending portion with a second trichobezoar in the distal ileum itself, which conditioned the patient's occlusive condition.

As mentioned, human hair is resistant to digestion and intestinal peristalsis, so the continuous ingestion of hair for a long period of time leads to its accumulation in the gastric chamber. In this case, the patient presented trichophagia for a period of approximately 8 years without receiving specialized management.

Usually the patient denies or hides information regarding trichotillomania. Trichophagia is a perversion of appetite determined by affective disturbances, social isolation and anxiety, conditions referred to by the mother in the present case. The depressive component and the conflictive family environment triggered by a stressful situation were decisive for the genesis of the trichobezoar.

Regarding surgical management, the gastrotomy must be longitudinal to the long axis of the stomach with sufficient width to cause the extraction of the bezoar and avoid tears of the incision with subsequent closure in two planes.

It is pertinent to highlight the importance of psychiatric management to avoid recurrences.

Conclusion

Rapunzel syndrome is a condition commonly associated with trichotillomania and trichophagia in young women, the diagnosis of which requires a thorough questioning, a complete clinical examination and a high diagnostic suspicion coupled with laboratory studies for confirmation. Surgical treatment

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is indicated in the context of giant trichobezoars or not accessible to endoscopic management. A multidisciplinary management is necessary to avoid complications and recurrence, being essential management by psychiatry.

Conflicts of interests

There was no conflict of interest during the study, and it was not funded by any organization.

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