A Giant Trichobezoar Causing Rapunzel Syndrome in a 12-year-old Female

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ABSTRACT

Bezoar is a tightly packed collection of undigested material that is unable to exit the stomach. Most bezoars are of indigestible organic matter such as hair–trichobezoars; or vegetable and fruit–phytobezoars; or a combination of both. Trichobezoars commonly occur in patients with psychiatric disturbances who chew and swallow their own hair. In very rare cases, the Rapunzel syndrome hair extends through the pylorus into the small bowel causing symptom and sign of partial or complete gastric outlet obstruction. A case report of trichobezoar in the stomach causing Rapunzel syndrome in a 12-year-old female is reported.

Key words: Rapunzel, syndrome, female, giant, trichobezoar

INTRODUCTION

A bezoar is an agglomeration of food or foreign material in the intestinal tract. It can be classified according to the primary constituent as trichobezoar or hairball, phytobezoar (food particles), trichphytobezoar (mixed), pharmacobezoar, lactobezoar, mycobezoar (fungal agglomerations) or pseudobezoar. Trichobezoars are the commonest type of bezoars. Occasionally, trichobezoars have a tail that extends to the cardia, pylorus, and duodenum, or even further to the jejunum and ileum. When the entire small intestine is involved, the disorder is called Rapunzel syndrome.

CASE REPORT

A 12-year-old female was referred to our surgical clinic with a history of abdominal pain, distension, weight loss, and attacks of vomiting of 4 months duration. Her personal history revealed nothing significant. Abdominal palpation revealed a well-defined mass occupying the upper half of the abdomen. The mass was not tender and was firm in consistency. The computed tomography (CT) scan revealed a well-circumscribed lesion in the region of the stomach that comprised of concentric whorls of different densities with pockets of air enmeshed within it. Oral contrast filled the more peripheral interstices of the lesion with a thin band of contrast circumscribing the lesion [Figure 1]. Upper gastrointestinal endoscopy revealed a trichobezoar occupying almost the whole gastric cavity. Removal of the trichobezoar endoscopically failed as it was possible to pull only few fibers of this huge ball of hair.

The patient underwent surgery, and through upper midline incision gastroscopy was done. A huge trichobezoar was identified which took the shape of the stomach [Figure 2]. The whole trichobezoar weighing
about 2 kg was removed. There was a long tail of hair extending through the pylorus into the proximal jejunum [Figures 3 and 4]. By this feature the diagnosis was clear of a Rapunzel syndrome. The opening in the gastric wall was closed with continuous Vicryl 3-0. Postoperatively, the patient accepted continuous ingestion of her own hair since 2 years. The patient had an uneventful postoperative course and was discharged after 8 days. The patient was also referred to psychiatric follow-up. Sertraline hydrochloride, starting from 50 mg/day to 100 mg/day, was given to the patient to provide additional help in reducing her hair-pulling urges. After several months of weekly psychotherapy and pharmacotherapy, the patient is presently reported to have no hair-pulling events.

DISCUSSION

The most commonly encountered bezoar is the “trichobezoar”. It is almost exclusively seen in young females, often associated with psychiatric problems. In our case, the presentation is in a very young age with hair extending down to the proximal jejunum, causing symptoms, which could mimic gastrointestinal infections and infestation especially in endemic areas. It is postulated that hair strands too slippery to be propelled are initially retained in the mucosal folds of the stomach and become enmeshed over a period of time. Trichobezoars are usually black from denaturation of protein by acid, glistening from retained mucus and foul smelling from degradation of food residue trapped within it.[1]

The patient generally presents with epigastric discomfort, pain, nausea, vomiting, satiety exacerbated at meal times, or complete gastric outlet obstruction. Contiguous extension of a trichobezoar into the small bowel can lead to the ‘Rapunzel syndrome’. This syndrome is named after a tale written in 1812 by the Brothers Grimm about a young maiden, Rapunzel, with long hair who lowered her hair to the ground from a castle, which was a prison tower to permit her young prince to climb up to her window and rescue her. This syndrome

Figure 1: CT scan demonstrates a mixed density mass with a whirled configuration containing multiple small pockets of air. Oral contrast circumscribes the lesion

Figure 2: Trichobezoar occupying whole stomach

Figure 3: Trichobezoar being delivered

Figure 4: The giant trichobezoar
was originally described by Vaughan et al. in 1968.\[^{1}\] The commonly accepted definition is that of a gastric trichobezoar with a tail extending to the jejunum, ileum, or the ileocecal junction. The complications of the Rapunzel syndrome ranges from attacks of incomplete pyloric obstruction to complete obstruction of the bowel to perforation to peritonitis and mortality.\[^{2,3}\]

Trichobezoars with small bowel extensions may produce other complications, namely bleeding, perforation, protein losing enteropathies,\[^{4,5}\] steatorrhea, pancreatitis, appendicitis, and intussusceptions.\[^{6}\] Diagnosis of trichobezoars rest on the clinical evidence of long standing trichophagy, abdominal mass and radiological investigations or CT scan and abdominal ultrasound but obtaining pieces of matted hair through endoscope is pathognomonic. The characteristic appearance on CT is of an inhomogenous non-enhancing mass within the lumen of the stomach/bowel. Oral contrast circumscribes the mass and may fill the interstices near the surface. Surgical removal at laparotomy or laparoscopically is the treatment of choice. If small, they may be removed endoscopically. Biopsy devices, water jets, bezotomes, and LASER devices may be used to fragment larger bezoars and lavaged out of the stomach.

Psychiatric follow-up is important although no conclusion has been reached with respect to whether the use of medication makes any difference in the progression of this condition.\[^{7}\] This follow-up care should be extended to family members, who should be vigilant with patients, since recurrences of the problem have been described.\[^{8}\] No clear evidence-based practice guidelines for the treatment of patients with (trichotillomania) TTM are available, and more research is needed. Behavioral techniques and selective serotonin reuptake inhibitors (SSRIs) are the most commonly used treatment modalities, having the most evidence for efficacy. The behavioral approaches, including habit-reversal therapy, have been shown to be effective.\[^{9}\] In habit-reversal therapy, patients learn to be aware of the times, cues, and situations in which they pull their hair. They practice movements such as those in knitting, crochet, and needlepoint that redirect their urges to pull their hair. Thus, they learn to “substitute a different and more adaptive behavior”\[^{9}\] and receive social approval for efforts to interrupt the hair pulling.\[^{10}\] Other approaches such as cognitive-behavioral treatment, negative practice, and variations of these interventions have been tried.\[^{11}\]

The SSRIs are the pharmacologic agents used most frequently in the treatment of patients with TTM. Other psychotropic agents that have been used for treating patients with TTM include the tricyclic antidepressants, the antipsychotic medications, and mood-stabilizing (anticonvulsant) medications. Clomipramine hydrochloride and fluoxetine hydrochloride have been used successfully.\[^{12}\] The need for adequate follow-up should be emphasized to avoid recurrences, although these are rare since the trauma of surgery may prevent the patient from provoking another episode.

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