Review

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Rapunzel Syndrome Reviewed and Redefined

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Key Words
Bezoar · Rapunzel syndrome · Trichobezoar · Trichotillomania · Intestinal obstruction

Abstract
Rapunzel syndrome is a rare presentation of a trichobezoar, with 24 cases having been reported in literature so far. This syndrome has been reported in 23 female and 1 male patient with a mean age of 10.8 years. The patients commonly present with abdominal pain, nausea, vomiting and signs of obstruction. The distal end of the bezoar may be in the jejunum, ileum or the colon. We evaluate the various cases reported and discuss the various modes of presentation of this uncommon syndrome, and also present a series of 3 cases, all females, aged 16, 18 and 21 years, and had a trichobezoar in the stomach with a tail extending into the ileum.

Introduction

Trichotillomania, i.e., the irresistible desire to pull out ones own hair, was first described in 1889 by Hallopeau, while the first case of a trichobezoar was reported a century earlier by Baudamant in 1779 [1]. Trichobezoars are usually without symptom until they reach a large size. Rapunzel syndrome is a rare form of trichobezoar, Vaughan et al. [2] first described this syndrome in 1968 in 2 patients. Though Vaughan et al. [2] did not give any strict definitions of the syndrome, both their patients had a trichobezoar with a tail-like extension and symptoms of obstruction. We report 3 cases with similar features.

Case Reports

Case 1

A 16-year-old female presented with a history of anorexia and occasional vomiting. Clinical examination revealed a mobile lump in the abdomen in the shape of a dilated stomach. She had a history of recurrent admissions for subacute intestinal obstruction. Her blood picture revealed hypochromic microcytic anemia. Barium contrast showed an intragastric mass with barium in the honeycomb interstices (fig. 1). The barium contrast outlined the margins of the mass and penetrated the interstices in a lace-like 'mottled' pattern. There was no connection to the stomach wall and the mass did not arise from it.

On laparotomy a large gastric trichobezoar was present. It had a tapering tail, which extended into the proximal ileum (tail length 116 cm; fig. 2a). The mass was removed by gastrostomy. The patient made an uneventful recovery. On retrospective questioning the patient denied trichophagia but her parents remarked that they had often seen her putting hairballs in her mouth.

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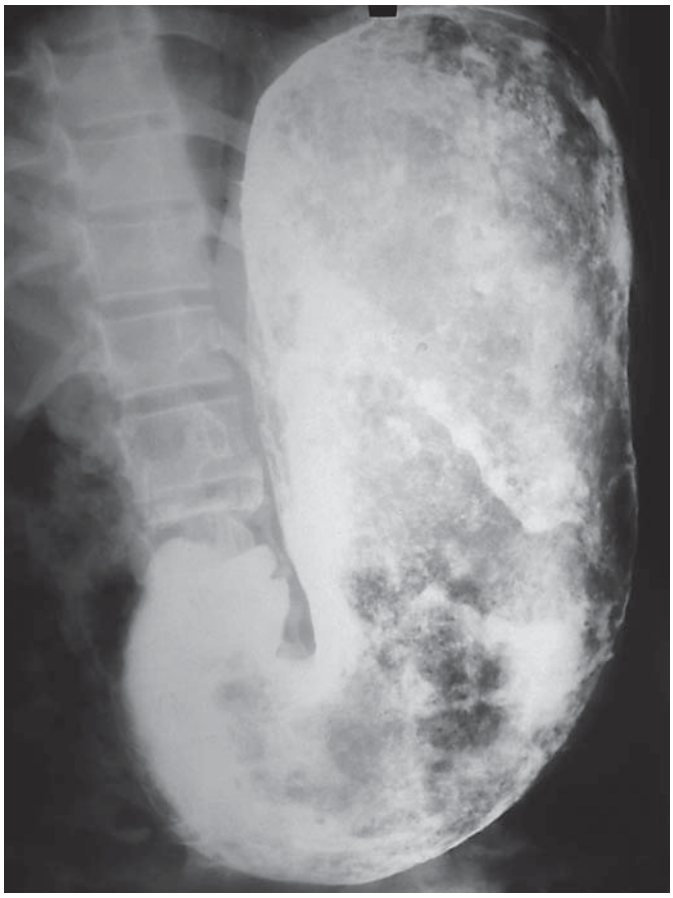

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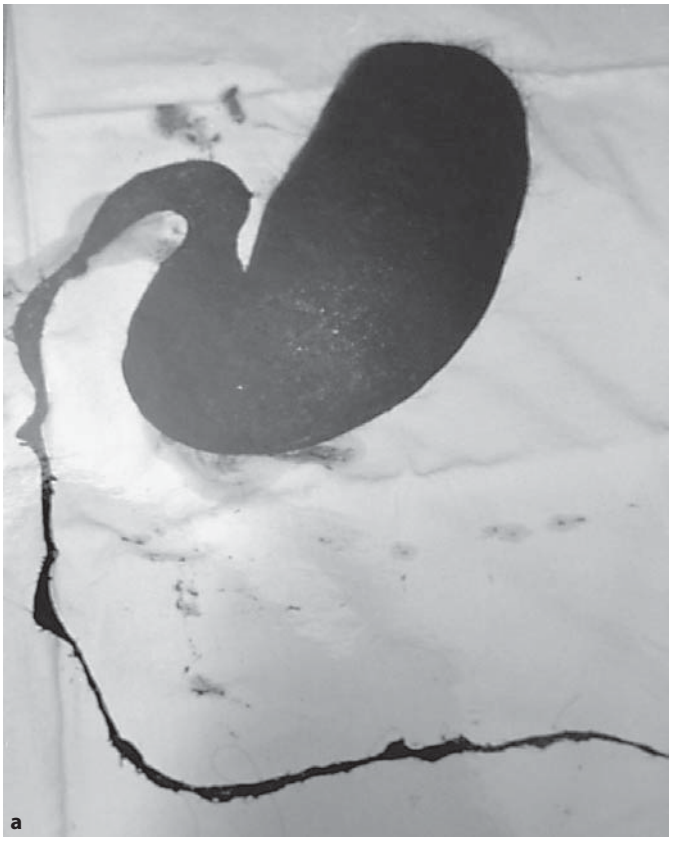

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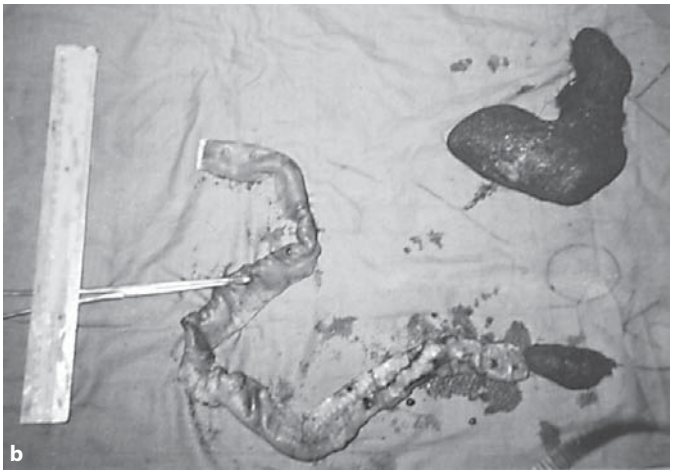

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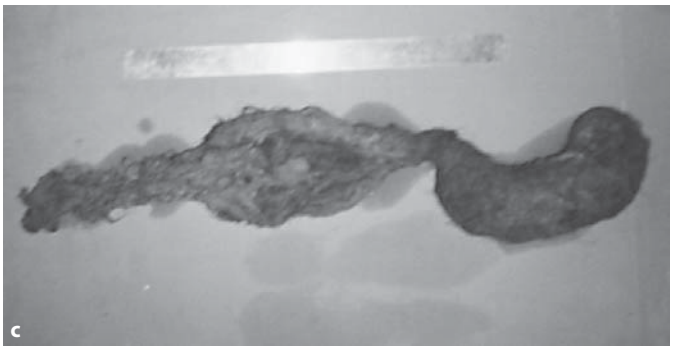

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Fig. 1. Barium contrast study showing an intragastric trichobezoar.
Fig. 2. Post-surgical specimens of case 1 (**a**), case 2 (**b**), and case 3 (**c**).

Case 2

A 18-year-old female presented with complaints of abdominal distension, pain in the abdomen and vomiting of 4 days duration. On examination she had tachycardia and abdominal distension with exaggerated bowel sounds. Plain radiograph of the abdomen showed multiple air fluid levels with dilated small intestine loops. Conservative management with fluids and nasogastric aspiration for 24 h failed to improve her clinical condition. She was subsequently explored and a trichobezoar was found in the antrum extending into duodenum and proximal jejunum. This was removed by gastrostomy. The distal small bowel had another trichobezoar which was the cause of the obstruction and was removed by a separate enterotomy (fig. 2b). Part of the distal small bowel was gangrenous with multiple impending perforations. It was resected and an end-to-end ileoileal anastomosis was performed. The patient recovered uneventfully and was discharged on the 10th postoperative day. On retrospective questioning she admitted having trichophagia and trichotillomania.

Table 1. Rapunzel syndrome: cases reported and their characteristics

	Author	Year	Age	Sex	Presentation	Extent	Outcome
1	Vaughan et al. [2]	1968	13	F	Peritonitis	Jejunum to ileocecal valve	Alive
2	Vaughan et al. [2]	1968	15	F	Obstruction	Jejunum to colon	Alive
3	Deslypere et al. [5]	1982	14	F	Nausea + vomiting	Stomach to transverse colon	Died
4	Buyukunal et al. [10]	1982	5	F	Sepsis	Details not available	Died
5	Wolfson et al. [11]	1987	5	F	Volvolus	Stomach to cecum	Alive
6	Hassan and Panesar [12]	1989	5	F	Weight loss	Stomach to ileocecal valve	Alive
7	Balik et al. [9]	1982	15	F	Abdominal pain, bilious vomiting	Stomach to jejunum (53 cm tail)	Alive
8	Duncan et al. [13]	1993	5	F	Intussusception	Stomach to ileocecal valve	Alive
9	Uroz Tristan et al. [14]	1996	8	F	vomiting + asthenia	Stomach to jejunum	
10	Pul and Pul [15]	1996	12	F	Vomiting distension anorexia	Stomach to ileocecal valve	Alive
11	Seker et al. [16]	1996	6	F	Obstruction	Stomach to cecum	Alive
12	Senapati and Subramanium [17]	1997	8	F	Pain + hematemesis	Stomach to jejunum (20 cm tail)	Alive
13	Dalshaug et al. [18]	1999	7	F	Intussusception + obstruction	Mid jejunum to transverse colon	Alive
14	Singla et al. [19]	1999	9	F	Weight loss + abdominal pain	Stomach to ileum	Alive
15	Kasparet al. [20]	1999	12	F	Obstruction	Stomach to ileum	Alive
16	Faria et al. [1]	2000	7	F	Peritonitis	Stomach to ? (not detailed)	
17	Hirugade et al. [21]	2001	6	M	Colic + vomiting	Stomach to terminal ileum (195 cm tail)	Alive
18	Couper [22]	2001	4	F	Epigastric pain	Stomach to jejunum	Alive
19	Curosio Vilchez et al. [23]	2002	22	F	Details not available	Details not available	
20	Klipfel et al. [24]	2003	14	F	Vomiting gastric emphysema	Stomach separate in terminal ileum	Alive
21	Memon et al. [25]	2003	10 and 12	F	Nausea + colic	Stomach to proximal ileum	Alive
22	Gockel et al. [26]	2003	4	F	Abdominal pain	Details not available	Alive
23	Deevaguntla et al. [27]	2004	12	F	Pain, vomiting	Stomach to distal small bowel	Alive
24	Eryilmaz et al. [28]	2004	19	F	Pain, vomiting	Stomach to ? (not detailed)	Alive
25	Naik et al. [present study]	2006	16	F	Pain, SAIO	Stomach to ileum	Alive
26	Naik et al. [present study]	2006	18	F	Peritonitis	Stomach to mid ileum	Alive
27	Naik et al. [present study]	2006	21	F	Peritonitis	Stomach to terminal ileum	Alive

SAIO = Subacute intestinal obstruction.

Case 3

A 21-year-old female presented with repeated episodes of pain and distension of the abdomen, and vomiting of 6 days duration. On examination her abdomen was distended and bowel sounds were exaggerated. Plain radiograph of the abdomen showed multiple fluid levels and dilated small intestinal loops suggestive of small gut obstruction. She was managed conservatively and improved. Enteroclysis was advised but the patient was lost to follow-up. She presented 2 months later with peritonitis and the abdominal X-ray showed free gas under the diaphragm. On exploration there was contamination of the peritoneum with ileal content. Intussusception of the small gut was present at multiple places. A trichobezoar, which extended through the whole length of the small gut, was seen in the stomach. The length of the hair was less than the length of the small bowel and hence it was causing a pursestring-like effect. Peristaltic tightening of the strong, wire-like, twisted hairs of the 'tail' of the bezoar had caused contraction and compression of the mesenteric aspect of the wall of small bowel.

Gastrostomy was performed along with multiple enterotomies and the entire mass was removed (fig. 2c). The perforated bowel was resected and an ileostomy was made about 0.9 m proximal to the ileocecal junction. She had an uneventful postoperative course and was discharged 32 days after surgery. She admitted having trichophagia but denied trichotillomania.

Discussion

Trichobezoars are commonly found in young females usually with an underlying psychiatric disorder [3]. Formation of trichobezoars occurs when the hair strands are

Table 2. Rapunzel syndrome: summary of cases reported and their characteristics

Total patients	27
Demographics	
Mean age, years	10.8
Range	4–22
Sex	
Male	1
Female	26
Presenting symptoms	
Abdominal pain, %	37.0
Peritonitis, %	18.5
Nausea and vomiting, %	33.3
Obstruction, %	25.9
Weight loss, %	7.4
Intussusception, %	7.4
Bezoar length	
Stomach to jejunum, %	14.4
Stomach to ileum, %	25.9
Stomach to colon, %	25.9
Jejunum to colon, %	11.6
Unknown, %	22.2

retained in the folds of the gastric mucosa because their slippery surface prevents propulsion by peristalsis. As more hair is added, peristalsis causes it to be enmeshed until a ball, too large to leave the stomach, forms causing gastric atony due to its large size. This large quantity of hair becomes matted together and assumes the shape of the stomach, usually as a single mass [4, 5].

The increased incidence of these casts forming in the stomach is probably because of the initial hold up by the pylorus, and the churning action of the stomach which helps entangle new hair into the already formed casts.

The mucus covering the bezoar gives it a glistening shiny surface. Decomposition and fermentation of fats in the interstices gives it a putrid smell [6]. The acidic contents of the stomach denature the hair protein giving it its black color regardless of the original color of the hair [7, 8].

Rapunzel Syndrome

Rapunzel syndrome is a rare form of trichobezoar. It is named after a charming tale written in 1812 by the Brothers Grimm about a young maiden, Rapunzel, with long tresses [2] who lowered her hair to the ground from high in her prison tower to permit her young prince to climb up to her window and rescue her.

This syndrome was originally described by Vaughan et al. [2] in 1968. Since then 27 cases have been reported in the literature, with variable clinical features (table 1). Various criteria have been used by different clinicians to report their cases as Rapunzel syndrome. Some have defined it as a gastric trichobezoar with a tail extending up to the ileocecal junction [20]; others have said it is simply a trichobezoar with a long tail, which may extend to the jejunum, ileum or the ileocecal junction, and still others have defined it as a trichobezoar of any size which presents in the form of an intestinal obstruction [1]. The cases reported to date and their differing clinical and anatomical characteristics are summarized in table 2.

The identical features which would thus qualify all these cases to be a Rapunzel syndrome are: (1) a trichobezoar with a tail; (2) extension of the tail at least to the jejunum, and (3) symptoms suggestive of obstruction.

All the cases reported in the literature are females, except one. This may be attributed to the traditional long hair of females which makes them more prone to entanglement and hence formation of trichobezoar casts. The only male was reported by Hirugade et al. [21]. This patient, however, ate the hair of his sisters.

Most of the cases have been reported in countries where women traditionally have long hair. The most common presenting features are abdominal pain (37%), nausea and vomiting (33.3%), obstruction (25.9%), and peritonitis (18.3%). Uncommonly though, patients have also presented with weight loss (7.4%), anorexia, hematemesis and intussusception (7.4%; table 2). Most of the patients deny any history of trichotillomania or trichophagia, even when specifically asked. A thorough psychiatric evaluation and management are hence advocated. Conservatively managed cases have had a poor outcome and hence surgical removal is advised whenever preoperative diagnosis is made. However, most cases are diagnosed at laparotomy. Surgical removal is advocated by gastrostomy and enterotomies when required. Three recurrences have been reported [13, 25, 28]. In 2 cases, the first episode was as a simple trichobezoar and the subsequent episode presented as Rapunzel syndrome, and in 1 case both the initial and the recurrent presentation was as Rapunzel syndrome [25]. Hence a long-term psychiatric follow-up is advised. A follow-up endoscopy or contrast study may also be advised if trichotillomania is suspected, as the patient usually never gives a positive history. Many of these patients report having parental discontent [1, 13], bereavement [13], or other family problems [26]. Parental or spouse counseling is also be advised as a regular part of treatment to prevent recurrence.

Conclusion

Rapunzel syndrome is an uncommon trichobezoar, with a tail extending into the small intestine. It has a varied presentation and is seldom diagnosed preoperatively. Management requires gastrostomy and enterotomies, when required. A psychiatric assessment and a long-term follow-up are advocated with parental or spouse counseling as a regular part of treatment to prevent recurrence.

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