The Rapunzel Syndrome: A Case Report and Review of the Literature

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Summary

The Rapunzel syndrome, found characteristically in girls with varying gastrointestinal symptoms, is a rare form of gastric trichobezoar extending throughout the bowel. The previously reported cases have different clinical and pathological features affecting the mortality. We describe a new case preoperatively diagnosed with none of the complications previously reported.

Key words

Trichobezoar – Rapunzel syndrome

Résumé

Le syndrôme de Rapunzel est une forme rare de trichobezoard gastrique qui s'étend à tout l'intestin se voit habituellement chez la fille et présente des symptomes gastrointestinaux variés. Les observations déjà rapportées de patients décédés présentaient des particularités cliniques et pathologiques. Nous décrirons un nouveau cas diagnostique avant l'intervention et qui n'a présenté aucune des complications décrites.

Mots-clés

Trichobézoard – Le syndrome de Rapunzel

Zusammenfassung

Das Rapunzel-Syndrom tritt charakteristischerweise bei Mädchen mit verschiedenen gastrointestinalen Symptomen auf, es ist eine seltene Form des Magentrichobezoars, der sich innerhalb des Darmes ausbreitet. Die bisher berichteten Fälle hatten verschiedene klinische und pathologische Eigenschaften, die sich auf die Mortalität auswirkten. Wir beschreiben einen neuen präoperativ diagnostizierten Fall, der ohne die bis dahin berichteten Komplikationen verlief.

Schlüsselwörter

Trichobezoar – Rapunzel-Syndrom

Introduction

Bezoars are impactions of swallowed foreign material in the stomach and occasionally in the intestine. Bezoars have been known to occur in animals and men for centuries. Since bezoars were collected and used as medicinal charms after preservation, the name bezoar is thought to be derived either from the Arabic "badzehr" or from the Persian "panzehr" both meaning antidote (7). Trichobezoars rarely have a "tail", extending through the pylorus and duodenum, or through the entire length of the small intestine

Only five such cases could be found in a review of the literature. In this report, we present the sixth case of "Rapunzel syndrome".

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Case report

Z. D., a 15-year-old girl, was admitted to our department with abdominal pain of 5 days' duration. She had had anorexia for the last three months. A detailed medical history revealed episodes of bilious vomiting in the last three months and trichophagia since the age of five. She also had a left nephrolithotomy procedure 1.5 years before her admission.

Physical examination revealed a 43 kg girl, lower than 10th percentile for her age (4) in relatively good general condition with no acute distress.

A large, firm, smooth, oval-shaped, painless and movable mass was palpated in the left upper quadrant and epigastrium. There was no abdominal distension, guarding, rigidity or tenderness. Laboratory examinations were all within normal range. At plain abdominal roentgenogram, lateral and inferior displacement of intestinal gas shadows could be detected. Ultrasonography revealed a solid mass in the upper abdomen giving a mixed echogenic pattern. Barium meal demonstrated a large filling defect in the stomach with a so-called "bubbly" appearance which gave the first clue to a bezoar (Fig. 1). Computerized tomography of the upper abdomen confirmed the diagnosis (Fig. 2).

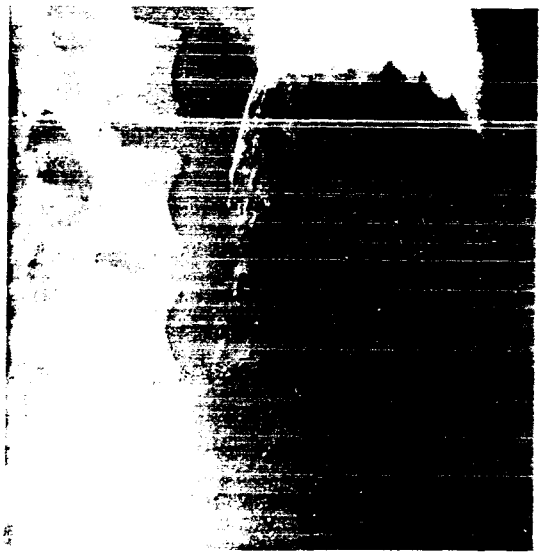

Fig. 1 After barium meal a large intragastric mass could be seen at abdominal roentgenogram.

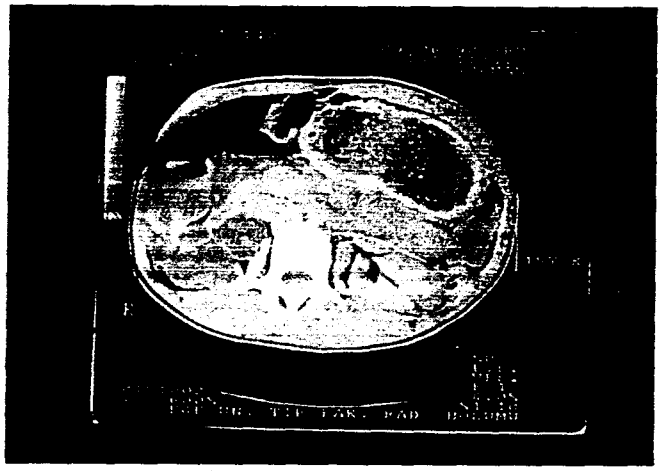

Fig. 2 Computerized tomography of the upper abdomen detecting intragastric trichobezoar.

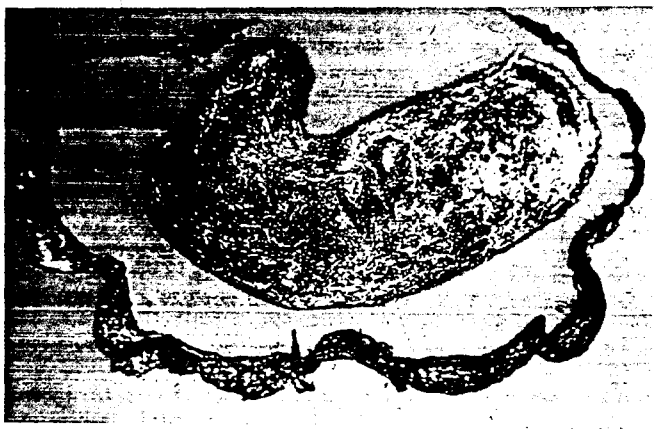

Fig. 3 Gastric trichobezoar and its intestinal extension.

At laparotomy, the stomach was extensively filled with a heavy solid mass. Gastrotomy revealed a large trichobezoar, extending to the jejunum through pylorus and duodenum. The gastric bezoar and the intestinal extension were extracted through the gastrotomy incision by stringing the intestine along the length of the bezoar. The trichobezoar was 1250 g, 20×10 cm in dimensions and with a tail extending 53 cm (Fig. 3).

The patient had an uneventful recovery. Psychiatric evaluation revealed no signs of active psychologic disturbance and ten months postoperatively she was in excellent condition with good appetite and weight gain.

Discussion

Although recent reports accept phytobezoars to be the most common type, trichobezoars are still the leading bezoars in childhood (1, 3, 7).

The typical child with a trichobezoar is an adolescent girl having anorexia, weight loss and abdominal discomfort with a history of trichophagia. As in our case, a psychological disorder could not always be identified in the reported cases (1–7). Physical examination sometimes reveals alopecia produced by intentional hair pulling which may be a clue to the diagnosis. A movable mass in the upper abdomen is usually palpated, although small masses may be easily overlooked in an uncooperative young child (7). Plain abdominal roentgenograms and an upper gastrointestinal barium meal are usually sufficient in confirming the diagnosis. However, in elective conditions, ultrasonography, computerized tomography and gastroscopy may be used for a more precise preoperative diagnosis.

The syndrome that bears Rapunzel's name, the long-haired heroine of a tale told by *Grimm* brothers, was first described by *Vaughan* et al who reported two cases in 1968 (6). *Deslypere* et al in Belgium and *Büyükünal* et al in Turkey reported the third and forth cases in 1982 with fatal outcomes (2, 3). An additional patient with Rapunzel syndrome was reported by *Wolfson* et al in 1987 (7) (Table 1).

Although all of these authors called their individual patients' pathological condition Rapunzel syndrome, the clinical and the pathological features of the patients were not all alike. Even the pathological features of the two patients of *Vaughan* et al were not identical. The trichobezoar mass, the tail extending through the small bowel, and the gastrointestinal symptoms of various degrees seem to be the only three common

Table 1 The Rapunzel syndrome cases reported in the literature.

	Age	Sex	Treatment	Complication	Outcome
1. *Vaughan* et al (1968)	13	F	Surgical	Peritonitis	Alive
2. *Vaughan* et al (1968)	15	F	Surgical	–	Alive
3. *Deslypere* et al (1982)	14	F	Medical	Metabolic	Dead
4. *Büyükünal* et al (1982)	5	F	Surgical	Sepsis	Dead
5. *Wolfson* et al (1987)	5	F	Surgical	Volvulus	Alive
6. *Balık* et al (1992)	15	F	Surgical	–	Alive

Table 2 Suggested clinical grading in Rapunzel syndrome.

Grade	Symptoms	Pathology	Treatment
Grade 1	Intermittent abdominal pain, anorexia, weight loss	Partial intestinal obstruction	Surgical: Gastrotomy and/or enterotomy
Grade 2	Abdominal distension, vomiting, obstipation	Complete intestinal obstruction	Surgical: Gastrotomy and enterotomy
Grade 3	Peritonitis, and Grade 2 symptoms	Bowel necrosis, perforation, peritonitis	Surgical: Gastrotomy, multiple enterotomies, excessive bowel resection

features of the Rapunzel syndrome. The pathophysiology of the events beginning with partial intestinal obstruction and leading to bowel necrosis and perforation, prompted us grading these patients according to their pathological findings. These pathological features seem unrelated to the size and length of the mass, although they are in close relation with morbidity and mortality.

In conclusion, we suggest a grading system which is thought to be helpful in predicting mortality in the patients with Rapunzel syndrome (Table 2). Such a grading system will obviously show the necessity of early diagnosis and accurate treatment of patients with suspected trichobezoars.

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