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Case Report

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A Case of Rapunzel Syndrome

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Rapunzel syndrome refers to a very rare condition in which swallowed hair forms a gastric trichobezoar that has a long tail extending into the small bowel. We describe a case of Rapunzel syndrome in an 8-year-old girl who presented with abdominal mass, epigastric pain and vomiting. Abdominal computed tomography scan showed a markedly dilated stomach filled with coarse heterogeneous materials. Upper gastrointestinal endoscopy revealed a huge hairy ball with a tail extending through the pylorus. We performed a surgical laparotomy and successfully removed a huge trichobezoar with a long tail extending into the middle portion of jejunum. Psychiatric consultation with review showed her past history of trichotillomania and trichophagia 4 years ago. But her parents denied further psychiatric therapy and she was lost to the follow-up. Rapunzel syndrome should be included in the differential diagnosis in children with chronic abdominal pain and trichophagia. **(Pediatr Gastroenterol Hepatol Nutr 2013; 16: 127～130)**

Key Words: Rapunzel syndrome, Bezoars, Trichotillomania, Trichophagia, Abdominal pain

INTRODUCTION

A bezoar is a concretion of undigested exogenous material that accumulates in the gastrointestinal tract of humans and some animals. The term "bezoar" is derived from Arabic *bedzehr* or *Persian* padzhar, meaning "counterpoison" or "antidote." Some societies have historically believed bezoars from animal guts had magical properties and used them as antidotes to certain poisons [1]. The following 4 types of bezoars have been described: phytobezoars (comprising vegetable or fruit fibers), lactobezoars (comprising milk curds), trichobezoars (comprising hair), and pharmacobezoars (comprising pills or capsules) [2].

Unlike other bezoars, trichobezoars are common in patients with underlying psychiatric disorders who chew and swallow their own hair (i.e., trichotillomania and/or trichophagia). Rapunzel syndrome is a very rare and severe form of gastric trichobezoar with a long tail extending into the small bowel . Rapunzel syndrome can be an unusual cause of chronic abdominal pain or gastric outlet obstruction in children and adolescents [2,3].

Here, we report a case of Rapunzel syndrome in an 8-year-old girl who presented with symptoms of gas-

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tric outlet obstruction.

CASE REPORT

An 8-year-old girl was referred to our pediatric clinic with a 3-month history of palpable abdominal mass, epigastric pain, and intermittent vomiting that worsened after meals. Her mother reported several episodes of vomiting, which consisted of undigested recently eaten food. There was no history of recent illness, fever, diarrhea, or ingestion of toxic substances. Physical examination of her abdomen revealed hyperactive bowel sounds and a palpable firm mass in the epigastric region (approximately 15×10 cm). No acute peritoneal signs were evident. The patient had long hair, and no erythema, scaling, or scalp edema was present. The rest of the physical examination was unremarkable.

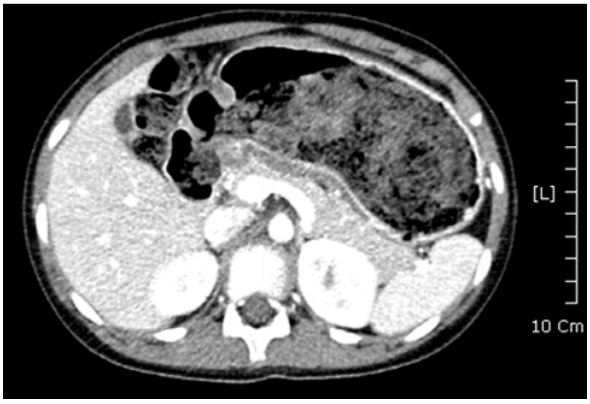

Fig. 1. Abdominal computed tomography scan reveals a huge, free-floating heterogeneous solid mass with mottled gas pattern in the markedly distended stomach.

Laboratory findings were all within normal limits, including complete blood cell counts, serum electrolyte levels, liver and renal function tests, complete metabolic panel, and pancreatic enzyme levels. Plain radiograph of the abdomen revealed an unusual gas pattern in the left upper quadrant with a rim of air inside. Abdominal computed tomography (CT) scan revealed a huge free-floating heterogeneous solid mass with a mottled gas pattern in the markedly distended stomach (Fig. 1). The diagnosis of gastric trichobezoar was made, and removal through an upper gastrointestinal endoscopy was considered. Upper gastroscopy revealed a huge gastric trichobezoar mixed with undigested food material occupying almost the entire gastric cavity. A long hairy tail was exiting the pylorus and extending into the duodenal bulb; the gastric mucosa appeared normal without evidence of ulceration (Fig. 2). An attempt to remove the trichobezoar using foreign body forceps failed because of its extensive size. Consequently, surgical laparotomy was performed. A gastrostomy was made in the anterior wall of the stomach, and the huge trichobezoar was subsequently identified and extracted. The stomach-shaped mass measured 14.0×10.0×5.5 cm, weighed 407 g, and had a long tail of hair extending through the pylorus into the middle portion of the jejunum (Fig. 3).

The patient had an uneventful postoperative course and was discharged and returned home 7

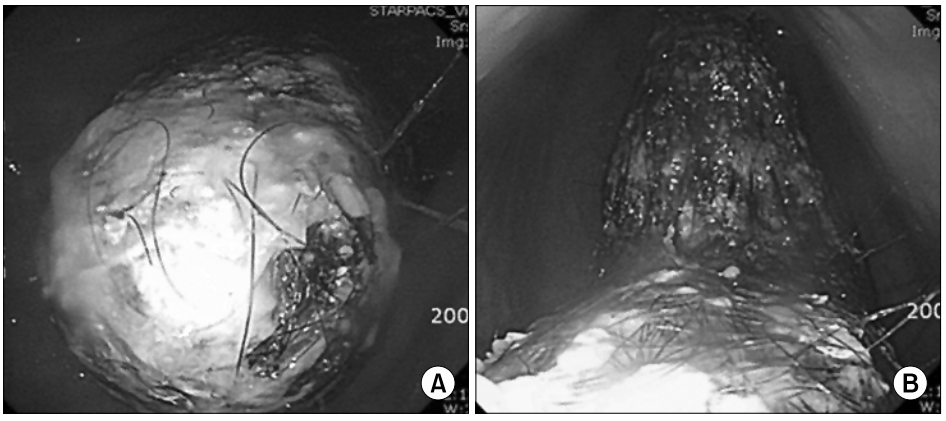

Fig. 2. Upper gastrointestinal endoscopy showing a huge gastric trichobezoar (A) with a tail which extends through the pylorus (B), and the gastric mucosa appears normal without evidence of ulceration.

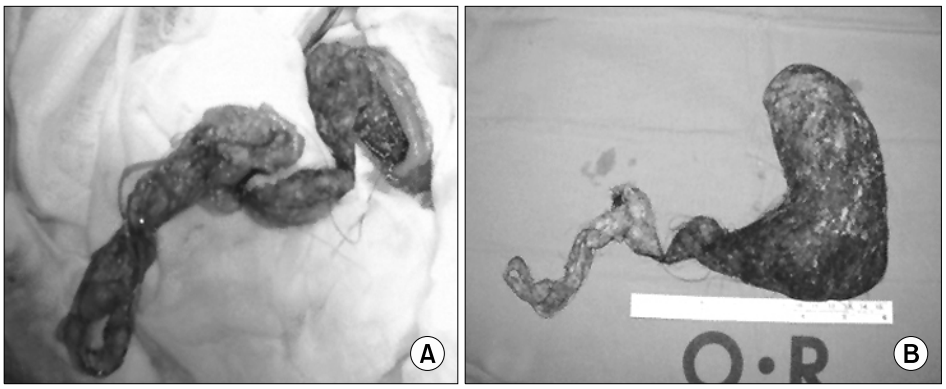

Fig. 3. (A) A very long extension of the hair and trichobezoar is extracted from the stomach through the gastrotomy. (B) A stomach-shaped trichobezoar (measuring 14.0×10.0×5.5 cm and weighing 407 g) with a tail extending into the middle portion of jejunum.

days later. During her admission, a psychiatric consultation with a review revealed a history of trichotillomania and trichophagia 4 years ago. However, we failed to find the fundamental causes of stress or anxiety disorder associated with her trichotillomania and trichophagia. Her parents were advised to visit a pediatric psychiatrist for further psychiatric intervention. However, they refused further psychiatric treatment, and the patient was lost to follow-up.

DISCUSSION

Rapunzel syndrome refers to a condition in which the swallowed hair forms a trichobezoar in the stomach with a long tail extending into the small bowel. It is named after the eponymous heroine of a German fairy tale written by the Grimm Brothers in 1812 about a 12-year-old princess imprisoned by a witch in a tall tower with neither stairs nor doors for many years; the princess lowered her long hair to the ground from her window, allowing a young prince to climb up and rescue her [4].

Rapunzel syndrome was first described by Vaughan et al. in 1968 [5]. Although there is no consensus about its definition, the commonly accepted one is a gastric trichobezoar with a tail extending up to the jejunum, ileum, or ileocecal junction [6]. The patient in our case presented with a gastric trichobezoar with a long tail of hair extending into the middle portion of the jejunum, causing symptoms of gastric outlet obstruction at a very young age.

Most cases of trichobezoars usually have underlying psychiatric disorders including trichotillomania and trichophagia, and more than 90% are reported in young girls [2,3,6]. Patients may remain asymptomatic for many years until the trichobezoar grows to the point of obstruction. The typical presentation of trichobezoars and Rapunzel syndrome includes palpable abdominal mass, chronic abdominal pain, nausea/vomiting, and constipation. Some patients present with weight loss, anorexia, hematemesis, iron deficiency anemia, and intussusception [2,6]. The reported complications of Rapunzel syndrome include gastric ulceration, incomplete pyloric obstruction, obstructive jaundice, acute pancreatitis, complete intestinal obstruction, gastric perforation, peritonitis, and mortality [7-10].

Elucidating a patient's history of trichotillomania and/or trichophagia is essential for the diagnosis of Rapunzel syndrome or trichobezoars. In some patients, putrid halitosis and patchy alopecia can be clues on physical examination. Imaging studies including plain abdominal radiograph, contrast upper gastrointestinal series, and abdominal CT scan may show the trichobezoar as a mass or filling defect in the stomach and small bowel. Among these modalities, CT scan with contrast can delineate the extension of trichobezoars [11]. Upper gastrointestinal endoscopy is considered the gold standard for the diagnosis of trichobezoars and Rapunzel syndrome. Upper endoscopy can enable direct visualization of trichobezoars as well as potential therapeutic intervention [12]. However, as in the present case, endo-

scopic retrieval is rarely a definitive treatment option, because gastric trichobezoars are often very large. Therefore, surgical removal remains the mainstay of treatment of large trichobezoars and Rapunzel syndrome [13]. Open surgery is usually accomplished by gastrostomy or enterotomy as in the present case, but recent reports describe successful laparoscopic removal of trichobezoars in pediatric patients [14,15]. Parental counseling, behavioral therapy, and long-term psychiatric follow-up are important parts of treatment and the prevention of recurrence.

Although case reports of pediatric patients with trichobezoars or Rapunzel syndrome are very rare, the present case indicates that a high index of suspicion is required to make a diagnosis of Rapunzel syndrome in order to provide timely and adequate management.

In conclusion, Rapunzel syndrome should be included in the differential diagnosis of pediatric patients with chronic abdominal pain, vomiting, and trichophagia. Early upper gastrointestinal endoscopy is recommended for making a correct diagnosis and treatment plans in pediatric patients with Rapunzel syndrome and/or trichobezoars.

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