Case Report

Rapunzel Syndrome in a Thai Girl with an Asymptomatic Abdominal Mass: A Case Report

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Rapunzel syndrome is an uncommon form of trichobezoar (a collection of ingested hair in the stomach that fails to pass through the intestine) with an extension of hair into the small bowel. The authors report in the present article a case of Rapunzel syndrome in a 10-year-old Thai girl with an asymptomatic abdominal mass incidentally detected during a hospital visit. She did not have nausea, vomiting, abdominal pain, weight loss, or any other symptoms. A movable, firm and smooth epigastric mass 10 x 12 cm in size was found upon examination. Eventually the trichobezoar mass was surgically removed after a failed endoscopic removal.

Keywords: *Abdominal mass, Trichobezoar, Rapunzel syndrome*

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Bezoar is the collection of foreign material in the stomach that fails to pass through the intestine. Its name is derived from a Persian term meaning an antidote. It can be categorized by content within it (*e.g.* lactobezoars from milk curd, pharmcobezoars or medication bezoar, phytobezoars from indigestible plant, or trichobezoar from hair). Trichobezoar is the most common type of bezoar in humans, usually occurring in emotional disturbance or mental-retarded patients. Most cases are female who eat their own hair. Rapunzel syndrome is an uncommon form of trichobezoar, first described by Vaughan et al in 1968[1], with an extension of hair into the small bowel. It obtains its name from that of a long-haired girl Rapunzel in the fairy tale of Brothers Grimm. Patients are usually presented with abdominal pain, nausea, vomiting, weight loss and sometimes with an abdominal mass and even gut obstruction and perforation[2]. To date, there have been quite a few reported cases of Rapunzel syndrome in children[3]. The authors report in this article a case of Rapunzel syndrome in a Thai girl with an asymptomatic abdominal mass.

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Case Report

A 10-year-old girl from a province on the western border of Thailand came with an abdominal mass for two months. The mass was incidentally found by a physician at a district hospital during the girl's visit due to muscle sprain. She did not have nausea, vomiting, abdominal pain, or weight loss. Her bowel movement was also normal. Despite the absence of constipation, laxative was given by the physician without improvement of the abdominal mass. Her parents had died of AIDS seven years before the episode and she had lived with her aunt ever since. She had received HIV prenatal prophylaxis and blood for anti-HIV yielded negative results. She had no other past medical illness and denied emotional disturbance. Upper GI study at the hospital revealed dilated stomach with a lot of gastric content and delayed passage of contrast into the small bowel (Fig. 1). She was referred to Phramongkut-klao Hospital for further evaluation.

Upon physical examination at Phramongkut-klao Hospital, a movable epigastric mass 10 x 12 cm in size with smooth surface and firm consistency was found without tenderness. Other physical findings were unremarkable. Complete blood count and complete metabolic panel were all normal. Abdominal ultrasonography showed that the stomach was dilated and filled with content but the mass was not seen. A large hair ball extending beyond the pylorus was

detected by upper endoscopy (Fig. 2). Endoscopic removal was tried without success; the hair ball could not be pulled out of the stomach. Eventually, surgical removal via gastrotomy was done and one piece of huge, foul smelling trichobezoar with a tail extending into the small intestine was successfully removed (Fig. 3). Additional history from her aunt later disclosed that the patient had habitually pulled out her own hair and eaten it (trichophagia) at around three to four years of age. After the girl had moved to stay with her aunt, the habit of trichotillomania and trichophagia was not noticed again. The patient herself denied any history of eating hair during recent years too. Psychiatric evaluation did not show serious emotional or behavioral problems. She was discharged after a few days without complication.

Discussion

Trichobezoar is commonly found in young females with underlying emotional disturbance or mental retardation. Most cases are reported from Asian countries or countries where women traditionally have long hair, although a number of cases from Europe or United States have also been reported[3-5]. Ingested hair strands are accumulated in the gastric folds and are not easily moved by peristalsis because, as explained by some authors[3,4], their slippery surface prevents enough friction required to push them out of the stomach. The size of hair ball thus increases overtime as a result of continuous accumulation of hair and other non-absorbable food or fibers. Definitions of Rapunzel syndrome vary; some authors define it as a trichobezoar with long tail extending up to the jejunum, ileum, or ileocecal valve while others indicate concomitant intestinal obstruction[4,6,7]. Its typical features involve both types of definitions where trichobezoar has a tail extending at least to the jejunum and symptoms insinuating intestinal obstruction[4]. Common clinical presentations of Rapunzel syndrome include abdominal pain, nausea, vomiting and gut

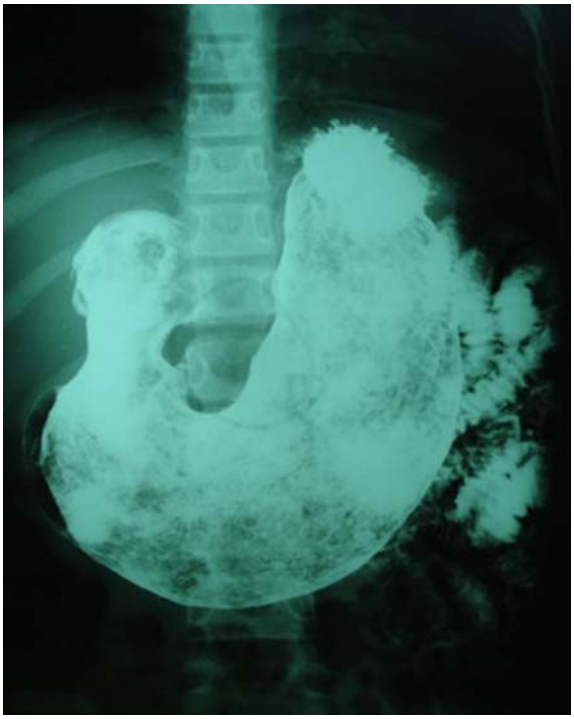

Fig. 1 Upper GI study shows dilated stomach with a lot of gastric content and delayed passage of contrast into the small bowel

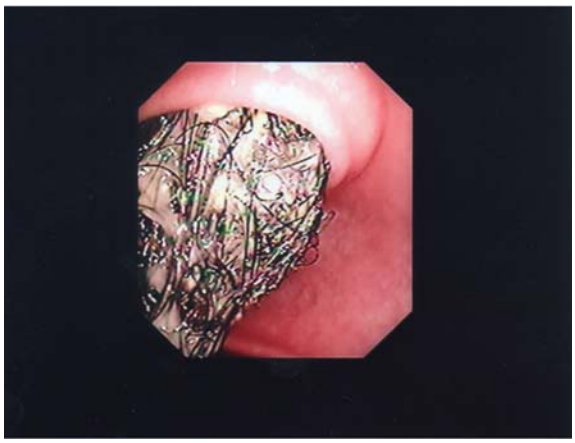

Fig. 2 Upper endoscopy demonstrates a large hair ball extending beyond the pylorus

Fig. 3 A huge, foul smelling trichobezoar with tail extending into the small intestine which was successfully removed

obstruction[4]. Less common presentations are peritonitis, weight loss, anorexia, hematemesis[3,4] and intussusception[8,9]. Mortality in a neglected child has been reported and was caused by gut perforation[10]. An abdominal mass without other symptoms can lead to delayed diagnosis as the syndrome is more likely to be overlooked[11]. Plain abdominal radiologic or barium study shows intraluminal filling defects and sometimes radiologic signs of gut obstruction. Diagnosis of gastrointestinal bezoar can be made by ultrasonography or CT scan of the abdomen as well. CT scan is more accurate and will show an intraluminal mass containing a mottled air pattern in such a case[12]. However, endoscopy is becoming an established diagnostic tool due to its ability to directly visualize the affected area while removal may be done in the same setting. Endoscopic removal can be achieved with small bezoar especially phytobezoar and lactobezoar but may be more challenging with trichobezoar, particularly in Rapunzel syndrome. In this circumstance, surgical removal is indicated[13].

In the present case, there were no typical symptoms of abdominal pain and gut obstruction concurrent with the abdominal mass despite endoscopic finding of a huge trichobezoar in the stomach occupying a large area of the pylorus with the tail extending into the small intestine; a finding that would make the diagnosis of Rapunzel syndrome. This lack of symptoms might be explained by slow and gradual accumulation of hair strands over a long period. Endoscopic removal unsurprisingly failed to pull out trichobezoar through the pylorus and laparotomy was needed to successfully remove the whole trichobezoar mass. Surgery by means of gastrotomy or enterotomy is still the mainstay for gastric trichobezoar removal especially those that extend into the intestine[3]. Nevertheless, a less invasive laparoscopic removal of gastric trichobezoars has been reported to operate successfully[14].

Psychiatric evaluation is an essential part of management in all patients. Trichotillomania and trichophagia, usually associated with psychiatric disorders, will lead to trichobezoar[15] though many patients deny a history of eating hair. Recurrent trichobezoar has been reported[16] hence a long-term psychiatric follow-up is necessary. The presented patient had a history of trichophagia at the time of parental illness and death, therefore parental deprivation was assumed as a precipitating factor in the present case. Although at the moment she lived happily with her aunt and refused trichophagia, psychiatric counseling and long-term follow-up was needed to prevent recurrence.

Conclusion

Trichobezoar or Rapunzel syndrome should not be overlooked in children coming with an asymptomatic abdominal mass. Without typical symptoms of abdominal pain, nausea and vomiting, making a prompt diagnosis may be difficult and this may lead to delay in management. History of pica eating should be sought out along with other evaluative measures. Abdominal ultrasound, CT scan and endoscopy can help establish the diagnosis. Small bezoars can be removed by endoscopy but larger ones or those with Rapunzel syndrome require surgical removal.

Potential conflicts of interest

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รายงานผู้ป่วย: กลุ่มอาการ "แรพพันซึ่ล" ในเด็กหญิงที่มาด้วยก้อนในท้องแบบไม่มีอาการ

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กลุ่มอาการ "แรพพันซึ่ล" (Rapunzel syndrome) เป็นลักษณะหนึ่งของผู้ป่วยที่รับประทานเส้นผมจนเกิดเป็นก้อนในกระเพาะอาหาร หรือที่เรียกว่า trichobezoar แบบที่มีส่วนของก้อนเส้นผมยื่นยาวผ่านกระเพาะอาหารออกไป ผู้นิพนธ์รายงานผู้ป่วยเด็กหญิงไทยอายุ 10 ปี ซึ่งมาด้วยเรื่องก้อนในท้องที่ถูกตรวจพบโดยบังเอิญ ในขณะที่ผู้ป่วยไปพบแพทย์ด้วยปัญหาอื่น ผู้ป่วยไม่มีอาการคลื่นไส้ อาเจียน ปวดท้อง น้ำหนักลด หรือความผิดปกติอื่น ๆ ก้อนมีขนาด 10 x 12 เซนติเมตร อยู่ที่บริเวณ epigastrium ก้อนนี้ถูกตรวจพบว่าเป็นก้อนเส้นผมโดยการส่องกล้องทางเดินอาหาร และได้ถูกผ่าตัดออกมาหลังจากไม่สามารถเอาออกผ่านทางกล้องส่องทางเดินอาหารได้