Case Report

Rapunzel Syndrome: An Uncommon Cause of Gastric Perforation

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ABSTRACT

Bezoars are due to the accumulation of indigestible substances in the digestive tract. In the case of trichobezoar, it is an accumulation of ingested hair fold. Rapunzel syndrome is a very rare variant where the trichobezoar developed in the stomach, extends the intestine. Reported for the first time in 1968 and since then, less than 70 cases are reported worldwide.

We report a case of Rapunzel syndrome, a complicated by a gastric perforation, in a 17-year-old patient presented for acute peritonitis. The trichobezoar was discovered operatively and the patient was treated successfully with open surgery (gastrectomy).

Keywords: Rapunzel syndrome; Trichobezoar; Stomach; Intestine; Gastric perforation

Introduction

Bezoars are due to the accumulation of indigestible substances in the digestive tract [1]. In the case of trichobezoar, it is an accumulation of ingested hair fold inside the stomach [2]. A special entity trichobezoar is the Rapunzel syndrome; it is a very rare condition where the trichobezoar, developed in the stomach, extends the intestine [3]. Bezoars can present to 4% of bowel obstruction but they can rarely form and stop in the stomach due to their flexible volume [1].

We report a case of Rapunzel syndrome complicated by a gastric perforation within a 16-year-old patient.

Case Report

A 17-year-old female patient presented to the emergency department with abdominal pain, bilious vomiting, and fever evolving for about 10 hours. Careful interrogation of the patient and his family dined any history of tobacco, alcohol, or illicit drug use. The physical examination revealed a mildly dehydrated patient. The body temperature was 38.7°C. The heart rate was 110 beats/minute, arterial blood pressure was 110/65 mmHg, and respiratory rate was 22 cycles/minute. Abdominal examination revealed rigidity. No abdominal mass was palpable or any other abnormalities were found. Laboratory tests revealed a biological inflammatory syndrome: white blood cells of 16200 elt/l and CRP of 38 mg/l. The chest X-ray showed a pneumoperitoneum. The diagnosis of peritonitis due to a digestive perforation was made and we decide to operate on the patient. At first, we started with a laparoscopy. Per operatively, we find a gastric antrum 5 mm perforation. While examining the stomach we found a foreign body all its form with hair exteriorized hair from the perforation. We decided to convert to a mid-line incision. We proceeded to a gastrotomy and extracted a large bezoar taking the gastric form and extending to the duodenum **(Figures 1 and Figure 2)**. A gastric biopsy was performed, and later histological exams showed no malignant proliferation.

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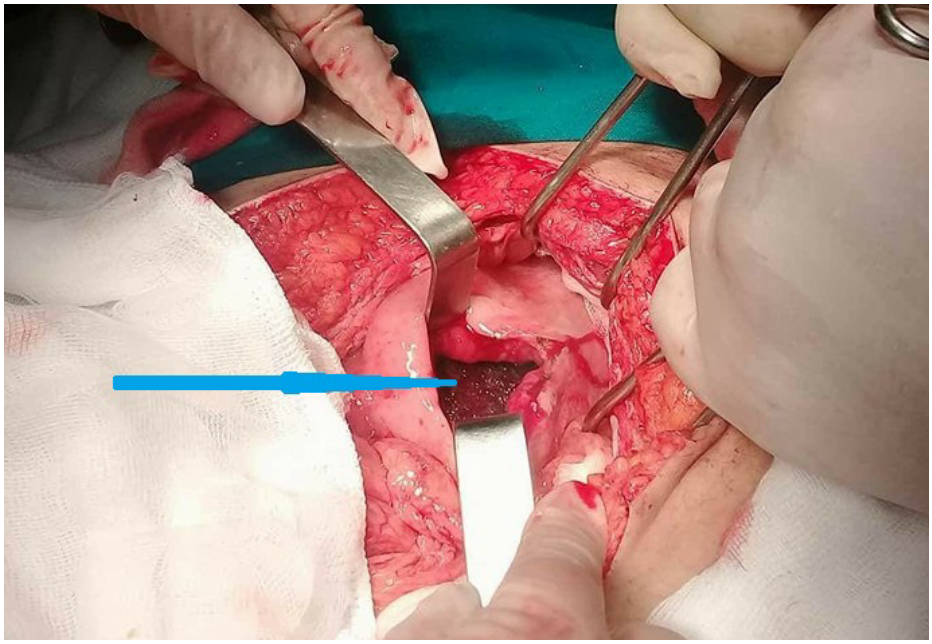

Figure 1: Hair folds inside the stomach (blue arrow).

Figure 2: The trichobezoar takes the form of the stomach and extends to the small bowel.

Postoperatively the patient was given intravenous pump inhibitors, antibiotics, paracetamol, and a daily prophylactic subcutaneous enoxaparin injection. Oral feeding was started on the third postoperative day. Outcomes were simple. She was discharged on the fifth postoperative day and she was referred for psychiatric follow-up. She attended two follow-up visits after 15 days and one month.

Discussion

Bezoars are due to the accumulation of indigestible substances in the digestive tract [1]. Those bezoars can be classified into four categories; phytobezoars which are the most common, trichobezoars, pharmacobezoars, and lactobezoars [4]. Trichobezoar was reported for the first time in Baudamant 1779[5]. This entity must be associated with trichotillomania and trichophagia [6]. Some recent studies found that 1.7% of people aged 16-69 suffer from trichotillomania, among them 30% were suffering from trichophagia [7]. This condition is usually found within preadolescent or adolescent

female patients with a psychiatric history or developmental delay [8].

In 1968 Vaughan and colleagues described for the first time a variant of trichobezoars called Rapunzel syndrome [5]. It was named after the long-haired heroine of the Brothers Grimm fairy tale [5,7]. It is due to the extension of the gastric bezoar to the small bowel [3]. Since discovered and until 2021, less than seventy cases were reported [7].

Unlike small bowel bezoars that are revealed by bowel obstruction, gastric bezoars are mostly asymptomatic [9]. However, they can be discovered while exploring various digestive or general symptoms like anemia, vomiting, nausea, constipation, or simple abdominal pain [10]. In some cases, such as ours, they can evolve to gastrointestinal ulceration, visceral perforation, and bleeding or pressure necrosis [11].

If not complicated, the visualization of the upper segment of the bezoar remains the key to the diagnosis and sometimes it permits to extract it [12]. CT-scan imaging has the superiority of showing the exact extension of the bezoar and detecting multifocal ones which permits to establish the therapeutic strategies [13].

Unlike phytobezoars that can be dissolved by using acetylcysteine, cellulase, phosphoric acid, or even Coca-Cola, the trichobezoars cannot be dissolved [14]. They must be removed surgically or using endoscopy [7]. The success extraction rates if surgery used are 100% with laparotomy while only 75% for laparoscopy [7,15]. For endoscopy, it is only indicated in the absence of gravity signs like signs of bowel perforation and with success rates only of 5% [15]. In our case, the patient presented for peritonitis and was operated on. However, we should keep in mind that this is an entity that is always associated with mental disorders [1,7,8]. That is why it is mandatory to set up psychotherapy and a psychological follow-up as soon as possible after the extraction to prevent relapses [7,15].

Conclusion

Trichobezoar is a rare condition mostly seen within female teenagers. It is generally associated with Trichotillomania that's why psychiatric care is essential for those patients. Symptoms are usually poor and are those of complications. The CT scan has proven its sensibility for the diagnosis in doubtful cases.

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