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Case Report

Successful endovascular treatment of superior mesenteric artery-duodenal fistula secondary to Rapunzel syndrome ☆

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ABSTRACT

Rapunzel syndrome is a rare clinical entity in pediatric patients with a history of trichotillomania and trichophagia that has only been mentioned a few times in the literature. It is characterized by abnormal gastric bezoar formation that sometimes extends to the duodenum, jejunum, or colon. Here, we present a case of a 16-year-old previously healthy female patient who had prolonged hospitalization due to complications related to a significant gastric bezoar that led to massive bleeding due to a superior mesenteric artery (SMA)-duodenal fistula successfully treated with stent graft placement. Undiagnosed trichobezoar can lead to rare and unexpected complications, such as SMA-duodenal fistula, with life-threatening hemorrhagic shock. Prompt activation of massive transfusion protocol and endovascular control of the hemorrhage was vital to successfully treating our patient.

Introduction

Trichotillomania is a psychiatric disorder in which the subject experiences compulsions to pluck and sometimes consume their hair (trichophagia). Trichotillomania is prevalent in around 1.7% of adults and commonly co-occurs with other psychiatric disorders [1]. In patients who exhibit trichophagia, large amounts of indigestible human hair in the stomach can sometimes accumulate to form a mass called a trichobezoar which can lead to epigastric pain, gastrointestinal obstruction, ulceration, and rarely perforation. When portions of the trichobezoar extend distally beyond the pylorus, the condition is termed Rapunzel Syndrome in reference to the long-haired fairy tale character. We report the case of a 16-year-old female with previously undiagnosed trichotillomania and trichophagia who developed a large gastric trichobezoar extending to the duodenum, which caused obstruction leading to perforation with surgical repair and a clinical course complicated by hemorrhagic shock requiring urgent intervention.

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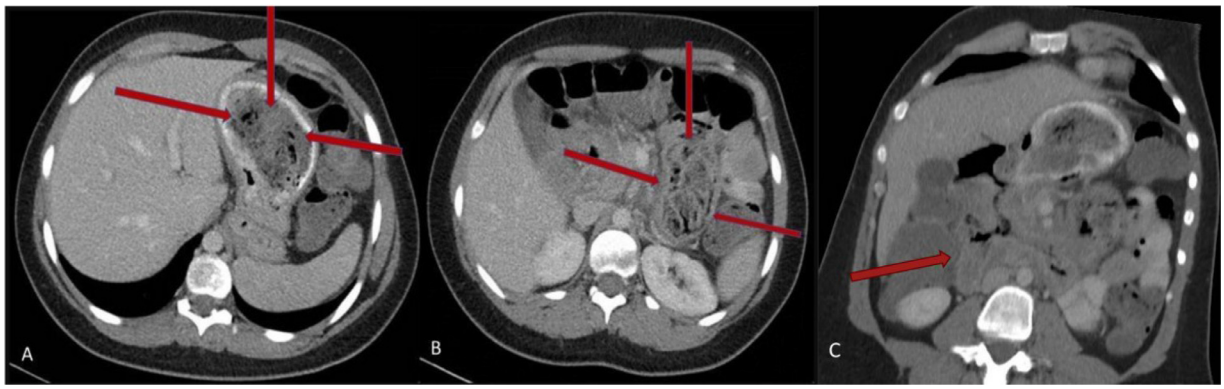

Fig. 1 – (A) CT abdomen and pelvis showing a large gastric bezoar. (B) Diffuse wall thickening and edema of the proximal duodenum with bezoar distending the ascending duodenum (C) Coronal oblique CT reconstruction showing the bezoar in the stomach with diffuse thickening of the duodenal wall and large duodenal ulceration.

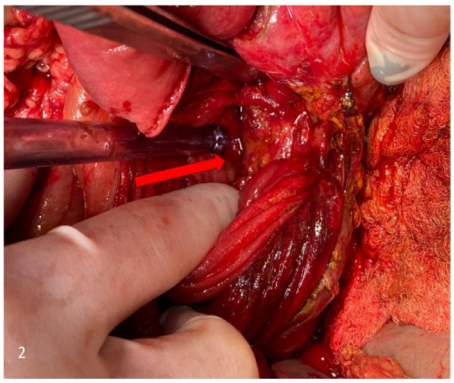

Fig. 2 – Surgical picture shows the large duodenal ulcer and ligated distal duodenum.

Case report

A 16-year-old female presented to the emergency department for hematemesis and respiratory distress. Before her arrival, she had been experiencing several weeks of weight loss, constipation, and abdominal pain and visited the emergency department 4 times. An abdominal CT showed duodenitis and heterogeneous masses with a mottled gas pattern within the stomach (Fig. 1A) and distal duodenum (Fig. 1B), suggesting a bezoar (Fig. 1C). An endoscopy performed upon admission confirmed the presence of a duodenal ulcer (Fig. 2) and a large trichobezoar, which could not be removed endoscopically. The patient's hemoglobin dropped to 6.9 g/dL the following day, requiring a blood transfusion. Repeat endoscopy followed by an attempt of laparoscopic removal of the bezoar was unsuccessful. Subsequently, the patient developed aspiration-related acute respiratory distress syndrome (ARDS) and was transferred to the University Hospital. After that, she underwent laparotomy with the removal of a large trichobezoar repair of 3 areas of duodenal ulcerations. Two reopening laparotomies were required in the following days with peritoneal washout, duodenectomy with duodenojejunostomy, duodenostomy, gangrenous ileum resection with ileocolic anastomosis, and gastrostomy tube placement.

The patient continued to recover for 1 week after her final abdominal surgery when she suddenly developed severe hypotension and bleeding from her surgical drains, jejunostomy, and gastrostomy. Massive transfusion protocol was initiated, and Interventional radiology was emergently consulted. A mesenteric arteriogram showed a large duodenal-SMA fistula (Fig. 3A). Hemostasis was temporarily achieved via a 4 mm x 4 cm Armada balloon placed across the area of active bleeding from the SMA. The balloon was periodically deflated and reinflated every 30 seconds. Upon discussion with vascular and pediatric surgery, a decision was made to place a stent graft placement across the fistula. A 6 mm x 2.5 cm Viabahn-covered stent (Gore Medical, Newark, DE) was deployed across the fistula with the proximal aspect of the stent distal to the inferior anterior pancreaticoduodenal artery. Poststenting angiogram demonstrated persistent bleeding at the distal end of the stent (Fig. 3B), and an additional 6 mm x 2.5 cm Viabahn-covered stent was deployed. A final angiogram confirmed good flow to all main SMA blanches and no residual bleeding (Fig. 3C).

Our patient's hospital stay was complicated by pancreatic necrosis, septic shock, DVT, and Posterior Reversible Encephalopathy (PRES). However, no further intervention was required for intrabdominal bleeding. The patient was discharged almost 4 months after her initial presentation to an acute rehabilitation facility due to deconditioning. One year after this incident, the patient has recovered well and was in good health at subsequent follow-up visits.

Discussion

Trichotillomania is a psychiatric disorder where the patient has an irresistible urge to pull out hair. It can be associated with compulsive hair-eating or Trichophagia [1]. First onset most commonly occurs in adolescence. Trichobezoar is defined as the abnormal accumulation of human hair in the stomach [2]. The majority of trichobezoars are found in

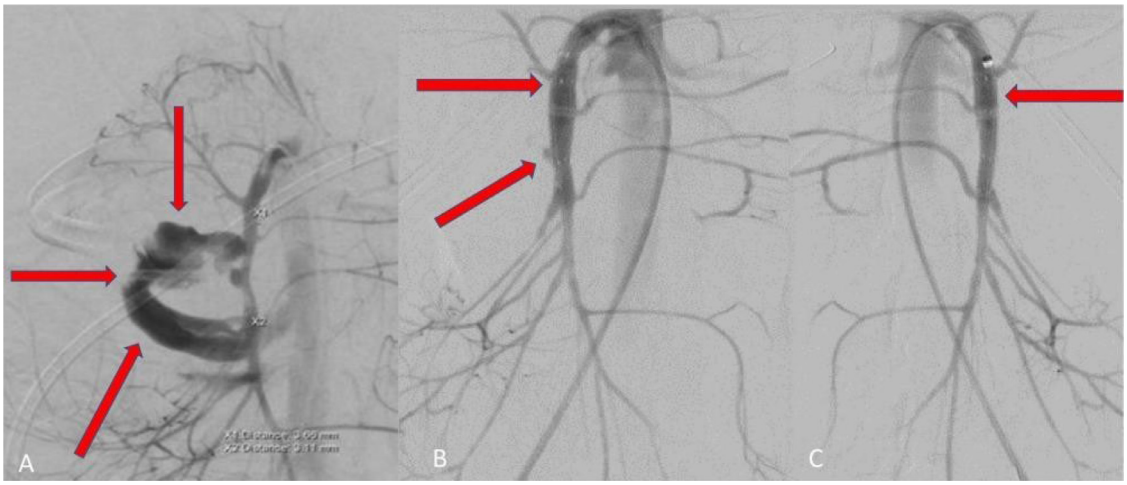

Fig. 3 – (A) Angiogram from the SMA origin showing massive SMA-duodenal fistula. Calipers are placed in the intended stent landing zone. (B) SMA angiogram after covered stent placement shows a small area of persistent bleeding at the distal margin of the stent. (C) SMA angiogram after deployment of the second covered stent shows resolution of the bleeding and preserved flow in the SMA branches.

young female patients (13-20 years old) with long hair. The presence of a trichobezoar can be associated with nonspecific gastrointestinal symptoms including abdominal or epigastric pain, nausea, vomiting, constipation, early satiety, and weight loss. However, more serious complications, including gastrointestinal obstruction, ulceration, perforation, and even severe bleeding and hemorrhagic shock, can occur if undiagnosed in its early stages. Given the potentially severe consequences of trichobezoar and the underdiagnosed nature of trichotillomania and trichophagia, providers should consider screening for these conditions in the adolescent population [3].

Only a few cases of SMA-duodenal fistulae were reported in the literature [4,5]. Furthermore, there are no previously reported cases of SMA-duodenal fistula secondary to trichobezoar. In our case, the trichobezoar remained undiagnosed for a prolonged period and eventually resulted in complications that required multiple surgeries and culminated with an SMA-duodenal fistula leading to life-threatening hemorrhagic shock. The management of this patient was particularly challenging, given her severe abdominal bleeding and hemodynamic instability. Embolization of the SMA was not an option due to inevitable catastrophic bowel ischemia, and additional surgery would carry high mortality risk. Ultimately, temporary balloon tamponade followed by covered stent placement in a time-sensitive setting successfully stopped the bleeding and saved the patient's life.

Conclusion

Our case emphasizes the need to consider trichobezoar during differential diagnosis, especially in young female patients with an unknown cause of acute abdominal pain or gastrointestinal bleeding. Moreover, it highlights the value of urgent endovascular interventions in treating arterio-enteric fistulae and severe bleeding.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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