Intestinal Obstruction and Acute Pancreatitis: An Unusual Presentation of Rapunzel Syndrome

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Abstract

Abdominal pain is a common complaint in children, and one potential cause is pancreatitis caused by mechanical obstruction of the pancreatic drainage canal. This case report describes a 14-year-old girl who presented with significant abdominal pain and a history of scattered hair loss. She was diagnosed with intestinal obstruction, caused by a large bezoar composed of hair and food debris (Rapunzel syndrome), leading to acute pancreatitis due to the blockage of the ampulla of Vater. Initial abdominal X-ray was inconclusive, but an abdominal CT scan confirmed the diagnosis. As non-surgical treatments proved insufficient, the patient was treated surgically by removing the hair mass causing the obstruction. After recovery, she was referred for mental health evaluation. This case illustrates the importance of considering rare causes of abdominal pain and recognizing the interplay between mental health disorders and physical symptoms.

Keywords: Trichobezoar; Pancreatitis; Trichotillomania; Rapunzel Syndrome; United Arab Emirates

Introduction

Abdominal pain is the most frequent symptom that brings children to emergency departments. However, acute pancreatitis is rare in children, with an incidence of only 1–13 cases per 100,000 children.[1] Diagnosis is based on symptoms of acute abdominal pain, elevated pancreatic enzyme levels, and radiological evidence of an inflamed pancreas. Several etiologies have been identified in cases of acute pancreatitis including infections, metabolic disorders, toxins, medications, trauma, and congenital malformations.[1]

Mechanical obstruction of the pancreatic drainage canal can cause stasis of the pancreatic enzymes, leading to acute pancreatitis. The obstruction can be due to stones, tumors, or trichobezoars. Rarely, a trichobezoar extending down from the stomach into the duodenum—known as Rapunzel syndrome—can trigger pancreatitis.

We describe an unusual presentation of acute pancreatitis in a teenager caused by Rapunzel syndrome, where a large trichobezoar caused mechanical obstruction of the duodenum and the ampulla of Vater.

Case Report

A 14-year-old girl presented to the emergency department (ED) with severe, stabbing epigastric pain for one day. The pain worsened by eating or changing positions and was not relieved by painkillers. She reported loss of appetite and frequent vomiting, which became bilious over time. There was no fever or jaundice. Her history

included trichophagia for almost two years. Examination revealed epigastric and right hypochondrial tenderness, poor personal hygiene, and patchy hair loss on the scalp. The patient's abdomen was soft and not distended.

Laboratory investigations revealed leukocytosis (12.9 $\times 10^3$/mcL; normal: 4–11 $\times 10^3$/mcL), neutrophilic predominance (11.4 $\times 10^3$/mcL; normal: 2.0–7.0 $\times 10^3$/mcL), and markedly elevated amylase (1,487 U/L; normal: 0-100 U/L) and lipase (2,196.0 U/L; normal: 4.0–39.0 U/L). Liver enzymes, C-reactive protein, urea, and creatinine were all within normal limits. Abdominal X-ray revealed only mottled lucent areas in the stomach, while ultrasound showed an edematous pancreas, consistent with acute pancreatitis.

Conservative management—bowel rest, hydration, pain relief, and antiemetics—failed to improve her condition, and pancreatic enzyme levels continued to increase. A computed tomography (CT) scan of her abdomen with contrast confirmed the diagnosis of Rapunzel syndrome and acute pancreatitis [Figure 1]. Within 12 hours, the patient started experiencing abdominal distension and vomiting of bile. As a result, she underwent a laparotomy.

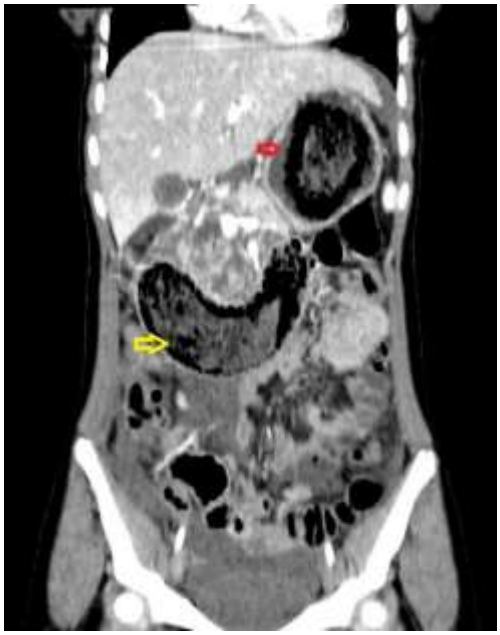

Figure 1: Coronal section of abdominal computed tomography (CT) scan showing the trichobezoar in the stomach (red arrow) and duodenum (yellow arrow).

During the surgery, an endoscopy was conducted which revealed the presence of trichobezoars in both the stomach and the second part of the duodenum. An attempt to remove them endoscopically was unsuccessful. Therefore, a laparotomy was performed by making an incision in the right supraumbilical transverse muscle. During exploration, a hard trichobezoar mass was found in the second part of the duodenum. Initially, it was firmly lodged, but the surgeon was able to move it to the jejunum. Thereafter, it was removed by conducting enterotomy in the proximal jejunum [Figure 2]. The second trichobezoar was removed from the stomach via a separate gastrotomy.

The patient had an uneventful post-operative recovery. There was immediate biochemical recovery from pancreatitis and the patient's condition improved within 48 hours. She was discharged without complications and was referred to the mental health team for management of her underlying psychological disorder.

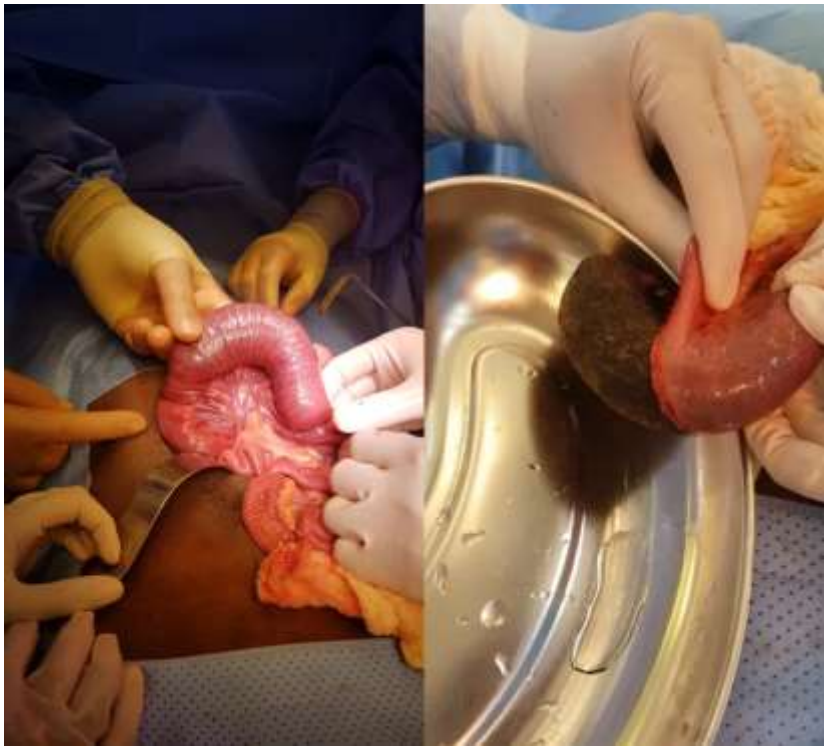

Figure 2: Trichobezoar in the intestine (on the left) and while milking it out of the stoma (on the right).

Discussion

Our patient presented with an uncommon case of intestinal obstruction where the ampulla of Vater was obstructed, leading to an impediment in the drainage of bile and pancreatic duct. This obstruction caused an episode of acute pancreatitis. The patient had a mental health condition, which involved ingesting her own hair after pulling them out. The hair accumulated with food debris to form a large mass, blocking the stomach and duodenum, including the second part where the ampulla of Vater drains. This rare condition was first reported in 1968 and is typically observed in young females. The bezoar may sometimes detach, and parts of it can migrate to more distal regions, resulting in intestinal obstruction. If the problem is not recognized promptly, the bezoar can become larger and worsen the condition.[2]

Obstruction is a common cause of acute pancreatitis in both adults and children. Gallstones often obstruct the ampulla of Vater, leading to stasis of pancreatic enzymes and bile. Other causes include strictures, tumors, edema, or impacted parasites.[3] A blocked pancreatic duct causes pancreatic enzymes to accumulate, increasing pressure. This can lead to secretions flowing back into the pancreas and activating spontaneously.[4] This leads to auto-digestion of the organ, leading to inflammation, which may not subside without the obstruction being resolved. There have been attempts to eliminate bezoars without resorting to surgery using substances that can dissolve them. Coca-Cola is recommended for small bezoars comprised mainly of food because its acidity can help dissolve the mass. Laparoscopic removal usually fails with big bezoars, as in the present case.[2]

Rapunzel syndrome is basically a psychological problem, often associated with underlying emotional trauma, including childhood neglect and abuse.[5] The underlying psychological problem has to be diagnosed and treated to prevent the recurrence of the condition. Pharmacological treatments, especially selective serotonin reuptake inhibitors (SSRI) such as fluoxetine, have shown promise in treating obsessive-compulsive type of behaviors such as trichotillomania.

Conclusion

We presented a rare case of Rapunzel syndrome complicated by acute pancreatitis which required prompt surgical intervention. It is essential to address the underlying psychological causes to prevent recurrence. By combining

effective psychological therapy, appropriate medication, and ongoing support, long-term recovery and relapse prevention are attainable.

Disclosure

The authors declare no conflicts of interest. Informed consent was obtained from the patient's father.

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