

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 pancreatitis is one of the potential causes, which can be due to mechanical obstruction of the pancreatic drainage canal. This case report describes a rare presentation of acute pancreatitis and intestinal obstruction in a teenage female caused by hair ingestion, forming a trichobezoar in the stomach and duodenum which is known as "Rapunzel Syndrome.". The 14-year-old girl presented with significant abdominal pain. Noted to have scattered and random hair loss. She was later diagnosed with intestinal obstruction, caused by a large bezoar composed of hair and food debris. This obstruction led to acute pancreatitis due to the blockage of the ampulla of Vater. Despite initial unclear findings on an abdominal X-ray, further investigation with an abdominal CT scan confirmed the diagnosis. Traditional treatments proved insufficient, prompting consideration of surgical intervention, emphasizing the importance of a comprehensive diagnostic approach in challenging cases like this. The patient was treated surgically by removing the hair mass causing the obstruction and the patient made a full recovery. She was referred to mental health for management of her underlying psychological disorder. This case illustrates the importance of considering rare causes of abdominal pain in children and highlights the potential link between mental health disorders and physical symptoms.

Keywords: Trichobezoar, Pancreatitis, trichotillomania, Rapunzel syndrome.

Introduction

Abdominal pain is the most frequent symptom that brings children to a pediatric emergency department. However, despite its prevalence, acute pancreatitis is rare in this population, with an incidence of only 1 to 13 cases per 100,000 patients.¹ Diagnosis is based on symptoms of acute abdominal pain with raised pancreatic enzymes and radiological evidence of an inflamed pancreas. Several etiologies have been identified as the main reason for acute pancreatitis including: infections (viral, bacterial, and protozoal), metabolic disorders such as hypertriglyceridemia, toxins, medications (azathioprine, anti-tumor drugs), trauma, congenital malformations among others.¹ Genetic mutation of certain genes has been studied in few experiments, but no causative mutation has been identified yet.¹ Moreover, mechanical obstruction can occlude the drainage canal and cause stasis of the pancreatic enzymes, subsequently causing pancreatitis. The obstruction can be due to a stone, tumors, or hair trichobezoar. On very rare occasions, the hair collection is extended down from the stomach to the duodenum causing what is known as "Rapunzel syndrome".

In this case report, we highlight an unusual presentation of acute pancreatitis in a teenager caused by Rapunzel syndrome due to large trichobezoar leading to mechanical obstruction of the duodenum and the ampulla of Vater.

Case Report

A 14-year-old girl arrived at the emergency department (ED) with severe, stabbing pain in the upper part of her abdomen, which had been going on for a day. The pain was worsened by eating or changing positions and was not relieved by painkillers. She also reported a loss of appetite and several instances of vomiting, initially non-bilious but later becoming bilious. The patient did not have a fever or jaundice and had no significant medical or surgical history other than trichophagia for almost 2 years. During the abdominal examination, she had tenderness and guarding in the epigastric and right hypochondrial regions, yet the patient's abdomen was soft and not distended. The patient's personal hygiene was poor, and she was noted to have spots of scalp hair loss, but the rest of her examination did not reveal any other notable findings.

Initial lab investigations revealed leukocytosis $12.92 \times 10^3/\text{mcL}$ (Normal range $4-11 \times 10^3/\text{mcL}$) with neutrophilic predominance $11.38 \times 10^3/\text{mcL}$ (Normal range $2-7 \times 10^3/\text{mcL}$). Amylase and Lipase were both elevated at $1,487 \text{ U/L}$ (Normal range $0-100 \text{ U/L}$) and $2,196.0 \text{ U/L}$ (Normal range $4-39 \text{ U/L}$) respectively. Liver enzymes, C-reactive protein, urea and creatinine were all within normal limits. Abdominal x-ray did not show any significant findings except mottled lucent areas at the site of the stomach. Abdominal ultrasound showed an oedematous pancreas indicating acute pancreatitis.

Initially, the patient was managed conservatively with bowel rest, hydration, pain relief, and medication to prevent nausea and vomiting. However, there was no improvement in her symptoms and her pancreatic enzymes continued to increase. Subsequently, a computed tomography (CT) scan of her abdomen with contrast was performed, which confirmed the diagnosis of Rapunzel syndrome - a rare condition in which a hairball extends from the stomach into the small intestine - and acute pancreatitis [Figure 1]. Within 12 hours, the patient started experiencing abdominal distension and vomiting of bile. As a result, she underwent laparotomy.

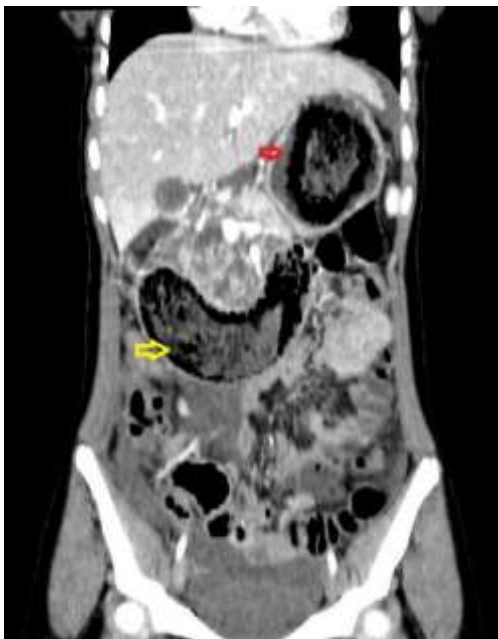


Figure 1: Coronal section of abdominal CT scan showing the Trichobezoar in the stomach and duodenum. Red arrow points to the gastric trichobezoar, and the yellow arrow points to the duodenal trichobezoar.

During the surgery, an endoscopy was conducted which revealed the presence of Trichobezoars in both the stomach and the 2nd part of the duodenum. An attempt was made to remove the Trichobezoars from the stomach but it was unsuccessful. As a result, a Laparotomy was performed by making an incision in the right supraumbilical transverse muscle. The Trichobezoars were found to be a hard mass in the second part of the duodenum. Initially, it was fixed in place but surgeon could later move it to the jejunum. The Trichobezoars were then removed by making an enterotomy in the proximal jejunum [Figure 2]. Another Trichobezoars 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. The

patient was discharged home without any complications and was referred to 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

The 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 of a foreign body with a long tail, made up of hair and food debris, was first reported in 1968, and is typically observed in young females. Medical practitioners refer to this condition as "Rapunzel" syndrome. 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 well-known cause of acute pancreatitis in adult and pediatric age groups. Commonly, gallstones can pass and obstruct the canal at the level of ampulla of Vater causing stasis of both pancreatic enzymes and bile flow. Other causes of obstruction can be due to strictures, tumors, edema, or impacted parasites.³ When the pancreatic duct becomes blocked, the pancreas will continuously produce digestive enzymes. Due to the blockage, these enzymes will build up in the duct and cause pressure to increase. This increased pressure can cause the secretions to flow back into the pancreas and become activated spontaneously.⁴ As a result of that, the enzymes will cause auto-digestion of the organ, leading to inflammation. Even with standard conservative treatments, this inflammation may not subside without the obstruction being resolved. There have been attempts to eliminate bezoars without resorting to surgery by utilizing substances that can dissolve them. Coca cola has been recommended for small masses that are primarily composed of food rather than hair. Laparoscopic removal usually fails with big bezoars.²

These patients require appropriate mental health care establishment. The underlying psychological problem has to be diagnosed and treated to prevent the recurrence of the condition. Antipsychotic medications such as Fluoxetine or other selective serotonin reuptake inhibitors have been used to treat trichotillomania. Child neglect and abuse have been linked to this condition.⁵

Conclusion

We present a case of Rapunzel syndrome complicated by acute pancreatitis which is a rare condition that must be promptly treated surgically. It is essential to address the underlying causes, including mental health issues, to prevent recurrence. Moreover, It's crucial to recognize that untreated Rapunzel syndrome may escalate, potentially leading to severe complications such as pancreatitis, which can be serious.

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