ISSN 2692-5877
DOI: 10.46998/IJCMCR.2021.10.000239

International Journal of
Clinical Studies & Medical Case Reports

Case Report

An Unusual Cause of Abdominal Pain and Iron Deficiency Anemia in a Three-Year-Old Girl. Rapunzel Syndrome the Missing Link

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Received: May 28, 2021 Published: June 11, 2021

Back ground

Bezoar is the accumulation of undigested foreign bodies or nutrients in the gastrointestinal tract. Several types of bezoar have been described including: hair (trichobezoar), fibers or seeds of vegetables and fruits (phytobezoar), or remnants of milk (lactobezoar), stones (lithobezoar), tablets (pharmacobezoar) and miscellaneous (fungus, sand, paper, etc.) [1,7].

Most cases of trichobezoar are reported in females, which may be attributed to the traditional long hair in females. [2]

Once the trichobezoar extends from the stomach into the jejunum or further on, it is referred to as "Rapunzel syndrome," which was first described in 1968 by Vaughan Jr. et al. [3].

The accumulated hair within the stomach and intestine is resistant to digestion, which occasionally presents as trichobezoar-induced bowel or gastric outlet obstruction [4].

Trichobezoar, is under-diagnosed entity and has to be considered in the differential diagnosis of iron deficiency anemia and abdominal pain in pediatric emergency department, especially if a history of PICA is reported. [5]

psychiatric referral after surgical treatment of a trichobezoar must be considered as an essential part of successful treatment and prevention of recurrence [6]

We hereby report the case of a female child presented with abdominal pain, constipation and iron deficiency anemia. She was found to have trichobezoars as reason for her symptoms. We highlight the difficulty of diagnosis without deep exploring of the clinical history.

Case Presentation

A three-year-old Caucasian girl presented with a two-weeks history of intermittent abdominal pain, abdominal distention, non-bilious vomiting, and constipation. She has been treated with laxatives by her GP without any discernible effects. She was subsequently admitted to the local hospital in view of reduced oral intake and weight loss.
On general examination, she looked pale and emaciated.

Abdominal examination revealed distention, mild tenderness, and irregular soft mass in the left lower quadrant of the abdomen. The rest of her systemic examination showed no abnormality.

The laboratory studies showed normal inflammatory markers, normal kidney and liver function with hypochromic microcytic anemia and low iron stores.

Abdominal X-ray demonstrated dilated bowel loops with no air-fluid level.
Emergency abdominal ultrasound was suggestive of intussusception.

She was referred to a paediatric surgery center where abdominal ultrasound was repeated and did not demonstrate any evidence of intussusception or obstruction.
The girl did not display any change in her symptoms, so the history was retrieved by her mother again, and it was identified that this girl had a history of eating hair, pieces of linen and tissue.

A new ultrasound study was repeated based on the new knowledge discovered from history which demonstrated a hyperechoic band with posterior acoustic shadowing in keeping with diagnosis of gastric trichobezoars with extension into the small bowel (Rapunzel syndrome).

A water-soluble contrast swallow and meal reported a filling defect extending from mid-gastric body to first part of duodenum confirmed the diagnosis.

She had a laparotomy and gastrostomy for removal of trichobezoar and recovered well post-operatively.

Outcome

She was discharged on iron supplement and referred to dietician and child mental health services.

Discussion

Trichobezoars one type of bezoars are concretions of swallowed hair retained within the digestive tract. Psychiatric ill-

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ness and mental health problems are commonly associated with trichobezoars [8-15].

Rapunzel syndrome is a rare form of trichobezoar that extends from the stomach into the small intestine.The name Rapunzel originates from the Brothers Grimm fairy tale of a 12-year-old princess locked inside a tower, who uses her long, golden hair to permit her young prince to climb up to her window and rescue her.

The disease often affects female children and adolescents who usually have a habit of hair pulling that is called trichotillomania with resultant patchy hair loss. Trichotillomania followed by swallowing the hair (trichophagia) is one type of pica, which is defined as 'the persistent craving and compulsive eating of non-food substances', such as hair, sponge, soap, sand and so on 22. The case we describe did not suffer any psychiatric illnesses or hair loss and this initially made the diagnosis challenging.

Patients with a trichobezoar may be asymptomatic, which can result in late presentation and, when left chronically undiagnosed, it can lead to severe anemia through either gastrointestinal bleeding or malabsorption [6].

Clinical findings of trichobezoars differ according to the settlement site. Whereas oesophageal bezoars lead to dysphagia, reflux and retrosternal pain, gastric bezoars present with abdominal pain, nausea, vomiting, loss of appetite, weight loss, ulcerations, and perforations. Intestinal bezoars lead to partial or complete obstruction or intestinal perforation. Colonic bezoars cause abdominal pain, constipation, and obstruction at the rectosigmoid level [12].
Our case presented initially with abdominal pain and constipation for two weeks which was thought to be functional and received laxatives without any effect. This shows the importance of investigating the causes of constipation before assuming it is functional, so serious pathologies are not missed.

On examination, a mobile, well-defined mass may be palpable in the epigastrium. This may be indentable on examination, Lamberton's sign, as quoted by O'Sullivan et al. [14]. In our case, there was soft palpable left abdominal mass which was thought to be faecal mass and after the initial ultrasound, it was suspected to be intussusception mass.

The most common complications that are reported include gastric mucosal erosion, ulceration and perforation of the stomach or small intestine, gastric outlet obstruction and intussusception.[10].

Iron-deficiency anemia is common in patients with trichobezoars, and could be the direct outcome of bleeding gastric ulcer due to pressure by massive trichobezoar [8].

Imaging is the mainstay of diagnosis and, therefore, both clinicians and radiologists need to have
a high level of suspicion to diagnose the condition early and prevent serious complications. [7] In our case the diagnosis was missed in the first setting due to lack of suspicion as it is a very rare cause for this presentation.
X-ray of the abdomen may show a radio-opaque mass in the stomach. Ultrasound reveals highly echogenic areas but is often not diagnostic 18. Our case had an x ray which showed dilated stomach without evidence of air fluid level, and ultrasound was done twice before the diagnosis was reached. On the first ultrasound scan, there was a suspicion of intussusception due to interpreting the appearance of the images as bowel in bowel. The second ultrasound scan was interpreted as normal.

Barium studies show barium in the stomach interspersed within the gastric mass. CT abdomen is often the investigation of choice in diagnosis and well-defined heterogeneous masses 9. The case we describe did not need CT abdomen as the diagnosis was strongly suspected after revisiting the clinical history and confirmed the presence of trichobezoar on contrast study. We can argue that detailed history of eating hair, linen and tissue has pointed towards the diagnosis and saved our patient unnecessary exposure to high radiation of CT abdomen.

Traditionally, a gastric trichobezoar was treated with laparotomy with either gastrotomy or enterotomy and this is still the typical treatment for large trichobezoars and Rapunzel Syndrome.

Laparotomy is the best method for treatment of trichobezoar. Retrospective study of 108 patients diagnosed with trichobezoar was performed by Gorter et al. The study concluded that the success rate of endoscopic treatment was 5%, while 75%for laparoscopic treatment. And 100% success rate was achieved by laparotomy [11]. The case we describe underwent laparotomy and gastrostomy with complete recovery.

In addition to the acute surgical treatment for a trichobezoar, psychiatric consultation is crucial to prevent relapses and to treat comorbid conditions that usually accompany this disorder [16-19]. The patient also received iron therapy for severe iron deficiency anaemia which has improved the pica.

Conclusion

1. Although rare, Rapunzel syndrome should be considered as a differential diagnosis in young patients who present with an iron deficiency anaemia, failure to thrive, abdominal mass and pain.
2. Professionals should be willing to re-evaluate the diagnosis and re-visit the history again as this was the clue to diagnosis after being missed. This will prevent missing serious conditions.

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