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Rapunzel Syndrome: Case Report

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Abstract **Case Report**

Rapunzel syndrome is a rare gastrointestinal pathology in the paediatric population, characterized by an extension of ingested intragastric hairs to an intestinal or even colonic extension. We report a case of gastroduodenal and intestinal trichobezoar in a 5-year-old girl, who consulted with an occlusive syndrome in whom the diagnosis of gastric and intestinal trichobezoar was diagnosed by ultrasound and confirmed intra-operatively. Our aim is to clarify the value of imaging in the diagnosis of this entity.

Keywords: Trichobezoar, Rapunzel syndrome, case report.

INTRODUCTION

First described by Vaughan *et al*., in 1968, it is an exceptional form of bezoar. It is described as the extension through the pylorus of a gastric bezoar which may extend into the caecum [1]. Various criteria have been used to describe it in the literature. Some define it as a gastric trichobezoar with a tail extending to the ileocaecal junction, some describe it as a simple trichobezoar with a long tail, which may extend to the jejunum or beyond, and some still define it as a bezoar of any size that can cause intestinal obstruction [2].

In reporting this recent observation, which is characteristic of this anatomical form and its mode of onset, we focus on the role of imaging in the diagnosis of this entity.

PATIENT AND METHODICAL

Patient information: This is a 5 and a half year old girl with a pathological history of trichophagia and textillophagia who presented to the emergency department with digestive symptoms such as intermittent abdominal pain associated with early vomiting of food which had been evolving for 12 days.

Clinical results: The child was in good general condition, apyretic, haemodynamically and respiratorily stable, with diffuse abdominal distension and tenderness with cessation of matter. On abdominal palpation, an epigastric mass was noted in the periumbilical region.

Diagnostic approach: The unprepared abdomen showed hydroaerobic levels, centred in relation to an acute intestinal obstruction. There was an associated dense calcium mass over the stomach (Figure 1). An abdominal ultrasound scan was requested, which revealed a curvilinear hyperechoic epigastric band with a posterior shadow cone (Figure 2) associated with two images of ileo-ileal invaginations in the right periumbilical region and on the left flank, centred by a hyperechoic band generating a shadow cone. This suggested a diagnosis of gastric trichobezoar extending into the small intestine, complicated by secondary intussusception (Figure 3).

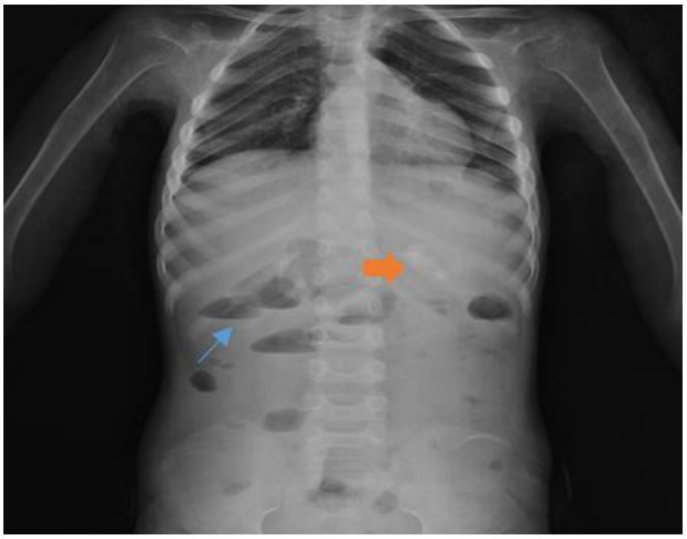

Figure 1: Showing hydro-aeric levels wider than high (blue arrow), centred with a stair-step appearance consistent with acute intestinal obstruction. There is a dense calcium mass over the stomach (orange arrow)

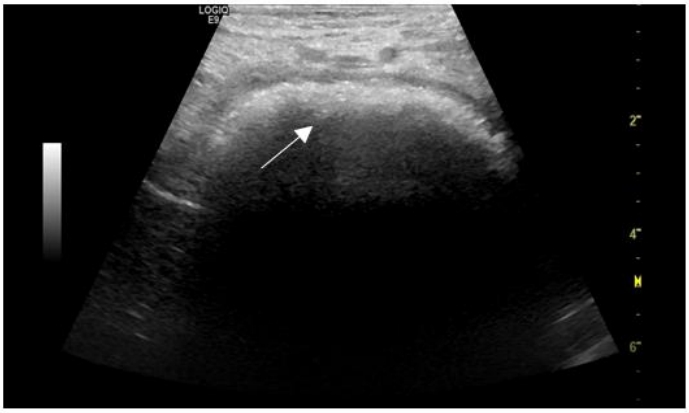

Figure 2: An arciform epigastric hyperechoic area generating a posterior shadow cone in relation to a trichobezoar (arrow)

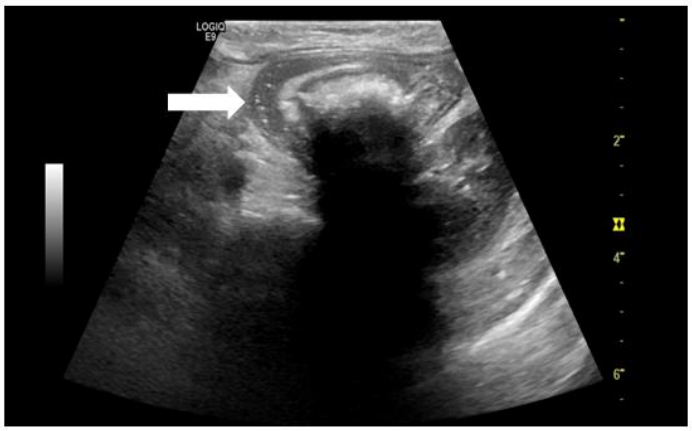

Figure 3: Image in cockade of an ileo-ileal invagination (arrow) centred on a hyperechoic band generating a shadow cone testifying to an invagination on a foreign body

Therapeutic Intervention: A median laparotomy was performed as an emergency and confirmed the diagnosis.

Patient's Perspective: The family stated that they were satisfied with the quality of the care provided.

Informed Consent: Consent was obtained from the patient's family.

DISCUSSION

Trichobezoar is a rare obstructive condition with an increased prevalence in older children aged 10-

19 years, females with an increased predisposition and a history of psychiatric illness such as trichotillomania (removing one's own hair) and trichophagia (ingesting hair) as a primary psychiatric disorder [3, 4]. Our observation may fit in with a patient with an eating disorder.

Rapunzel's syndrome is a rare progression of trichobezoar, because over time the accumulation of hairs will lead to the growth of trichobezoar beyond the stomach and into the small intestine [5, 6], as in the case of our patient with trichobezoar extending from the stomach into the small intestine.

Initially asymptomatic in the early stages, the mass increases and then leads to obstruction, vomiting, generalised abdominal pain and even an associated complication, as in the case of our patient who presented with an occlusive syndrome.

Imaging plays an essential role in the positive diagnosis. Abdominal X-rays can be used to assist in the diagnosis of gastric trichobezoar. Conventional findings are a mottled opacity of soft tissue filling the stomach, which is often distended and may show a calcified rim. However, in the literature review for this case report, no research supported abdominal radiography as an aid to the diagnosis of trichobezoar [7]. In our case the unprepared abdomen demonstrated intestinal-type hydro-aeric levels with a mass of calcific tone projecting from the stomach.

Ultrasound is often used as first-line imaging, and many authors have reported that ultrasound offers good sensitivity and specificity in the diagnosis of intestinal obstruction [7]. In a series reported by Ripollés *et al*., on 17 cases, ultrasound revealed signs of intestinal obstruction in 93% of patients with intestinal bezoars showing an echogenic curved band with a posterior acoustic shadow intra-intestinally and on the basis of this characteristic image, they suspected the presence of a bezoar before surgery in 15 (88%) of the 17 patients in the series [8].

Abdominal CT is the preferred modality for the diagnosis of trichobezoar, with an identified diagnostic accuracy of between 73% and 95%. The CT scan shows a well-demarcated mottled oval intraluminal mass. The mottled appearance of a trichobezoar on CT is due to hair mixed with trapped air and ingested food. It can be difficult to distinguish retained food from a bezoar, but a bezoar is usually oval/round and fills the gastric lumen with diffuse air bubbles throughout the mass [9]. Furthermore, according to some authors, the appearance of obstructive small bowel bezoars on CT scan is sufficient for diagnosis [10]. However, digestive endoscopy is considered the best diagnostic tool [11].

CONCLUSION

Rapunzel syndrome is a rare gastrointestinal disorder occurring in a paediatric population with an underlying psychiatric disorder. Patients present with a range of symptoms, from being asymptomatic to abdominal pain with a palpable abdominal mass, to occasionally developing complications. Ultrasound plays an essential role in the positive diagnosis of this condition.

Conflicts of Interest: No conflicts of interest.

Authors' Contributions: All the authors contributed to the diagnostic management of the patient and to the writing of this report.

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