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Radiological Case Report / Caso Clínico

An Unknown Abdominal Mass in the Pediatric Population – Rapunzel Syndrome

Uma Massa Abdominal Incaracterística na Idade Pediátrica – Síndrome de Rapunzel

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Abstract

Rapunzel syndrome is a rare condition, a term also used for a trichobezoar that presents a tail to the duodenum and/or jejunum through the pylorus. This is caused by a "hairy ball" within the stomach, leading to gastric outlet obstruction. We present a case of an 11-year old child with abdominal pain and vomiting episodes, with an abdominal mass depicted at physical examination, whose imaging findings were compatible with this condition, posteriorly with surgical confirmation.

Keywords

Rapunzel syndrome; Trichobezoar; Gastric obstruction.

Resumo

O síndrome de Rapunzel é uma condição rara, resultante de um tricobezoar que se prolonga para o duodeno e/ou jejuno através do piloro. Este é condicionado pela presença de uma "bola de cabelo" ocupando o lúmen gástrico, sendo responsável por obstrução do esvaziamento gástrico. Apresentamos um caso de uma criança de 11 anos com dor abdominal e vómitos, com evidência de uma massa abdominal ao exame clínico, tendo-se documentado achados radiológicos compatíveis com esta condição, posteriormente confirmados na exploração cirúrgica.

Palavras-chave

Síndrome de Rapunzel; Tricobezoar; Obstrução gástrica.

Introduction

Rapunzel syndrome is a term also used for a trichobezoar and it constitutesa rare entity.
It occurs more frequently in adolescent girls, often with history of a psychiatric illness or of a previous surgery. The authors present a clinical case of this disease, including imaging and histological findings, reviewing its main characteristics.

Clinical History

An 11-year old female patient was admitted at the Emergency Room of a Hospital Unit with abdominal pain and distention, accompanied by vomiting episodes. She had no relevant previous clinical history. At physical examination there was a significant abdominal distention with a palpable upper abdominal mass. Laboratory results were mainly unremarkable, with the exception of a slight neutrophilia. At first approach, an abdominal radiography was requested, revealing gaseous distention of some small bowel loops in the left lumbar and iliac region, with evidence of some air-fluid levels. Additionally, at detailed evaluation, one could perceive the presence of heterogeneous content occupying the epigastric and right lumbar area with a mottled gas appearance (Figure 1). To better characterize these findings, an ultrasound was performed, which was somewhat difficult due to the significant abdominal gas distention. It demonstrated a significant gastric distention and also a large upper abdominal mass of about 12cm, causing an intense shadowing effect, which was difficult to access if it were endoluminal or intraperitoneal. In addition, liquid distention of some small bowel loops was demonstrated at the left lumbar region (Figure 2). As the patient continued to present vomiting episodes and signs of intestinal sub-occlusion, an abdominal and pelvic CT was performed after IV contrast. It confirmed massive gastric distention caused by the presence of a reticular, non-enhancing mottled gas mass, occupying the whole gastric lumen, protruding into the proximal duodenum, being the reason of the obstructive symptoms. Some thickening, edema and enhancement of the gastric wall was seen as a sign of congestion. A similar and smaller lesion was depicted at the left iliac region, this one being the cause of the distention of the small bowel loops in the left lower quadrant (Figure 3). Considering the size of the mass, the endoscopic approach was not attempted, and a minimal invasive surgery (gastrostomy and enterotomy) was performed to remove both masses, the one inside the stomach which was protruding into the first portion of the duodenum and the other one inside a small bowel loop, 40cm away from Treitz angle, causing the proximal small bowel occlusion. Surgery revealed its hairy nature.
During the stay at the Emergency Department, the mother was interrogated about the girl's behavior and she then confirmed that the patient, for the past three years, had the habit of eating not only hair, but also finger nails, remote control buttons and plastic bags. After recovery, the patient was finally provided with psychiatric guidance.

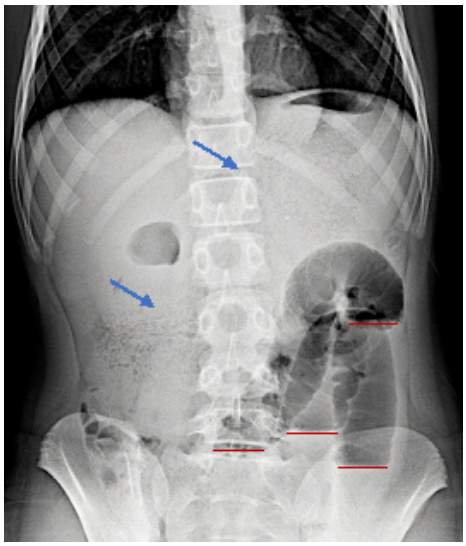

Fig. 1 – Abdominal radiography, posteroanterior projection, demonstrating small bowel distention in the left lower quadrant with some air-fluid levels (red lines). One could also depict gastric distention with a mottled air appearance (blue arrows).

of gastric surgery or a mental illness accompanied by the habit of eating hair, called trichotillomania. One per cent of patients with trichophagia will develop a trichobezoar. Hair is indigestible because of a slippery surface, allowing it to escape peristaltic propulsions and accumulate in concretions between the gastric folds.[2] As it grows bigger, it turns into a mass adjusting to the shape of the stomach. Rapunzel syndrome is a rare form of a trichobezoar, presenting an elongation through the pylorus into the duodenum, jejunum or even ileum. Patients can remain asymptomatic for a long period of time where symptoms are usually unspecific: nausea, abdominal pain, post-prandial fullness, dyspepsia, loss of appetite, vomiting or obstruction. Gastric ulceration, perforation, gastric emphysema, peritonitis, intussusception, obstructive jaundice, pancreatitis or malabsorption syndromes can show as complications.[1,2] Gastrointestinal occlusion can occur in 10% and gastrointestinal bleeding in about 6%.[3] Physical examination can depict a patchy alopecia, halitosis and abdominal non-tender mass.[1]

Diagnostic evaluation can include an abdominal radiography, fluoroscopy, ultrasound and/or CT.[2,3]

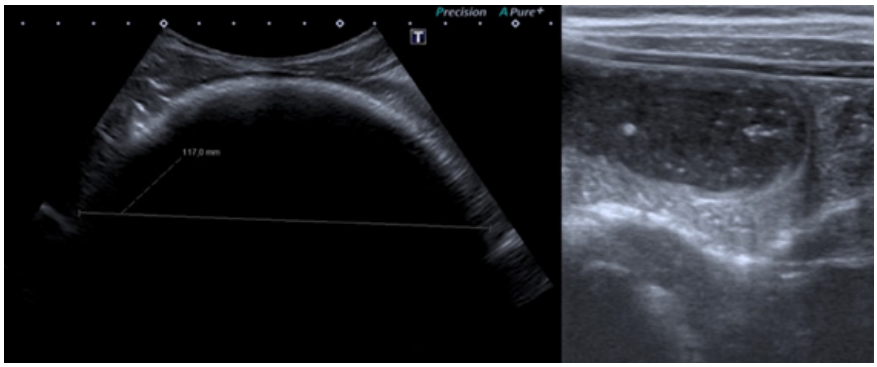

Fig. 2 – Ultrasound images. On the left side demonstrating large upper abdominal mass, with a hyperechoic band causing a sharp posterior attenuation. On the right side there was confirmation of liquid distention of small bowel loops in the left lower quadrant.

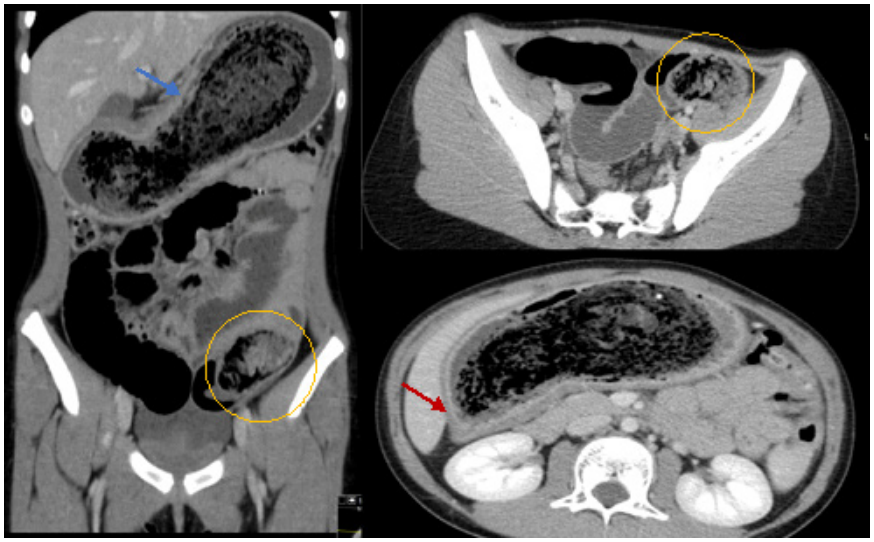

Fig. 3 – Computerized tomography (CT) images, on the left on coronal plane and on axial plane on the right side. CT confirmed the presence of a massive mottled gas mass occupying the entire lumen of the stomach (blue arrow), insinuating to the first portion of the duodenum (red arrow). Another similar and smaller mass was depicted inside a jejunal loop in the left lower quadrant (orange circle), which was the cause of the small bowel obstruction and air-fluid levels on radiography.

Discussion

Bezoars consist of foreign ingested materials that accumulate in the gastrointestinal track, classified according to its components/original materials and are usually located inside the stomach.[1,2,4] When composed by hair bezoars are called trichobezoar, which is the second most common type of bezoar,[2] and patients often have a previous history

- Abdominal radiography: distended "J"-shaped stomach, with a mottled gas appearance with surrounding lucency and some air-fluid levels in the abdomen.
- Fluoroscopy: endoluminal gastric filling defect with no wall affiliation with a lacelike pattern.
- Ultrasound: hyperechoic curvilinear dense strip with sharp acoustic shadowing with no through transmission. It maintains its appearance regardless of probe angulation or water intake.

- CT: presence of a large mass within the gastric lumen, with a heterogeneous appearance with entrapped air and food debris. The lesion extends through the pylorus to the duodenum/jejunum/ileum, consisting of a concentric compressed ring. Stomach and sometimes small bowel dilatation with or without air-fluid levels.

Diagnostic differentials include pancreatic pseudocyst, gastrointestinal stromal tumors (GIST) or phytobezoar (bezoar composed of indigestible food materials).

Management can be endoscopic or surgical, usually considering the size of the mass. Endoscopic approach is particularly difficult for trichobezoar, especially when it is large (>20cm), although some medical devices such as bezotomos or bezotriptors can be a useful tool. Quite often patients with Rapunzel syndrome undergo minimally invasive surgery for the mass removal (gastrostomy/enterotomy).[1] Long term prognosis is excellent as long as the patients maintain behavioral therapy with psychiatric follow-up.

In summary, Rapunzel syndrome is an underdiagnosed condition that should be taken into account as a diagnostic hypothesis in the pediatric population, especially in the presence of an abdominal mass with obstructive symptoms. Radiologists should be acquainted with this condition for proper patient management, which, in this case, was surgical.

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Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.
Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).
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