Journal Homepage: -www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI:10.21474/IJAR01/16395
DOI URL: http://dx.doi.org/10.21474/IJAR01/16395

RESEARCH ARTICLE

RAPUNZEL SYNDROME COMPLICATED BY ACUTE INTUSSUSCEPTION: ABOUT TWO CASES

Abide Zakaria*, Andour Hajar, Cherraqi Amine, ALL Ali Nazik, CHAT Latifa, ELHADDAD Siham
Departement of radiology, Mother and Child Hospital, CHU Ibn-Sina, Bp 6527, Rue LamfadelCherkaoui Rabat Morocco.

Manuscript Info

Manuscript History
Received: 05 January 2023
Final Accepted: 09 February 2023
Published: March 2023

Key words:
Rapunzel's Syndrome, Trichobezoar, Acute Intussusception, Standard Radiography, Ultrasound, Computed Tomography

Abstract

Rapunzel syndrome is a rare form of gastric Trichobezoar extended to the intestine. There is little literature describing the complications associated with this condition. The diagnosis can be guided by the results of conventional radiography and ultrasound, but computed tomography (CT) is the gold standard, as it confirms the diagnosis and provides crucial information on its severity.

Introduction:-

Trichobezoar is a mass of hair and fiber trapped in the gastrointestinal tract.

Rapunzel syndrome is a rare form characterized by extension of the tail into the small intestine. The first case was reported by Vaughan in 1968. (1)

Occlusion, peritonitis, and hemorrhage are the most serious complications reported. (2)

The diagnosis is based on the findings of conventional radiography, ultrasonography, and computed tomography.

Through these two case reports, we aim to outline the radiological semiology of Rapunzel syndrome (RS).

First Case Report

A 4-year-old girl with celiac disease, iron deficiency anemia, and a history of geophagia, which was accentuated during a maternity stay for a new pregnancy, presented to the emergency room with intermittent abdominal pain and vomiting.

Clinical examination revealed a distended abdomen, deformed by a hard and tender epigastric mass.

Radiography (figure 1) showed an opacity centered on the epigastric region, pushing back the digestive structures with clear greensacral distension without hydro-aeric levels.

Abdominal ultrasound (figure 2) is in favor of a strongly attenuating hyperechoic arciform epigastric image hiding the structures behind, making evoke a Trichobezoar given the clinical and anamnestic context associated with a hydric and hydrocholecystic distension.

Corresponding Author:- Abide Zakaria
Address:- Departement of radiology, Mother and Child Hospital, CHU Ibn-Sina, Bp 6527, Rue LamfadelCherkaoui Rabat Morocco.

The CT scan (figures 3,4,5) showed a hypodense and heterogeneous mass containing air bubbles, molding the gastric wall and taking the shape of the stomach and extending to the duodeno-jejunal level.

It is associated with duodeno-jejunal invagination (figure 6) with distension of the small intestines upstream of a non-stenosing parietal thickening.

Surgical exploration found a large gastric bezoar reaching the jejunum (figure 7). The patient underwent gastrostomy and the bezoar was extracted.

Second Case Report:
A 5-and-a-half-year-old girl with a history of trichophagia, seen in consultation for abdominal pain for 1 month and in whom the clinical examination found a palpable mass at the epigastric level.

Ultrasound exploration showed the presence of an echogenic curvilinear mobile shadow in the stomach with a dense posterior acoustic shadow.

CT scan (Figure 8, 9, 10) objective the presence of a diffuse intra gastric mass extending from the bottom of the stomach to jejunum. The lesion appears heterogeneous with entrapped air and food debris. There is an associated cocardial image on the left flank in relation to a jejuno-jejunal invagination coil.

The patient underwent an exploratory laparotomy. Findings included gastric distension, a palpable mass extending from the gastric lumen to the first section of the duodenum, and jejunojejunal intussusception, which was released by manual revision (figure 11).

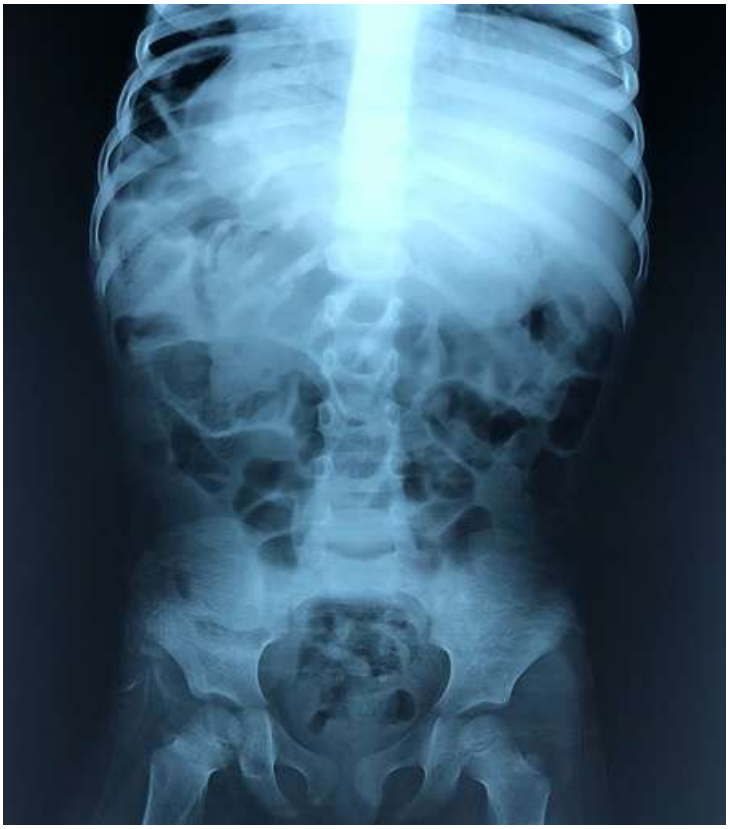


Figure 1:- Opacity centered on the epigastric region, pushing back the digestive structures with a clear grelo-colic distension without hydro-aeric levels.

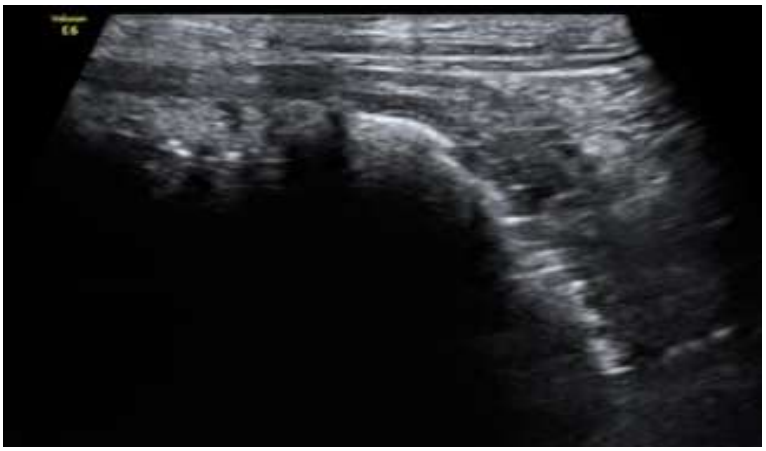
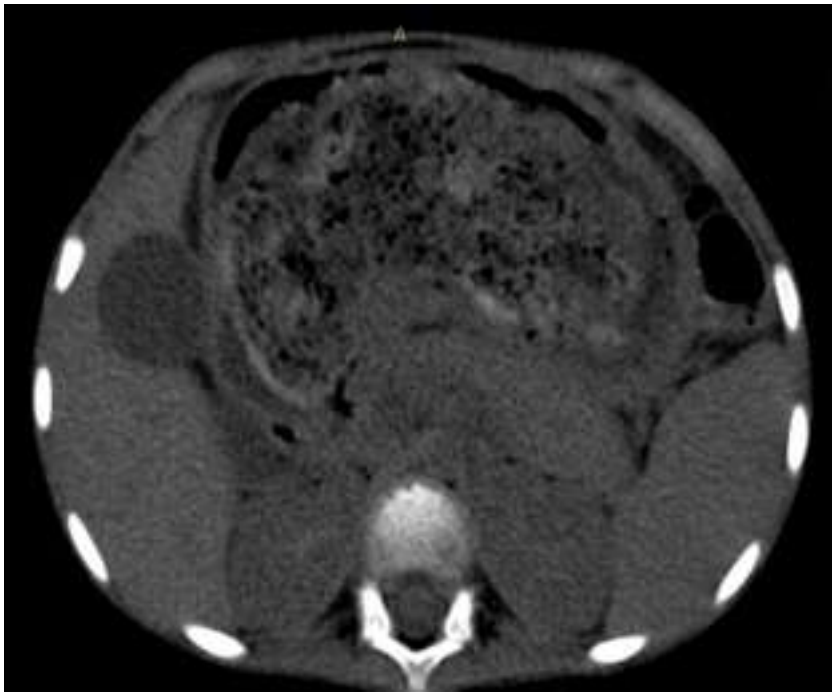


Figure 2:- Abdominal ultrasound showing a curvilinear echogenic moving shadow in the stomach with dense posterior acoustic shadow.



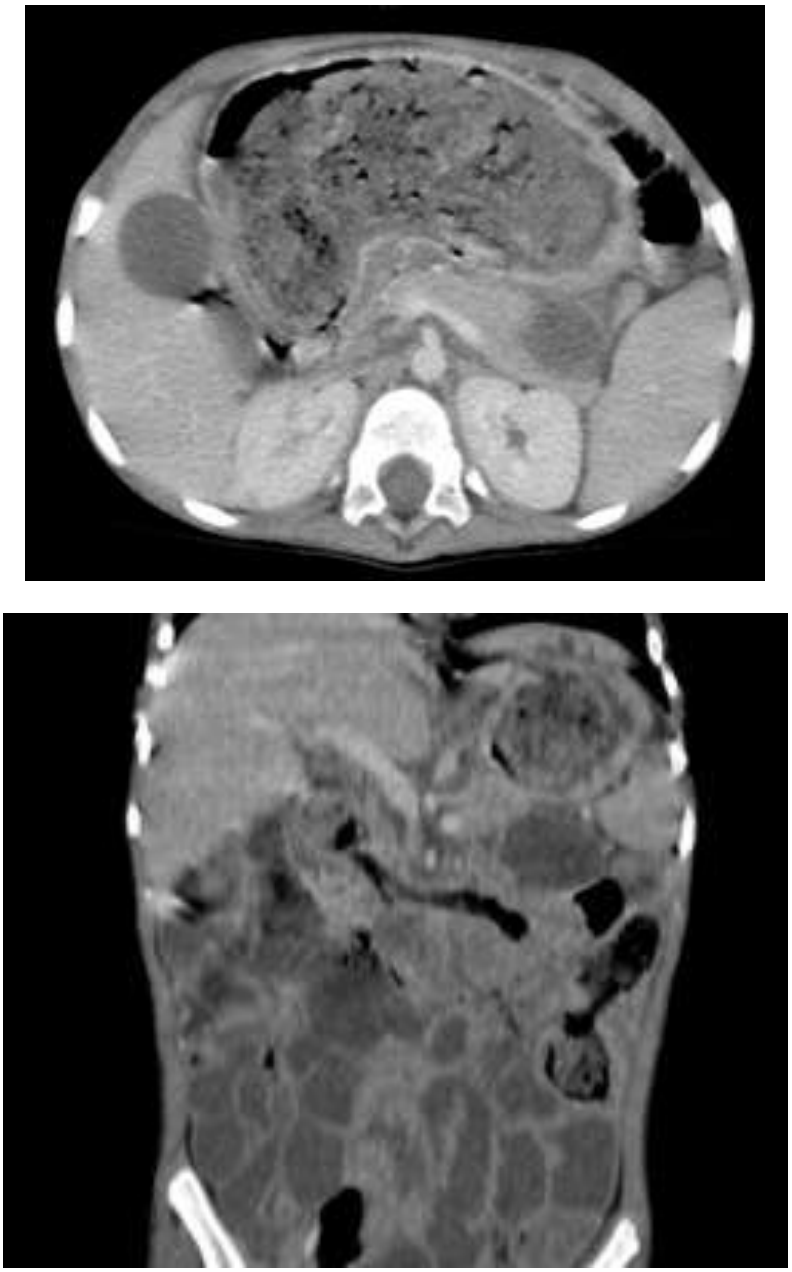


Figure 3, 4, 5:- Axial (a) without injection and axial (b) and coronal (c) CT sections with injection of contrast product: heterogeneous gastric endoluminal mass extending to the jejunum, containing air bubbles similar to the digestive contents.

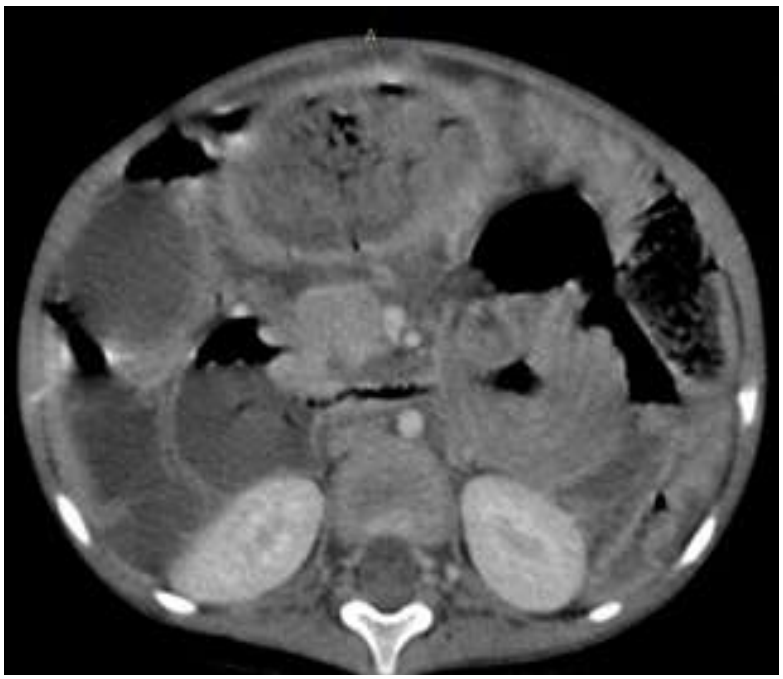


Figure 6:- Image in cocarde in the left flank made of superimposed layers of the digestive walls in relation to a duodeno-jejunal invagination coil.

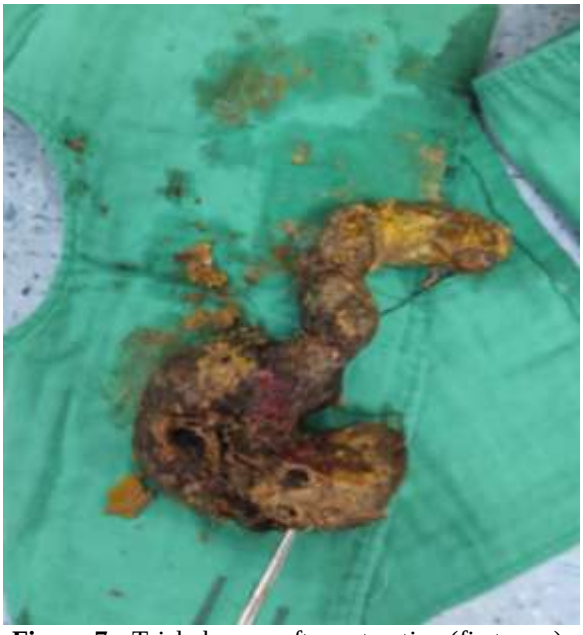
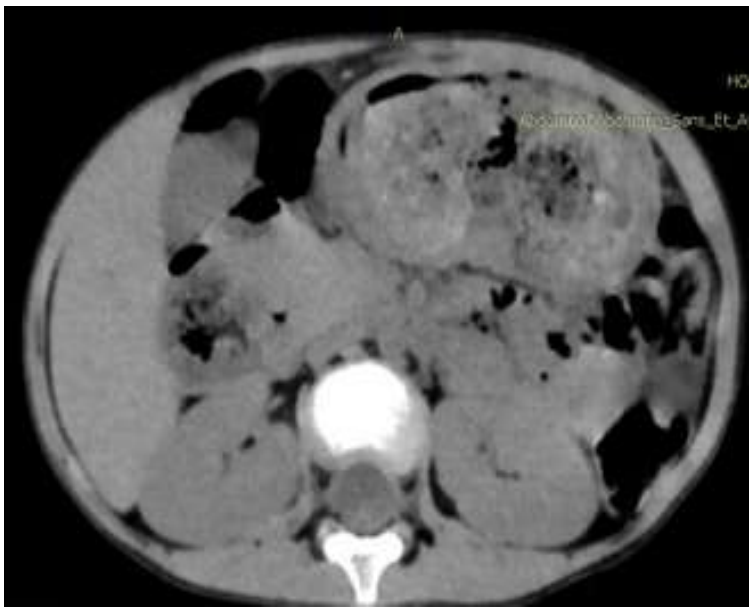
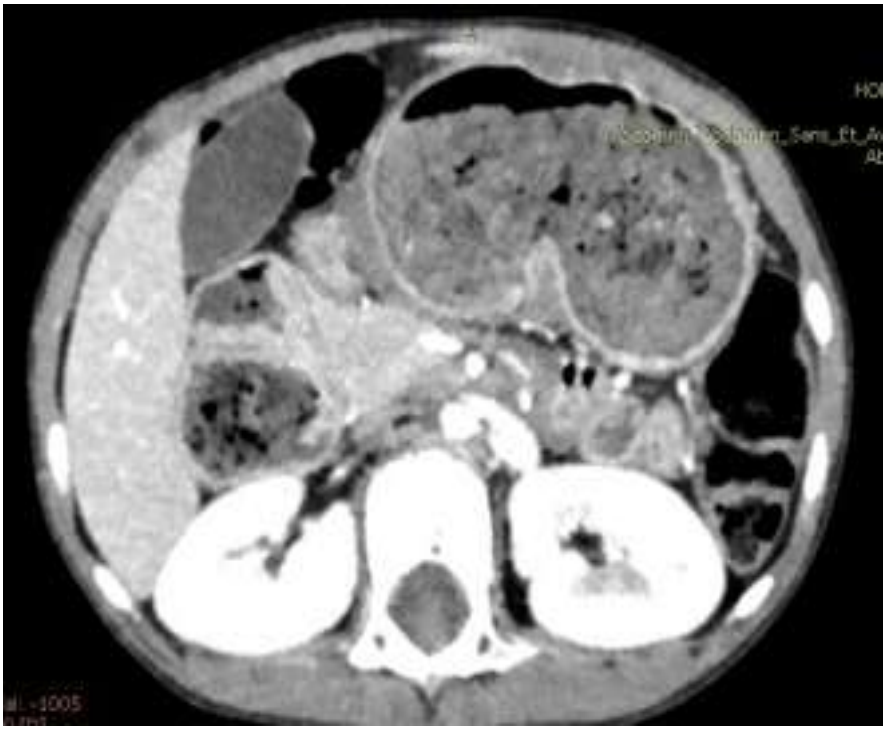


Figure 7:- Trichobezoar after extraction (first case).





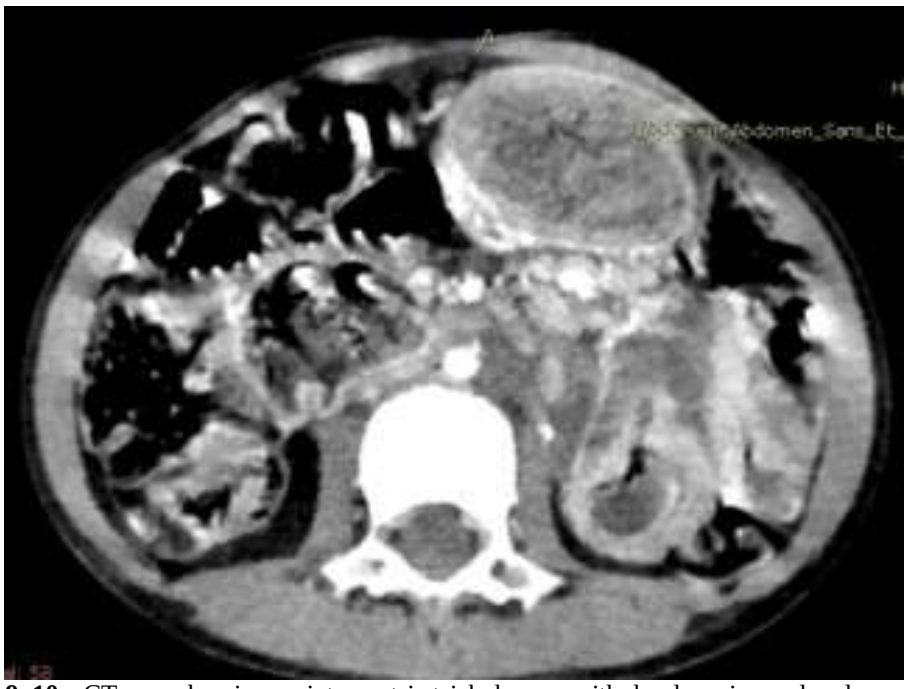


Figure 8, 9, 10:- CT scan showing an intragastric trichobezoar with duodeno-jenunal and presence of an invagination bladder on the left flank.

Figure 11:- Trichobezoar after extraction (second case).

Discussion:-

Rapunzel syndrome is a rare form of Trichobezoar, which gets its name from its resemblance to a tail. (3)

It is well known that trichophagia is the most common condition associated with Rapunzel syndrome, but various studies have reported other factors: history of gastrotomy or vagotomy, gastroparesis, and eating habits. (1, 2)

The revealing symptoms are not very specific: abdominal pain, anorexia, weight loss, epigastric satiety aggravated during meals, vomiting, peptic esophagitis, diarrhea or constipation (4). It can be revealed by complications such as peritonitis (18.3%) or by an occlusive syndrome, with a prevalence of 25.9% of all complications (5).

On plain abdominal radiography, bezoar can be visualized as a mottled epigastric mass similar in appearance to a stomach filled with food.

Sonographically, bezoar presents as a mass of increased echogenicity in the stomach area, with complete loss of posterior echoes. (6)

CT is the gold standard for diagnosis of the syndrome. The bezoar forms a heterogeneous, mobile, intraluminal mass without any parietal attachment; sometimes showing concentric rings surrounded by the contrast medium. The marbled gas pattern is pathognomonic of bezoar. The presence of air and food is responsible for the density mixing of the mass.

CT also provides crucial information about severity by revealing intestinal ischemic signs. (7)

Small Trichobezoars can be removed endoscopically. Endoscopic removal is difficult and risky, esophageal perforation may occur.

The exact location of the obstruction must be analyzed because it conditions the surgical approach: a proximal bezoar is a formal indication for gastrotomy or enterotomy. The closer the obstruction is to the ileo-caecal junction the less invasive the attitude would be(8).

Conclusion:-

Diagnosis of Rapunzel syndrome is only possible on imaging. Ultrasound and radiography can guide the diagnosis, but CT scan confirms it. It also gives precise information about extension and complications.

References:-

[1]Delabrousse E, Brunelle S, Saguet O, Destrumelle N, Landecy G, Kastler B. **Small bowel obstruction secondary to phytobézoard in a child: CT Diagnosis.**AJR 2001;177:255-6.

[2] Rubin M, Shimonov M, Grief F, Rotestein Z, Lelcuk S, **Phytobézoar: a rare cause of intestinal obstruction.**Dig Surg 1998; 15:52-4.

[3]Gonuguntla V, Joshi DD. **Rapunzel syndrome: a comprehensive review of an unusual case of trichobezoar.** Clin Med Res. 2009;7(3):99–102.

[4]Balik E, Ulman I, Taneli C, Demircan M. **The Rapunzel syndrome: a case report and review of the literature.**Eur J Pediatr Surg 1993;3: 171-3.

[5] Vaughan ED Jr, Sawyers JL, Scott HW Jr. **The Rapunzel syndrome. An unusual complication of intestinal bezoar.**Surgery 1968; 63: 33943.

[6] Newman. B, Girdany.BR. **Gastric trichobezoars- sonographic and computed tomographic appearance.** PediatrRadiol. 1990; 20: 526-527.

[7]Billaud Y, Pilleul F, Valette PJ. **Mechanical small bowel obstrcution due to bézoard: correlation between CT and surgery findings.** J Radiol 2002 ;83 :641-6

[8]Jeyasingh S, Sundari M, Kannan VV. **Trichobezoar—two case reports.** TEJMS. 2011;2:41-42.