Available online at www.elixirpublishers.com (Elixir International Journal)

Radiology

Elixir Radiology 128 (2019) 52769-52770

ISSN: 2229-712X

Rapunzel's Syndrome: A Bezoar Like No Other

Omar El Aoufir, Ittimade Nassar and Nabil Moatassim Billah
Central Radiology Department, University Hospital Center Ibn Sina, Rabat.

ARTICLE INFO

Article history:
Received: 21 January 2019;
Received in revised form:
20 February 2019;
Accepted: 28 February 2019;

Keywords
Trichobezoar,
Rapunzel's Syndrome,
Computer Tomography.

ABSTRACT
Rapunzel's syndrome is a rare form of gastric trichobezoar extended to the bowel. The diagnosis may be oriented by the findings of the conventional radiography and echography, but the computed tomography remains the gold standard as it confirms the diagnosis and provides crucial informations on it's gravity

Introduction

The trichobezoar is a mass made of hair and fibbers trapped in the gastrointestinal system.

Rapunzel's syndrome is a rare form characterized by the extension of the tail to the small bowel. The first case was reported by Vaughan in 1968 [1].

Occlusion, peritonitis and digestive hemorrhage or perforations are the most severe complications reported [2].

The diagnosis rests on the findings of the conventional radiography, echography and computed tomography (CT).

Through this case report, we aim to expose the radiologic semiology of Rapunzel's syndrome (RS).

Case Report

A 16 years old female, with a known history of iron deficiency anemia and a trichophagia, presented to the hospital with an occlusive syndrome.

The clinical examination revealed a distended abdomen, deformed by an epigastric hard sensitive mass.

The Xray (Figure 1) showed a central opacity, highlighting the gastric curvature and repelling the bowel to the left, without air-fluid levels.

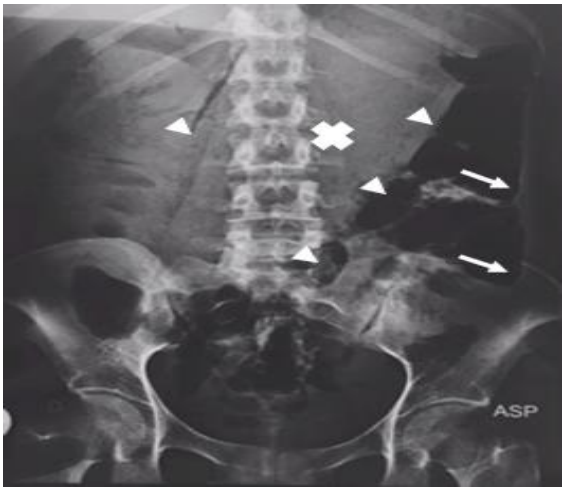

Figure 1. Plain abdominal film showing a central abdominal opacity (X), a thin gas line that marks out the gastric curvature (➢). Intestines are repelled (➔).

The CT (Figure 2, 3) brought the light on a structure filling practically the entire gastric lumen, expanding to the duodenum and the duodenojejunal junction.

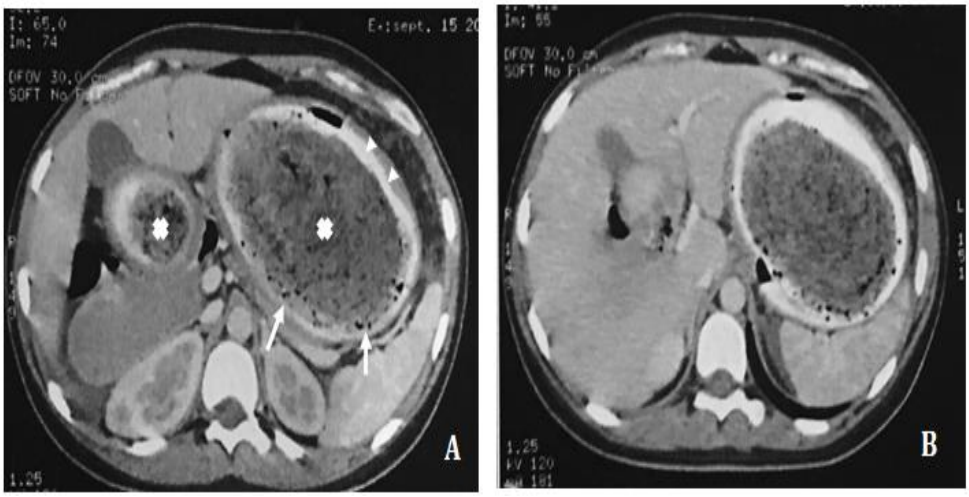

Figure 2 A, B. Axial computerized tomography with Injection of a contrast agent. Heterogeneous mass in the gastric lumen and duodenum (X), containing air ➔ molded by the gastrographine (➢) and without any parietal attachment.

Tele: +212672262306
E-mail address: omarelaoufir@gmail.com

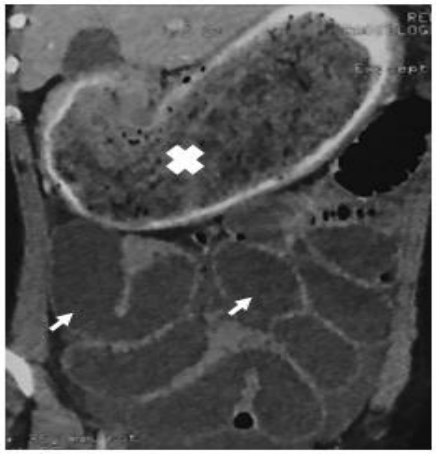

Figure 3. Coronal opacified computerized tomography: Endoluminal heterogeneous gastric mass X extended to the duodenum with an important distention of the small bowel (→).

Figure 4. Foreign materiel extracted surgically. It is made of compacted hair.

It had an ovoid shape, a mixed density, heterogeneous content, composed of concentric rings molded by the contrast agent. It had no parietal attachments. We also noted an important distention of the small bowel.

The surgical exploration (Figure 4) found a wide gastric bezoar reaching the jejunum. The patient underwent a gastrostomy and the bezoar was extracted.

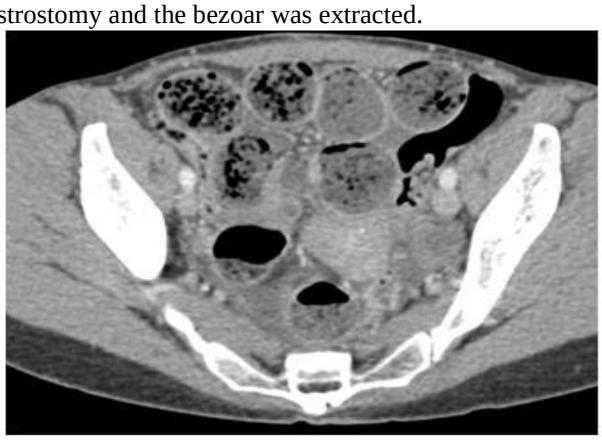

Figure 5. Axial computerized tomography: intestinal occlusion on a bow. Note the small bowel feaces with a mixed density image in the small bowel, the less compacted aspect and the wider extension (jejunum and ileus), at the opposite of the bezoar, which rarely exceeds the proximal jejunal portion

Discussion

Rapuzel's syndrome is an extremely rare form of gastric trichobezoar with a duodenal and jejunal extension. It mainly affects young women with eating disorders.

It is well known that trichophagia is the most frequent condition associated with RS, yet, different studies reported other factors: history of gastrotomy or vagotomy, gastroparesis and eating habits [1,2].

The clinical presentation depends on the size of the bezoar. In most cases, it is asymptomatic; nevertheless, it can be revealed by complications such as peritonitis (18.3%) or by an occlusive syndrome, as it was the case for our patient, with a prevalence of 25.9% of all the complications [3].

Typically, the plain abdominal film shows a prominent gastric outline with an intragastric mottled mass, outlined by gas in the distended stomach (figure 1). In the delayed forms, calcifications can be observed [4].

Sonographically, the bezoar presents as an increased echogenicity mass in the stomach region, with complete loss of posterior echoes [4].

The CT is the gold standard for the diagnosis of the syndrome. The bezoar forms a heterogenic intraluminal mass, mobile, without any parietal attach; sometimes presenting concentric rings surrounded by the contrast agent. The mottled gas pattern is pathognomonic of bezoar. The presence of air and food are responsible of the mixture of density of the mass. The gastric location is a fundamental sign of orientation of Rapunzel's syndrome.

It also provides crucial informations on the gravity by revealing ischemic bowel signs.

The exact location of the obstacle must be analyzed as it conditions the surgical approach: A proximal bezoar is a formal indication to a gastrotomy or an enterotomy.

The more the obstruction is closer to the ileo coecal junction; the attitude would be less invasive [5].

The essential CT differential diagnosis is faeces sign [6] (figure 5), as it has the same nature; it reflects a decline in the intestinal transit.

The CT diagnosis sensitivity can be improved by using larger scanning windows, looking for gas patterns in the bezoar [5]. A window between -100 et +500UH provides 97% sensitivity [6].

Conclusion

It is clear that imagery is the key of the management of Rapuzel's syndrome. Sonography and x ray may orientate the diagnosis, but the computed tomography confirms it. It also gives precise informations on the extension and the 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. Vaughan ED Jr, Sawyers JL, Scott HW Jr. The Rapunzel syndrome. An unusual complication of intestinal bezoar. Surgery 1968; 63: 33943.
4. Newman. B, Girdany.BR. Gastric trichobezoars-sonographic and computed tomographic appearance. Pediatr Radiol. 1990; 20: 526-527.
5. 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
6. Catalino O. The faeces sign: a CT finding in small bowel obstruction. Radiology 1997; 37: 417-9.