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Rapunzel Syndrome: A Rare Case Report

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Abstract **Case Report**

Bezoards are concretions of human fibres that accumulate in the digestive tract. Trichobezoards are often related to underlying psychiatric disorders such as depression and eating behavior disorder. Teatment is often medical or endoscopic while surgery is required for extended forms. We present the only documented case of Rapunzel syndrome in Morocco.

Keywords: Trichobezoard, Rapunzel syndrome, Mallory-weiss syndrome, Gastrotomy.

INTRODUCION

Rapunzel syndrome, first described in 1968, is a rare entity, defined as gastric trichobezoar with duodenal or jejunal extension [1]. They are often related to psychiatric disorders. Complications are common and may be the mode of disclosure. The upper digestive endoscopy is the reference examination with diagnostic and therapeutic interest.

CASE REPORT

A 12-year-old girl was admitted to the emergency department for hematemesis of moderate severity with epigastric pain that had been evolving for more than five months. The interrogation found a notion of trichomania and trichophagia with iterative postprandial vomiting. The physical examination revealed a patient who was apyretic, dehydrated, and malnourished with diffuse cutaneous-mucosal pallor and slightly discolored conjunctivae with the presence of a solid and mobile epigastric mass. The examination also showed multifocal alopecia.

After hydroelectrolytic resuscitation with blood transfusion, an oesogastroduodenal fibroscopy was performed, showing a laceration of the oesophageal mucosa and a fragile and erythematous gastric mucosa with the presence of a ball of hair and whitish material occupying the entire gastric cavity, extending into the pylorus and into the duodenum.

The rapunzel syndrome was confirmed by the upper digestive endoscopy which also specified the origin of the hematemesis in relation to the mallory-weiss syndrome secondary to the vomiting efforts associated with the gastritis induced by the trichobezoar. The endoscopic extraction was difficult and the patient benefited from a longitudinal gastrotomy (image 1 and 2) with simple postoperative and psychiatric follow-up.

Image 1: Gastrotomy for extraction of the trichobezoar and its duodenal extension

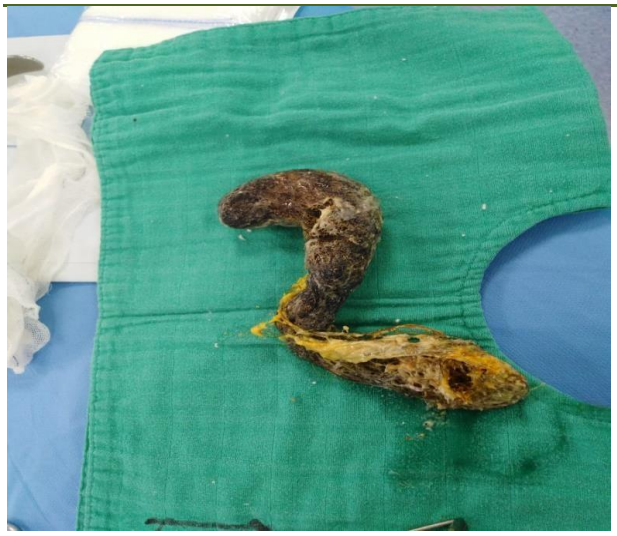

Image 2: Trichobezoar covered with whitish material and food debris

DISCUSSION

Rapunzel syndrome, first described in 1968, is a rare entity, defined as gastric trichobezoar with duodenal or jejunal extension [1]. Indeed, trichobezoar is a rare condition, common in females (90% of cases) with a peak incidence between 10 and 19 years of age, representing 0.15% of gastrointestinal foreign bodies [2]. Trichobezoar is often associated with underlying psychiatric disorders, particularly depression, mental retardation, and behavioral and eating disorders [3].

Trichobezoar can remain asymptomatic for a long time, thus explaining the delay in diagnosis. The symptoms develop gradually and insidiously as the mass becomes larger. They are often intermittent and usually vague and non- specific. They are primarily digestive in nature. Complications are frequent and may be the mode of revelation. They may be mechanical (occlusion, intussusception, volvulus, appendicitis), traumatic (ulcers, gastritis, pancreatitis) or related to malabsorption syndrome (anemia, hypoprotidemia) [4].

The upper digestive endoscopy is the reference examination with diagnostic (allowing visualization of a process composed of chevron hairs and food debris) and therapeutic (endoscopic extraction of small trichobezoard) interest. CT is considered an imaging technique of choice for confirmation of gastrointestinal bezoar. It can demonstrate a well- limited oval intraluminal mass occupying almost the entire gastric lumen and consisting of a multitude of concentric circles of different densities distributed in onion bulb [4].

Treatment can be medical, based on copious drinks associated with transit gas pedals, instrumental by extraction or fragmentation (endoscopic, by laser or extracorporeal lithotripsy) or surgical for extensive forms [2]. The prognosis is generally good. It is linked to a better knowledge of the clinical and radiological signs with the search for the primary anamnestic elements which are trichophagia and trichotillomania. Recurrences are possible as a consequence of non-adherence to psychiatric treatment [5].

CONCLUSION

Rapunzel syndrome is a rare condition, often related to psychiatric disorders, and can remain asymptomatic for a long time. Complications are frequent and can be the mode of revelation. The upper digestive endoscopy is the reference examination with diagnostic and therapeutic interest. Recurrences are possible as a consequence of non-adherence to psychiatric treatment.

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