Rapunzel syndrome: a rare cause of recurrent abdominal pain in 8 year-old child.

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ABSTRACT

Trichobezoar or Rapunzel syndrome is a rare condition in childhood, secondary to unusual aggregation of hair and indigestible fibers inside the digestive tract and especially in the stomach leading to a solid mass. It affects mainly young women with history of trichotillomania and trichophagia. We report an unusual case of gastric trichobezoar in 8year-old female with history of recurrent abdominal pain and unclear history of trichophagia. The treatment was surgical after failure of endoscopic extraction. A psychological follow-up was initiated and the outcome was favorable.

Keys word : Trichobezoar; Trichophagia; Children; Recurrent abdominal pain; Endoscopy; Gastrotomy.

INTRODUCTION :

Baudamant, a French doctor, was the first to describe a case of human trichobezoars in 1779 [1]. Trichobezoars are compact "cluster" of hair and indigestible organic fiber which agglomerate and remain in the gastrointestinal tract for long time. Like fishing net, the recurrent ingestion of hair and indigestible material enlarges the hairball. It's a rare condition that affects mainly young women. Trichobezoar in children are almost exclusively reported as isolated cases in the literature [2]. We report a case of a child with unclear psychological disorder admitted for recurrent abdominal pain due to huge gastric trichobezoar.

CASE REPORT :

An 8 year-old female referred to our pediatric gastroenterology out-patient clinic for recurrent abdominal pain and weight loss. The child suffered from gastric fullness and discomfort, frequent postprandial non-bilious vomiting and bad smell mainly in the morning for the last two years. She received several symptomatic treatments without any improvement. On examination, her weight was 21.5 kg (-1.8 DS) and her height was 121 cm (-0.9 DS). The respiratory rate and the blood pressure were within normal ranges. No pallor or jaundice. A marked halitosis smelt on throat examination. The abdomen was lax soft with mild epigastric tenderness. No abdominal mass was palpable. Biochemical studies showed normal levels for hemoglobin (12.1 mg/dl), glucose (5.2 mmol/1), blood urea nitrogen, serum creatinine, and transaminases. Celiac serology was negative.Upper gastrointestinal endoscopy showed a large trichobezoar occupying almost the whole gastric cavity extending from the cardia to the pylorus and duodenum (Figure1)

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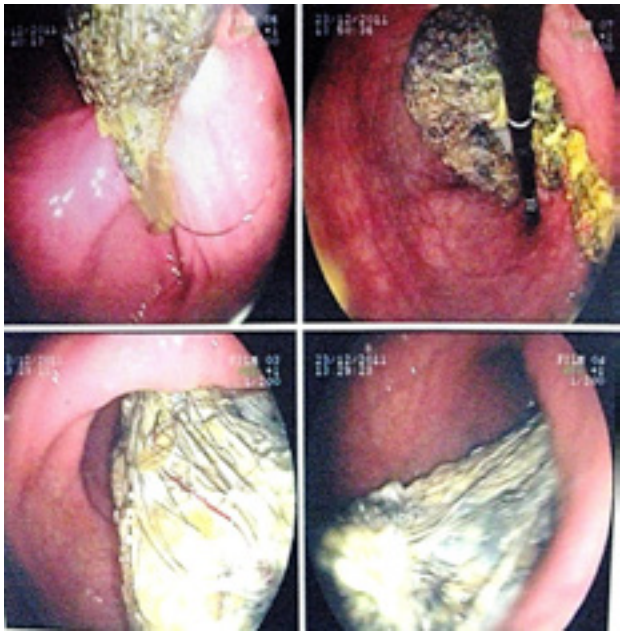

FIGURE 1 : Upper endoscopy. Gastric trichobezoar extended to the duodenum.

the gastric wall was intact and mucosa was normal.By reviewing the history of our patient, we found that the parents never seen their child eating her hair and they didn't observe any strange behavior. However, they reported that she was spoiled and had educational difficulties since the first grade of the primary school and she was often punished by her teachers because she didn't fulfill her homework. The examination of the girl's hair and scalp didn't show any area of hair loss.A longitudinal anterior gastrotomy was performed with extraction of huge ball of hair mixed to fragments of clothes (Figure 2 and 3).

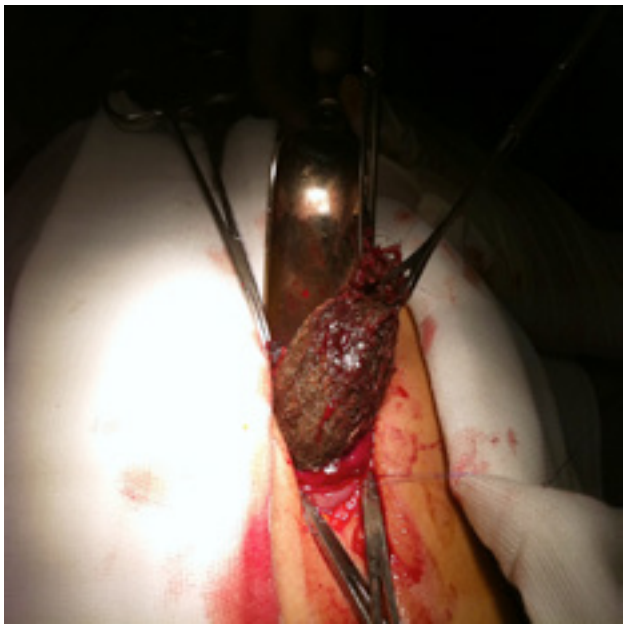

FIGURE 2 : Intraoperative view of trichobezoard extracted by longitudinal anterior gastrotomy

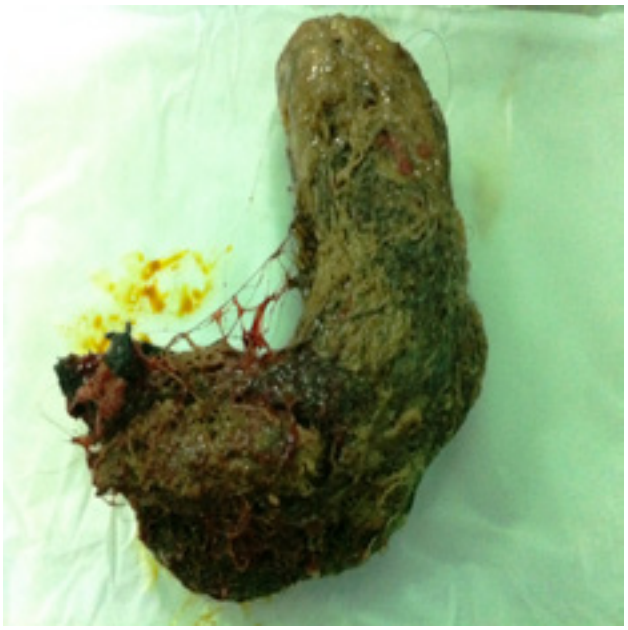

FIGURE 3 : A large trichobezoard 18 × 4 cm J-shaped. Agglomerate of hair and indigestible material weighing about 250 g.

The outcome was favorable and the girl was discharged on the 7th postoperative day. A psychiatric follow up was initiated. The weight became normal within 1 year of follow-up and we didn't observe recurrence of trichphagia.

DISCUSSION :

Our case showed that Rapunzel syndrome could be a cause of recurrent abdominal pain in children. Up to 90% of trichobezoar are found in adolescent girls younger than 20 years [2,3]. Almost 10% of patients have psychiatric disorder, and about 50% of patients have a history of trichophagia or trichotillomania [4,5]. The diagnosis is often delayed because of trichobezoar are often not recognized at the initial presentation [2]. In our patient, the behavioral disorder wasn't evident initially, and trichophagia was not observed by parents. May be this behavior was limited to school due to difficulties of integration. Upper abdominal pain and weight loss are the main complaint symptoms [2,5]. Some patients develop recurrent episodes of nausea, vomiting and dyspepsia with upper abdominal discomfort, gastric heaviness, burn or fullness such as our patient [6]. A non-tender and firm abdominal mass may be palpable in cases with huge trichobezoar [6,7]. A focal area of hair loss could be found on examination [2]. Though to be benign, this condition may cause serious complications

such us intestinal obstruction, perforation, peritonitis, hematemesis, anemia, malabsorption and gastric ulceration. Rarely, pancreatitis, obstructive jaundice, protein-losing enteropathy, intussusception, and appendicitis are observed [2, 8-11]. Those complications were absent in our case.The radiological investigations including upper gastrointestinal opacification, abdominal ultrasound or CT scan contribute to the diagnosis but they are not necessary if endoscopy is done first [2,12]. Endoscopy is the gold standard to diagnose and classify bezoars [2,10,13-15]. The upper endoscopy shows the trichobezoar as a mobile dark greenish ball of hair with a slimy surface [16]. The upper endoscopy guides the modality of treatment [10]. Small bezoars may be fragmented into small pieces and aspirated in one or several sessions [16]. In our case, endoscopic fragmentation failed because of the large size of the trichobezor. In those cases the treatment of choice remains surgical by a laparotomy and gastrotomy removal [2,8-10,15]. Enterotomy may be required in cases of trichobezoars extended to the bowels [8-10]. Although, trichobezoar in our patient was extended to the duodenum (second portion), full extraction was possible only by gastrotomy. This was explained by the highly aggregated trichobezoars allowing a complete withdrawal through the pylorus. Laparoscopy could be used but extraction is more difficult and need more time than conventional laparotomy [2]. To prevent recurrence, the underlying psychological disorder (trichophagia) should be treated. Behavioral training and sometimes drugs help to treat this disorder and prevent recurrence. Prognosis is often excellent after removal of the bezoar and regular follow up in psychiatric consultation [2,3,5].

CONCLUSION:

The diagnosis of trichobezoar in children with recurrent abdominal pain could be difficult not only because of its low prevalence, but also behavioural disorders are difficult to diagnose in paediatric population. The endoscopy has a high diagnostic value as well as a possible therapeutic alternative to surgery which remain unavoidable in huge trichobezoar. Outcomes are generally favorable after trichobezoar extraction and psychological management.

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