Rapunzel syndrome

Ali H. Al-Wadan, MD, Mohamed Al-Absi, MD, Azan S. Al-Saadi, FRCS, Mohamed Abdoulgafour, MSc.

ABSTRACT

An 18-year-old single female patient, presented with non specific gastrointestinal symptoms of anorexia, abdominal pain, and change in bowel habit. Clinically she was anemic, cachectic, and depressed. Abdominal examination revealed mobile epigastric mass. The scalp alopecia and endoscopy coupled by computed tomography scan, confirmed the diagnoses of trichobezoar, but it was not diagnosed as Rapunzel syndrome except after laparotomy, gastrotomy, and enterotomy. There are less than 16 cases of "Rapunzel syndrome" described worldwide, and this is the first case to be described in the middle east.

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Bezoars are defined as foreign bodies formed in the stomach or small bowel, or both, due to an accumulation of swallowed substances. The term bezoar, is believed to be derived from the Arabic "Badzehr" or Persian "Padzahr", meaning counter poison or an antidote. Trichobezoars are gastric concretions composed of hair or hair like fibres, in rare cases, trichobezoar may extend as tail from the stomach into the intestines as far down to the caecum, and this was named and described by Vaughan et al[1] in 1968 as "Rapunzel syndrome".

Case Report. An 18-year-old single female patient was presented to Rada Hospital in Yemen, with a one year history of anorexia, weight loss, weakness, and abdominal pain. The symptoms were non-specific and were mimicking those of other gastrointestinal conditions. She went to different clinics and hospitals prior to her diagnosis. On examination, she was pale, dehydrated, malnourished, depressed, and with areas of patchy alopecia of the scalp. Abdominal examination revealed a mobile non tender mass, occupying the upper part of the abdomen. Blood examination showed low level of hemoglobin and albumin. A plain x-ray of the abdominal showed mild dilatation of small bowel, while abdominal CT with oral contrast revealed an intra-gastric mass consisting of "compressed concentric rings", with a mixed density pattern due to the presence of entrapped air and food debris, the mass was extending and occupying the whole stomach, from the fundus down to the pylorus, the left copula was almost elevated to the right side **(Figures 1 & 2)**.

Endoscopy confirmed the diagnosis, but failed to remove the trichobezoar. She was operated after correcting the anemia, dehydration, and the protein deficiency. Laparotomy was performed under general anesthesia, which revealed an extended huge mass occupying the whole stomach and the pylorus with the tail extending and filling the upper part of small bowel. Gastrotomy was performed, which revealed a huge trichobezoar mixed with food debris, the incision on the stomach wall was extended to deliver a mass approximately 16 x 7 cm, which was gently pulled out of the stomach, the pylorus, and the duodenum. The remaining part of the tail

From the Department of Surgery, (Al-Wadan, Al-Saadi, Abdoulgafour), and the Department of Radiology (Al-Absi), Sana'a University, *Yemen*.

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Address correspondence and reprint request to: Prof. Azan Al-Saadi, Department of Surgery, Sana'a University, PO Box 2236, Sana'a, *Yemen*. Tel. +96 (771) 1476094. E-mail: azanalsaadi@gmail.com

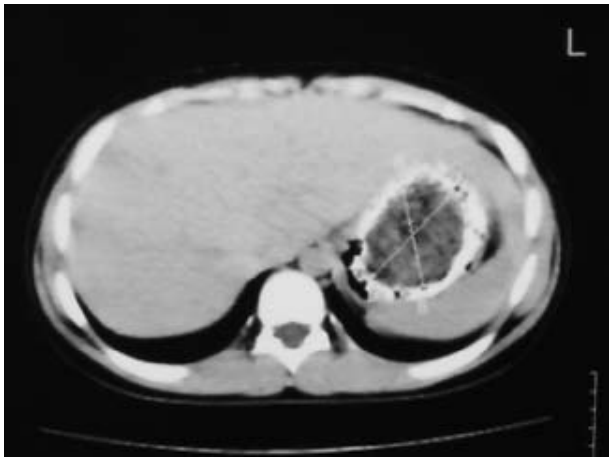

Figure 1 - Abdominal CT, after an oral contrast, in the supine position, showing non-adherent mass at the fundus level.

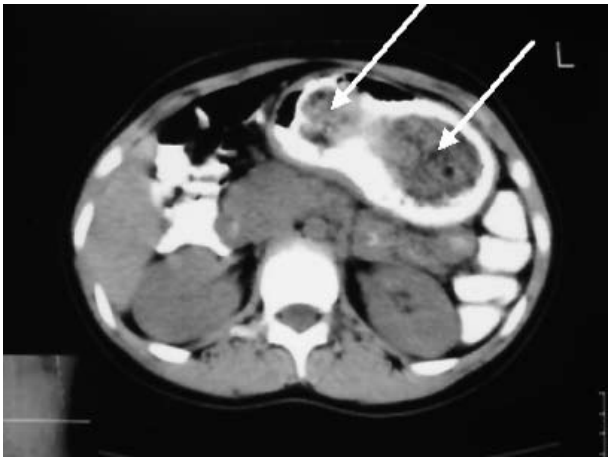

Figure 2 - Abdominal CT, after an oral contrast, in the supine position, showing non-adherent mass at the level of the antrum and extending to the pylorus (white lines).

Figure 3 - Retrieved trichobezoar, the large part was in the stomach with the tail extending into the small bowel.

was removed from the small bowel through an enterotomy incision **(Figure 3)**. The rest of the small bowel was palpated thoroughly. Incisions on both the stomach and small bowel were repaired. She recovered well after the surgery, and was referred for psychiatric therapy.

Discussion. Trichobezoar is related closely to Trichotillomania (pulling at one's own hair) and trichophagia, the first report of a trichobezoar case was in 1779 by Baudamant; and the first surgical excision was performed by Schonborn in 1883. Over 90% of the trichobezoars are found in female, younger than 20 years,[2,3] some with psychiatric problems and some are mentally retarded.[4] The symptoms of trichobezoars are very unspecified, abdominal pain is found in 70%, anorexia, nausea, and vomiting 64%, weight loss, and a sense of weightily oppression in the epigastrium, usually of insidious onset, alteration of bowel habits is seen in 32%. Only half of the patients had a history of trichophagia. The most characteristic physical finding was a large palpable and freely movable upper abdominal mass usually found in the epigastrium, but sometimes, occupying the lower positions with a well-defined, smooth outer surface, and uniform firmness. With time the patient may develop gastric outlet obstruction, ulceration, and perforation.[5] Distal extension (Rapunzel syndrome) of the trichobezoar can lead to obstructive jaundice, acute pancreatitis, protein-losing enteropathy, steatorrhea, constipation, mechanical small bowel obstruction alone or with perforation.

The diagnosis of Rapunzel syndrome may be suspected from an abdominal radiograph that shows a mottled mass. Barium examination may reveal an irregular intraluminal mass, which may confirm the diagnosis. Computed tomography is helpful in the preoperative diagnosis, especially in patients with small-bowel obstruction. In patients with learning difficulties and gastrointestinal symptoms, a low threshold for upper gastrointestinal endoscopy should be maintained.[6,7] The delay in diagnosis may be due to its non specific presentation,[8] but once it is expected, endoscopy and upper gastrointestinal contrast study are diagnostic. Early diagnosis and treatment of the Rapunzel syndrome is very important, in order to avoid fatal complications such as, gastric perforation and intestinal necroses.[9] The therapy of choice of Rapunzel syndrome is surgery; endoscopic removal fails in most cases due to the large gastric ball as well as its intestinal "tail". After surgery, intensive psychiatric follow-up is mandatory for preventing relapses, especially in mentally retarded or depressed patients, and regular upper gastrointestinal endoscopy is recommended.

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