Rapunzel Syndrome: Diagnosed After Laparotomy in a Young Girl

Rapunzel syndrome is an unusual and rare form of trichobezoar found in patients with the habit of hair pulling (trichotillomania) and swallowing it (trichophagia), consequently leading to collection of bezoars in stomach and intestines. We describe this syndrome in a very young girl who presented to us as a case of moderate acute malnutrition with intestinal obstruction.

Bezoars have been known to occur in the form of undigested masses found in the stomach but Rapunzel syndrome involves the presence of a gastric trichobezoar with a long tail extending beyond the duodenum and till terminal ileum [1]. Around 100 cases have been described in the literature since then, with a mean age of presentation of 10.8 years [2,3]. It is usually seen in young girls with or without known psychiatric disorders [4].

A 5-year-old girl presented to us with poor growth and not eating well. There was history of pain abdomen and occasional vomiting after meals. On examination, the child looked pale, emaciated, and stunted. Her hemoglobin level was 9 g/dL. Her weight was 11.2 kg (< 1st centile WHO weight-for-age chart and her height was 92.5 cm (<1st centile WHO height-for-age chart). The abdomen was mildly distended with normal bowel sounds. The patient’s mother admitted that she had a habit of picking hair from floor and secretly swallowing it. A diagnosis of moderate acute malnutrition with moderate anemia with subacute intestinal obstruction was made. The patient was kept nil per orally and received intravenous fluids, and surgical opinion was sought. After two days, the child passed stool mixed with hair strands with relief of abdominal distention. The child was allowed to eat semi-solid food, which resulted in vomiting of strands of hair and recurrence of abdominal distention. CBC showed moderate anemia with dimorphic picture. X-ray abdomen showed dilated gut with multiple air fluid level at various level. Serial USG abdomen failed to detect any intra luminal mass in the stomach or duodenum and ileum. Computed tomography was not available at that time.

Exploratory laparotomy was performed through a supraumbilical midline abdominal incision. A longitudinal 4 cm gastrotomy made on the anterior surface of the corpus of the stomach revealed an intraluminal smooth contour mass occupying bulk of stomach with post pyloric extension. There was a continuous thin strand of trichobezoar in duodenum with thick tail of 2-3 cm diameter along jejunum and terminal ileum, which were removed with separate enterotomy.

After discharge child was provided nutritional rehabilitation. Five months post-surgery, the child has gained 3 kg weight and is on iron supplements. She is receiving behavioral modification advice but not on any psychiatric treatment.

Rapunzel syndrome is rare but should be taken into consideration while investigating a malnourished child with intestinal obstruction. Trichotillomania and trichophagia as a diagnosis are considered on finding some bald patches on scalp or history of habit of pulling out hair and swallowing it [4]. This child did not have bald patches on her scalp.

Upper gastrointestinal endoscopy is the gold standard diagnostic modality but was not available at this hospital [3]. USG abdomen, although is said to be specific for trichobezoar, but in our case serial ultrasound abdomen failed to detect the nature or presence of any intra luminal mass. Ultrasound as an imaging modality has often missed subtle finding of a trichobezoar [5]. CT scan reveals the nature and the extent of the trichobezoar, and is regarded as the best modality for diagnosis. Obstruction of the upper digestive tract is the most common clinical manifestation of this disorder [1].

In the present case, surgery was attempted as an exploratory method due to uncertain cause of intestinal obstruction. Psychiatric follow-up is important, and care should be extended to family members, who should be vigilant with patients since recurrences of the problem have been described [4]. The need for adequate follow-up should be emphasized to avoid recurrences, although these are rare since the trauma of surgery may prevent the patient from provoking another episode.

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