CASE REPORT

Abdominal Mass in a Child with Pica; Rapunzel Syndrome

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ABSTRACT

Rapunzel syndrome is an unusual and rare presentation of gastric trichobezoar that can extends from stomach to small gut or beyond. This case report is about a 6-year-old girl who presented with complaint of severe abdominal pain and vomiting in OPD of pediatric gastroenterology, Lahore General Hospital, Lahore. There was a palpable mass in epigastric region along with pallor. A trichobezoar was diagnosed on esophagogastroduodenoscopy that was extending to the colon and was removed surgically. After treatment and cognitive behavioral therapy symptoms have resolved completely. Trichobezoar must be kept as a differential diagnosis in a patient with abdominal pain and palpable mass.

Key Words: *Bezoars, Trichobezoar, Trichophagia, Trichotillomania*

INTRODUCTION

Rapunzel syndrome is a rare condition which occurs as a result of trichophagia. This syndrome is named after a fairy tale, Rapunzel. Around 110 cases have been reported in medical literature and 90% of them are adolescent female.[1]

Bezoars are accumulations of non-absorbable food and fiber in gastrointestinal tract in patients with mental retardation, psychiatric disorders and pica.[2] Trichobezoar is not very common type of bezoar, it is formed due to swallowed hair and threads. Trichobezoar typically present with nausea and abdominal pain but rarely present with abdominal mass which can lead to intestinal obstruction and perforation. The mass is formed mainly due to gradual accumulation of hair/threads in the mucosal folds of stomach that expands to intestine over time. As it grows slowly symptoms usually appear in adolescent age. Trichophagia (habit of eating hair) and trichotillomania (habit of pulling hair) may be observed along with psychiatric illnesses and mental retardation.[3]

CASE REPORT

A 6 years old, malnourished, developmentally normal girl presented in outpatient department of pediatric gastroenterology, Lahore General Hospital Lahore, Pakistan with history of abdominal pain from last 3 years. Pain was gradually increasing, she visited multiple hospitals and was investigated, but no improvement was observed. Now for last 3 months there was severe postprandial abdominal pain associated with non-bilious vomiting and loss of appetite.

On physical examination, weight and height were at 10th percentile, patient was sick looking and pale. There was generalized abdominal tenderness with a firm, oval mass of 6cm x 4cm in epigastric region. Bowel sound were audible. Patches of alopecia on scalp were observed. Rest of the systemic examination was normal. Laboratory results were as following: white blood

cells: 16.9 x 10^3/ul; hemoglobin: 8.8 g/dl; HCT: 27.3%; MCV: 54.1 fl; MCH: 17.5 platelets: 896 x 10^3/ul; iron: 60 ug/dl; ferritin: 6 ng/ml, anti-tissue transglutaminase IgA <0.5 U/ml and total serum immunoglobulin IgA 6g/L.

Patient had history of pica (trichophagia) and trichotillomania since 2 year of age. Her psychiatric evaluation was done, compulsive behavior was noticed and cognitive behavioral therapy was advised.

A heterogeneous mass was noted in stomach on abdominal radiograph, while abdominal ultrasonography was unremarkable. Esophagogastroduodenoscopy was done and large trichobezoar was seen in stomach which was extending beyond the 2nd part of duodenum (fig 1). Computed tomography (CT) was done to see the extent of trichobezoar, soft tissue densities were seen in stomach extending up to ileocecal junction (fig 2). Because of very large size of trichobezoar laparotomy was planned and done by the pediatric surgeon. The trichobezoar, which had filled the stomach and extended out to the cecum, was removed. Two perforations were found in distal jejunum and proximal ileum, through which bezoar was retracted back and removed. Length of bezoar was 1.6 m. Both perforations were repaired (fig 3). Oral feed was allowed after 6 hours of surgery with ERSA (enhanced recovery after surgery) protocol and patient was discharged on 3rd post-operation day.

Patient is on regular follow-up at the department of pediatric gastroenterology and pediatric psychiatry, Lahore General Hospital. By cognitive behavioral therapy and habit reversal training, patient's habit of trichophagia has improved.

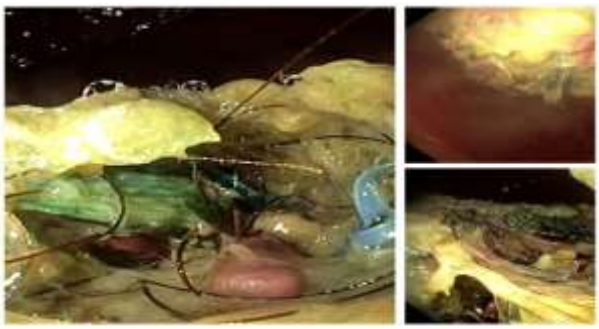

Fig 1: Endoscopic view of trichobezoars

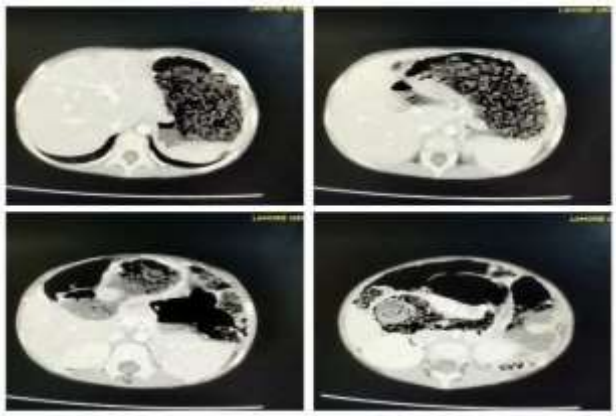

Fig 2: Computed tomography views: soft-tissue densities compatible with a bezoar in the stomach extending throughout the small intestine.

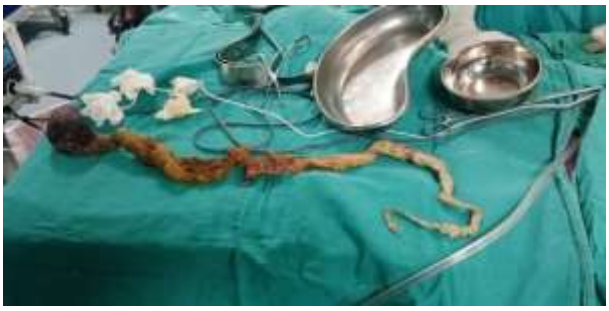

Fig 3: Surgically removed trichobezoar that filled the stomach and was extended to the cecum

DISCUSSION

Trichophagia and trichotillomania is mostly observed in adolescent girls.[4] Patient with trichobezoar usually present with abdominal pain (upper abdomen), nausea, vomiting, diarrhea/constipation, weight loss and epigastric mass.[5] The diagnosis is usually delayed due to non-specific symptoms. Rarely serious complications like intestinal obstruction and perforation can occur.[6,7] Points which made index case worth reporting are very young age of the patient, intestinal obstruction at level of ileocecal junction, two perforations at distal jejunum and proximal which is a rare complication and very long length of trichobezoar i.e. 1.6 m. Trichobezoar usually develops over years that is why majority of patients present and are diagnosed at adolescent age, but index case was

only 6 years old and 1.6 m long trichobezoar was removed.

Abdominal radiography and ultrasonography can be used in diagnose. Esophagogastroduodenoscopy has the highest specificity and sensitivity in terms of diagnosis. Computerized tomography can provide information about presence and extent of the bezoars.[8,9] In index case, the bezoar was not picked on USG abdomen, but was diagnosed on endoscopy and CT scan.

Treatment modality depends on size of bezoars. Endoscopic removal is opted in patients with bezoar length of <20 cm OR if there are multiple small bezoars. The best modality for large size trichobezoars (>20 cm) is removal of the mass by laparotomy/ laproscopically.[10,11]

Neuropsychiatric disorder (obsessive- compulsive disorder) and nutritional anemia was the cause of pica (trichophagia) in the index case. Cognitive and behavioral therapy was initiated in this regard and micronutrient supplementation and nutritional rehabilitation was done. The patient's trichophagia is fully reverted by treatment.

Diagnosis of trichobezoar must be considered in patients especially in young girls with complain of abdominal pain, vomiting and palpable mass in epigastric region. Detailed history about trichophagia and trichotillomania is mandatory.

Conflict of interest: The authors declare no conflicts of interest to this case report. There are no financial, personal or professional affiliations that could have influenced the preparation or presentation of this work.

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