CASE REPORT

Rapunzel Syndrome - Trichobezoar in a 9 Year Old Girl: A Case Report

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Abstract:

We here report a case of 9 years old girl presented in casualty with an acute abdomen and small bowel obstruction and on laprotomy a large trichobezoar was revealed extending into small intestine along with intussuception. Rapunzel syndrome is a rare but potentially fatal trichobezoar. Trichobezoar, an under-diagnosed entity, has to be considered in the differential diagnosis of abdominal pain and a non-tender abdominal mass even in young children.

Keywords: Trichobezoar, Rapunzel syndrome

Introduction:

Bezoars are concretions in the gastrointestinal tract that increase in size by continuous accumulation of non-absorbable food or fibre. The first reference to a bezoar in a human was in 1779 during an autopsy of a patient who died from gastric perforation and peritonitis. It was first described by Vaughan *et al* in 1968 [1]. Most cases are seen in young adolescent girls. Only one case has been reported in male who ate her sister hairs. A cotton bezoar with Rapunzel syndrome was recently reported in an 18-year-old male [2]. Rapunzel syndrome is an unusual manifestation of a trichobezoar in which the mass extends from the stomach and duodenum through a large portion of the small intestine. It is named after a beautiful long haired girl name Rapunzel in a German fairy tale by Grimm brothers (published in 1812) in which Rapunzel let her long golden hair down the prison in the castle over which her lover prince climbed upon to rescue her [3]. The word 'trich' mean hairs in Greek [3]. The term "bezoar" is derived from Arabic, "badzehr" from Persian "panzehr", both meaning counter poison and antidote [4]. De Bakey and Ochsner reviewed 172 cases of trichobezoar of which 90% were females with age range from 10 to 19 years [5]. Bezoars have been shown to be the cause of 0.8% of small bowel obstructions that were treated laparoscopically [6].

Case Report:

A 9 years old girl presented in casualty with severe pain in abdomen, nausea and vomiting and constipation since 2 days. Since 1 month she vomited after most meals. The parents gave the history of patient being developmentally delayed, reduced appetite, weight loss along with habit of trichophagia. Her Hb was 6.6gm% with microcytic hypochromic anemia, TLC= 99000, DLC=P82%, L10%, E4%M4%. She had patch of alopecia on her head. Her CECT scan of the abdomen and pelvis done at private radiologist was reported as grossly dilated stomach with a large non enhancing mixed density intraluminal gastric mass with foci of air and oral contrast and circumscribed by oral contrast. There was similar tubular nonenhancing intraluminal mass in distal small intestinal loops. The duodenum was dilated along with marked dilatation of proximal jejunal loop in left lumbar region with abrupt narrowing. Features were suggestive of gastric and intestinal bezoar with acute small bowel obstruction. Small bowel intussuception was noted in right iliac fossa along with signs of perforation peritonitis. On per abdominal examination an oblong mobile well

defined mass was felt occupying the upper half of the abdomen. Laprotomy performed revealed a trichobezoar measuring 51x7x5cm in stomach extending into the small bowel. Grossly dilated stomach was easily visualized. Thick strands of hair could be felt in the jejunum and ileum and two small perforations were noted 6 and 8 cm at the mesenteric border from gastroduodenal junction. An intussuception was noted 60 cm from duodenojejunal junction. Gastrostomy was done and trichobezoar removed from the stomach. Intussusception of jejunum reduced. Two perforations of jejunum were closed primarily in one layer. Feeding jejunostomy done 10 cm distal to perforation and taken outside from left side of abdomen. The patient died after two days because of septic shock.

Fig.1: Grossly Trichobezoar Measured 51x7x5cm and Comprised of Hair with Very Few Threads.

Discussion:

Trichobezoars result from the swallowing of hair plucked from head or fibers from fur rugs, garments, or woollen clothing and blankets. Trichobezoars are formed when hair strands are retained in the folds of the gastric mucosa because of their slippery surface which prevents their propulsion by peristalsis. As more hair is added, peristalsis causes it to be enmeshed until a mass forms and eventually assumes the shape of the stomach. These bezoars are usually seen in psychologically disturbed young women or mentally retarded children with disorders called trichotillomania (hair plucking) and trichophagia (ingestion of hairs) [3]. There are only forty-one cases reported in the medical literature about Rapunzel syndrome [7]. Maximum number of cases especially more than 30% has been reported in India [8]. Gonuguntla and Joshi reported the youngest documented case of Rapunzel syndrome in the United States: a 5-year-old girl with mental retardation with abdominal pain, vomiting, and a nontender abdominal mass [9].

Diagnostic criteria for Rapunzel Syndrome by Naik *et al* [10]:

1. Trichobezoar with a tail
2. Extension of this tail at least until the jejunum
3. Obstructive symptoms

Conditions predisposing to formation of bezoars include changes in gastric or intestinal anatomy, commonly after gastroduodenal surgery or disorders of gastrointestinal tract motility and gastroparesis.

Of 131 collected cases of trichobezoar, a palpable abdominal mass was present in (87.7%), abdominal pain (70.2%), nausea and vomiting (64.9%), weakness and weight loss (38.1%), constipation, diarrhea (32%)and haematemesis (6.1%).The laboratory investigations revealed low hemoglobin in about 62%(average)[11].

Diagnosis may be aided by abdominal plain films contrast upper gastrointestinal radiography, ultrasonography, CT scan, or upper gastrointestinal endoscopy. Complications of Rapunzel syndrome include pyloric obstruction, bowel obstruction, peritonitis, protein-losing enteropathy, iron deficiency, and megaloblastic anemia and mortality.

Treatment of a bezoar requires removal of the mass and prevention of recurrence. Depending on the clinical status, Rapunzel syndrome may be treated with gastrotomy and/or enterotomy (in cases of intestinal obstruction), or extended resection of the intestine associated with gastrotomy and enterotomy (in cases of necrosis, perforation or peritonitis) [12]. Endoscopic removal is possible for small trichobezoars. Other methods like extracorporeal shock wave lithotripsy, laser ignited mini-explosive technique, intragastric administration of enzymes (pancreatic lipase, cellulose), and medications (metoclopramide, acetylcysteine) have been reported with varying results. Laparoscopy has been also used with limited success [9, 11]. Open surgery still remains the corner stone of large trichobezoar.

Most of the previous cases of Rapunzel syndrome were diagnosed early and had uneventful recovery except a few but in our case the patient reported very late with multiple complications like bowel obstruction, perforation and intussuception and the patient could not be saved. The patient must be having recurrent abdominal pain in the past but either the patient was not taken to the hospital because of psychiatric disorder or the she was not diagnosed of having a trichobezoar.

Conclusion:

Rapunzel syndrome is a rare but potentially fatal trichobezoar. Trichobezoar, an under-diagnosed entity, has to be considered in the differential diagnosis of abdominal pain and a non-tender abdominal mass even in young children, and a history of pica should always be obtained. As recurrences are known, each patient should have a proper psychiatric evaluation and follow-up.

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