Case Report

Rapunzel syndrome trichobezoar in a twelve year old girl: a case report

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ABSTRACT

A bezoar refers to a mass of undigestible foreign material found in the gastrointestinal tract, mainly stomach. The second most common bezoar is the trichobezoar, which usually occurs in the young psychiatric female patients with history of trichotillomania and trichophagia. A 12-year-old female psychiatric patient came with complains of diffuse abdominal pain, vomiting, fever, and constipation. Ultrasonography and CT scan were done, which suggested trichobezoar. Thus, trichobezoar should be considered as differential diagnosis of abdominal pain in young psychiatric female patients. In the literature several treatment options are proposed, including removal by conventional laparotomy, laparoscopy and endoscopy. According to our experience and in line with the published results, conventional laparotomy is still the treatment of choice. In addition, psychiatric consultation is necessary to prevent relapses. We here report a case of 12 years old girl presented with complain of vomiting, pain and lump abdomen, loss of appetite and on laprotomy a large, approximately 156 cm trichobezoar was removed which was extending from stomach to ileocaecal junction. Trichobezoar, an underdiagnosed 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, Psychiatric female

INTRODUCTION

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).

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. Most cases are seen in young adolescent girls. 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. The word ‘trich’ mean hairs in Greek. The term “bezoar is derived from Arabic, “badzehr” from Persian “panzehr”, both meaning counter poison and antidote. The removal
of trichobezoars is mandatory because of the risk of potentially life-threatening complications such as intestinal obstruction, gastric bleeding, and perforation (Ellabban GM).² Gastric trichobezoars are unusual cause of outlet obstruction in the emergency department. De Bakey and Ochsner reviewed 172 cases of trichobezoar of which 90% were females with age range from 10 to 19 years.³

CASE REPORT

A 12-year-old female patient presented with chief complaints of off and on vomiting for six months, progressive loss of appetite for four months, pain and lump in abdomen for two months. The parents gave the history of patient being developmentally delayed, along with habit of trichophagia. On general physical examination, the patient was thin built well looking female with normal basal parameters. Abdominal examination revealed a well-defined, smooth and hard intra-abdominal lump of 8 × 10cm in the epigastic region. The lump was having palpable curved (convex) lower border but upper border not reachable (fingers could not be insinuated between the lump and costal margin). The lump was moving with respiration and dull on percussion. Differential diagnosis of gastric trichobezoar and gastric malignancy was made clinically. Abdominal ultrasound showed a large mass in the epigastric region with echogenic anterior margin. Contrast enhanced computed tomography (CECT) showed grossly distended stomach with heterogenous intraluminal content showing areas of curvilinear band like hyper densities and scattered lucencies suggestive of gastric trichobezoar. Patient was having a habit of hair eating for last one year. Based on the above findings we decided to perform surgery for the trichobezoar. Her Hb was 7 gm% with microcytic hypochromic anaemia, TLC=11000, DLC=P82%, L10%, E4% and M4%. She had patch of alopecia on her head. Laprotomy performed which revealed a long trichobezoar measuring approximately 156 cm (Figure 2) in length extending from stomach to ileocaecal junction. It was removed en bloc after anterior gastrostomy (Figure 1).

DISCUSSION

Bezoars are collections of non-digestible matter that usually accumulates in stomach and can extend to small bowel. Different types of bezoars are phytobezoar (vegetable origin), trichobezoar (composed mainly of hair), lactobezoar (concentrated milk formula), pharmacobezoar (mixed medicine bezoars), and food bolus bezoars. Trichobezoars (concretions of hair) are unusual and are usually found in young psychiatric females, who often deny eating their own hair (trichophagy). It is caused by the pathological ingestion of hair, which remains undigested in the stomach. Human hair is resistant to digestion and peristaltic movement because of its smoothness. Continuous ingestion of hair, over a period of time, can lead to their impaction along with mucus and food material into the stomach. In some cases, however, the trichobezoar extends through the pylorus into jejunum, ileum or even colon, this condition is called as Rapunzel syndrome.

There are only forty-one cases reported in the medical literature about Rapunzel syndrome. Maximum number of cases especially more than 30% has been reported in India. Gonuguntla et al 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.⁴ Diagnostic criteria for Rapunzel syndrome by Naik et al, trichobezoar with a tail, extension of this tail at least until the jejunum and obstructive symptoms.⁵ Conditions predisposing to formation of bezoars include changes in gastric or intestinal anatomy, commonly after gastroduodenal surgery or disorders of gastrointestinal tract motility and gastroperesis.

Bezoars may present with abdominal pain, nausea or vomiting, early satiety, weight loss, intestinal obstruction, ulceration leading to bleeding and/or perforation. Rarely intussusception can also happen (Dalshaug 1999).⁶ Our patient presented with off and on nausea, progressive loss of appetite, upper abdominal pain and lump. An upper abdominal mass remains the commonest presenting sign
(Debakey).\textsuperscript{3} In our case also there was a palpable intraabdominal lump in epigastric region which was moving well with respiration and hard in consistency with curved (convex) lower border. Diagnostic modalities include US, CT scan and upper endoscopy, CT scan has a high accuracy rate, but the accuracy of US in such cases is not so high (Sharma).\textsuperscript{7} The diagnosis is made easily at endoscopy or, indeed, from a plain radiograph (John).\textsuperscript{8} We also diagnosed and confirmed our case with US and CT. Although a psychiatric trouble is usually present, this is not always the case as the syndrome may affect healthy women (Salaam, Coulter).\textsuperscript{5,10} Firm or hard masses in the mid upper abdomen are usually suspected for malignant processes, especially in adults and in old age. Trichobezoar should be considered as a differential diagnosis in young females who present with either history of ingestion of hair or complain of epigastric pain, weight loss and epigastric mass. 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. Management options for trichobezoar include, endoscopic removal, laparoscopic removal, or via laparotomy. The surgical treatment is usually the first choice of treatment in a big trichobezoar. Gorter et al, in a retrospective review of 108 cases of trichobezoar, evaluated the variable treatments tried in these cases; it was noted that whereas 5% of attempted endoscopic removals were successful (small trichobezoars may respond to endoscopic fragmentation and vigorous lavage), 75% of attempted laparoscopic surgical extractions were successful.\textsuperscript{3} However, laparotomy followed by gastrotomy was 99% successful and thus favoured as their management of choice and still remains the cornerstone of large trichobezoar.

We also managed our case by laparotomy and anterior gastrotomy. Medical treatment as enzyme therapy with papain, cellulase, or acetylcysteine may be tried but usually ineffective. So non-operative treatments are discouraged because of high failure rate.

**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. Complications are not infrequent, a diagnosis of trichobezoar and its treatment well in time will prevent these complications. Also, recurrences are known. A psychiatric evaluation, counselling and treatment are helpful in preventing these recurrences.

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