CASE REPORT

RAPUNZEL SYNDROME

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A trichobezoar may be confined to the stomach or unusually it may extend from stomach to the small intestine or beyond when it is labelled as Rapunzel syndrome. Bezoars are concretions in the gastrointestinal tract that increase in size by continuous accumulation of non-absorbable food or fibre. Most bezoars in children are trichobezoars from swallowed hair. Repunzal syndrome is an uncommon condition in children with less than 40 cases reported. We present a case of an 8 year old girl with Rapunzel syndrome. She presented with vague abdominal pain and epigastric fullness of one year duration. On investigation she was found to have a trichobezoar. At laparotomy a large dark foul smelling trichobezoar that occupied the stomach, duodenum and proximal jejunum was removed via gastrostomy.

Keywords: Trichobezoar, abdominal mass, Rapunzel syndrome

INTRODUCTION

Bezoars are accumulations of undigested materials in the GI tract. They are broadly classified according to their primary constituents which may be hair (trichobezoar) vegetable/fruit fibre (phytobezoar), milkcurd (lactobezoar and miscellaneous materials (medications, fungus, foreign bodies, tar and sand etc.). A teenage girl is a typical presentation in 90% of cases and 80% of bezoars occur by the age of 30 usually with underlying psychiatric disorder.[1] Presentation may range from chronic abdominal pain to a previously asymptomatic abdominal mass leading to obstruction or perforation. Rapunzel syndrome is a rare form of trichobezoar. It is named after a charming tale written in 1812 by Brothers Grimm about a young maiden, Rapunzel with long tresses, who lowered her hair to the ground from high in her prison tower to permit her young prince to climb up to her window and rescue her.[2] It is an uncommon diagnosis in children with less than 40 cases reported. Though Vaughan *et al*[2] did not give any strict definitions of the syndrome, both their patients had a trichobezoar with tail-like extension and symptoms of obstruction.

CASE REPORT

An 8 year old girl presented to the paediatric OPD of Fauji Foundation Hospital, Rawalpindi, in March 2010 with vague abdominal pain and early satiety. Mother gave history of poor dietary intake and diarrhoea on and off for the last one year. There was no history of vomiting. On general physical examination, except for pallor, she appeared to be of normal height and weight for her age. Abdominal examination revealed a firm, non-tender mass, approximately the size of a tennis ball in the epigastric region. Baseline laboratory investigations were all within the normal limits. Abdominal ultrasound revealed 'Mild ascites, and several echogenic foci in the stomach'. Upper GI endoscopy revealed 'a trichobezoar extending from the stomach into the duodenum'. Barium meal showed 'a grossly distended stomach with irregular filling defects extending into the proximal gut' (Figure-1). Gastrotomy was performed and a huge trichobezoar was extracted in one piece which took the shape of the stomach, pylorous and duodenum with a tapering tail extending into the proximal jejunum (Figure-2, 3).

Postoperatively, the patient made an uneventful recovery. On repeated questioning, the mother gave a history of noticing her daughter eating threads from various clothes but denied observing trichophagia on any occasion. On careful re-examination the girl had mild alopecia all around her hairline. Before discharge, a thorough psychiatric evaluation and regular follow-ups were arranged.

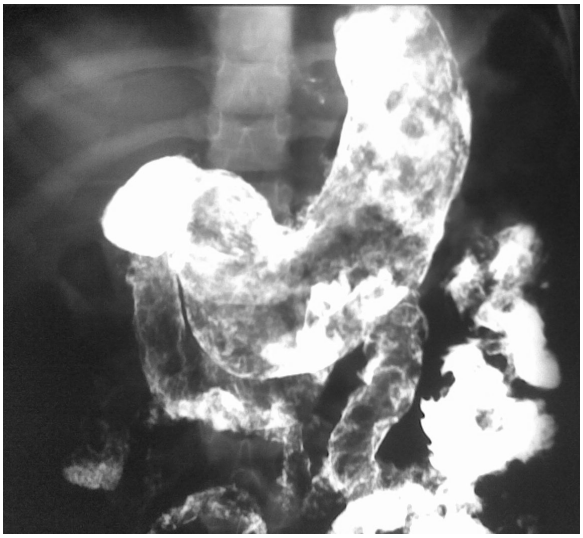

Figure-1: Barium meal examination showing filling defects in stomach, duodenum and proximal jejunum

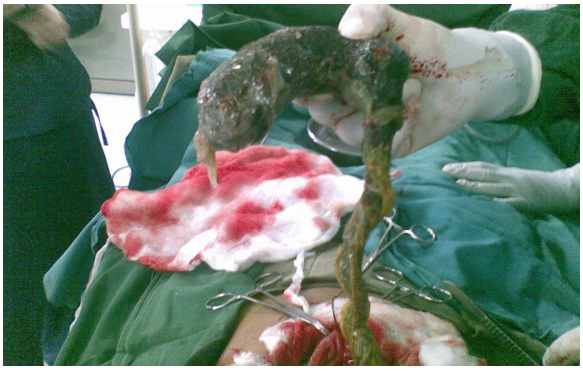

Figure-2: The bezoer being extracted at laparotomy

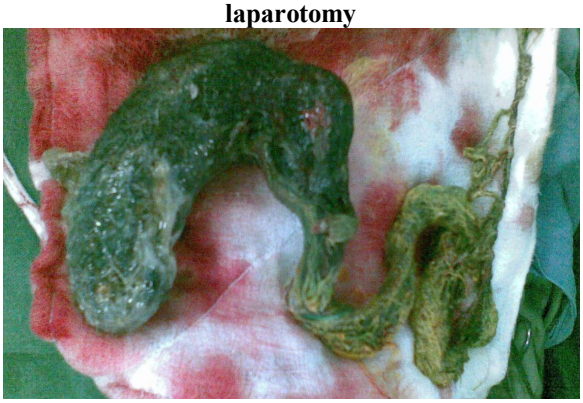

Figure-3: The extracted bezoer

DISCUSSION

Trichobezoar are commonly found in young females usually with an underlying psychiatric disorder.[1] Other social factors predisposing to the psychopathology include: an unstable family constellation, exposure to marital arguments, loss of a parent or even change of a school. The first reference to a bezoar in human was in 1779 during an autopsy of a patient who died from gastric perforation and peritonitis.[3] Trichotillomania or hair pulling was first described in literature in 1889. Among those who suffer from trichotillomania, only 30% will engage in trichophagia or eating their hair, and of these, only 1% will go on to eat their hair to the extent of requiring surgical removal.[4] Trichobezoars form when hair are retained in the stomach, escaping its peristaltic activity due to their smooth and slippery surface and eventually get enmeshed into a ball which usually assumes the shape of the stomach. Trichobezoars are black due to protein denaturation by acid, glistening (from retained mucus) and foul smelling due to the decomposition of fat residues within hair, also resulting in halitosis. Rapujnzel syndrome has been defined in several way, as a gastric trichobezoar with a tail extending up to the ileo-coecal junction[5], or simply a trichobezoar with a long tail, which may extend to the jejunum or beyond or as a bezoar of any size which can cause intestinal obstruction.[6] Patient may remain without symptoms for a long period and may only seek clinical attention when the bezoar becomes large enough to cause intestinal obstruction or a sudden perforation. In early stages anorexia, malaise, vague abdominal pain may occur. Weakness and weight loss may be noticed. With gradual increase in size nausea, vomiting, epigastric pain, and occasionally hematemesis or intussusception can occur. Bezoar may cause chronic inflammation of gastric mucosa, Menetrier disease, gastric polyposis and protein losing enteropathy with steatorrhoea. Iron deficiency anaemia is present in many cases. On one hand it may be due to impaired gastrointestinal absorption or it may present as a form of pica syndrome leading to trichophagia and bezoar formation.[7] Pressure necrosis may lead to mucosal erosions and ulceration. This eventually leads to bleeding perforation and peritonitis. Jaundice and acute pancreatitis due to obstruction of the ampulla of vater may occur.[8]

In history, stress should be placed upon trichophagia and finding out the social situation surrounding the patient. On physical examination, halitosis, alopecia and a firm, non tender, mobile mass, palpable in the epigastrium may be noticed. On investigations a plain abdominal radiograph may show a prominent gastric outline with an intragastric mottled mass, out lined by gas in the distended stomach, mimicking a food filled stomach. On barium study, a large intraluminal filling defect is seen which may extend beyond the stomach. Study should continue till the contrast reaches the ileo-caecal junction to look for any extension or synchronism of gastric bezoar. An abdominal U/S demonstrates a superficially located broad band of high amplitude echoes along the anterior wall of mass with sharp, clean post acoustic shadowing. Plain abdominal CT usually shows a mobile intragastric mass consisting of compressed concentric rings, with a high density pattern due to the presence of entrapped air and food debris.[9] The gold standard for diagnosis is upper GI endoscopy, which apart from diagnosis may render therapeutic intervention possible. Treatment depends on the size and location of bezoar. Endoscopic removal may be carried out for small sized bezoars. Bezotomes, devices which pull verize bezoars either mechanically or with acoustic waves have been used to fragment large bezoars. Extracorporeal shock wave lithotripsy and endoscopy with the use of laser ignition with mini-explosions may be used. Laproscopic removal also has been described. Intragastric administration of enzymes (pancreatic lipase, cellulose and medications (metoclopramide, acetylcysteine) have been used with varying success. Surgery remains the main treatment option for removal of a giant bezoar. A

traditional laparotomy with a gastrotomy and enterostomy, single of multiple if required, is carried out. After removal, rest of the GI should be inspected and palpated for any perforation or for a synchronous bezoar. Memon *et al*[10] have reported a case of recurrent Rapunzel syndrome due to non-compliance of psychiatric medication. Psychiatric evaluation, treatment and parental counselling is an important part of treatment. Regular follow-up is essential to prevent recurrence.

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