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Case Report

RAPUNZEL SYNDROME IN CHILDREN: CASE REPORT

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ABSTRACT

Rapunzel syndrome refers to a very rare condition in which swallowed hair forms a gastric trichobezoar that has a long tail extending into the small bowel. We describe 2 cases of Rapunzel syndrome in female child who presented with abdominal mass, epigastric pain and vomiting. Barium study showed a markedly dilated stomach filled with coarse heterogeneous materials. Upper gastrointestinal endoscopy revealed a huge hairy ball with a tail extending through the pylorus. We performed a surgical laparotomy and successfully removed a huge trichobezoar with a long tail extending into the middle portion of jejunum. Psychiatric consultation revealed trichotillomania and trichophagia and followup done. Rapunzel syndrome should be included in the differential diagnosis in children with chronic abdominal pain and trichophagia.
Keywords: Rapunzel syndrome, Bezoars, Trichotillomania, Trichophagia, Abdominal pain

INTRODUCTION

A bezoar is a concretion of undigested exogenous material that accumulates in the gastrointestinal tract of humans and some animals. The term "bezoar" is derived from Arabic *bedzehr* or *Persian* padzhar, meaning "counterpoison" or "antidote." Some societies have historically believed bezoars from animal guts had magical properties and used them as antidotes to certain poisons [1]. The following 4 types of bezoars have been described: phytobezoars (comprising vegetable or fruit fibers), lactobezoars (comprising milk curds), trichobezoars (comprising hair), and pharmacobezoars (comprising pills or capsules) [2].

- Bezoar is a tightly packed collection of undigested material that is unable to exit the stomach.
- Most bezoars are of indigestible organic matter such as hair-trichobezoars; or vegetable and fruit fibre – phytobezoars; or a combination of both. Other rare substances were also described in literature.
- Trichobezoars have been described in literature and they comprise 55% of all bezoars.
- They commonly occur in patients with psychiatric disturbances who chew and swallow their own hair. Only 50% will have history of trichophagia.
- Rapunzel Syndrome in very rare cases, the hair extends through the pylorus into the small bowel causing symptoms and signs of partial or complete gastric outlet obstruction.
- Here , we report two cases of Rapunzel syndrome who presented with chronic gastrointestinal symptoms.

CASE 1:

A 6 year old female child referred by a local pediatrician to our outpatient clinic with a clinical suspicion of malignancy .Complains of mass in the upper abdomen, intermittent pain and non bilious vomiting . Her history reviled trichophagia with anorexia and loss of weight. Child 's general examination behavior was shy type with anemia and malnourished and scalp showed an area of local sparseness in the front parietal region.(fig 1). Local examination revealed single well defined, intraabdominal mobile, firm mass of size 8 x5 cm in the epigastric region ,non tender,non pulsatile, surface smooth(fig2) .Investigations of barium follow and upper gastroendoscopy revealed gastrictrichobezoar .(fig 3). Elective Laparotomy – gastrotomy and extraction under general anaesthesia. Operative findings were gastrotrichobezoar with tail extending into the jejunum.(fig 5). Postoperative period uneventful .(fig 7).Psychiatric consultation was given.

CASE 2:

A 9 year old female child presented with vomitings and abdominal distension since 5 months.History revealed pica , loss of hair, decreased apetite.General examination revealed, conscious and coherent , malnourished and anemia , sparse hair(fig 8).Local examination found transverse ,oval shaped 15 x 12 cm non tender, firm in consistency with smooth surface palpable occupying epigastric , left and right hypochondrium.Investigations ultrasound shown Echogenic mass with dense after shadowing noted in the epigastric region suggestive of intragasric bezoars.Barium study revealed grossly distended stomach, evidence of large ill defined intraluminal defct occupying the cistal body of stomach with extension into the duodenal c- loop and proximal jejunum.(fig 9) Multiple luminal narrowing of mid jejunum.Impression shown multiple gastric and duodenal trichobezoars and multiple jejunal strictures. Psychiatric evaluation done diagnosed as trichotillomania with mild mental retardation.Elective laparotomy – gastrotomy done and extraction of trichobezoar .Operative findings with gastrobezoar with extention into jejunum (fig10-12) .Postoperative period uneventful and psychiatric revaluation done and monitored.

DISCUSSION:

Trichobezoar is the most commonly encountered ,exclusively seen in young females often associated with psychiatric problems . We report two cases , presentation in the young age group with hair extending down to the proximal jejunum with chronic gastrointestinal symptoms which mimic infectios and infestations especially in endemic areas. It is postulated that hair strands too slippery to be propulsed are initially retained in the mucosal folds of the stomach and become enmeshed over a period time. Trichobezoars are usually black from denaturation of protein by acid , glistening from retained mucus and foul smelling from degradation of food residue trapped within it.(3)

Fig 1 showing focal area of hair sparseness over the scalp.

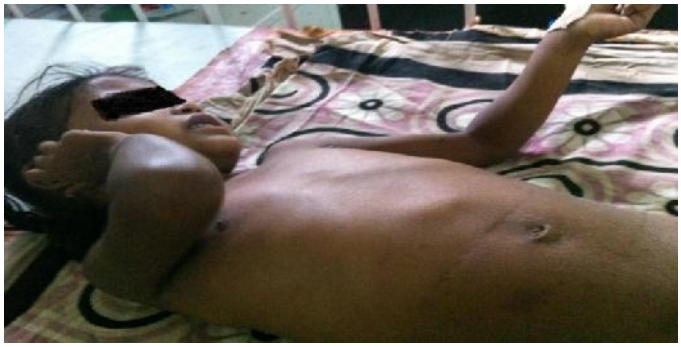

Fig 2. Showing mass in the epigastric region

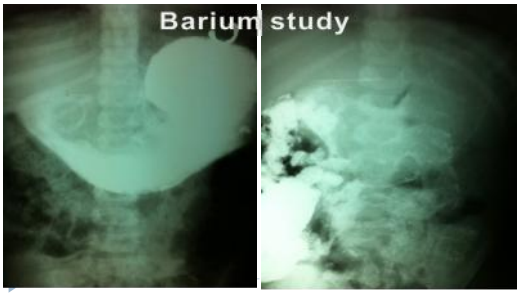

Fig 3 Barium study showing filling defect .

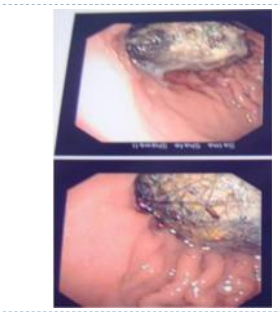

Fig 4.Upper GI Endoscopy showing Trichobezoar.

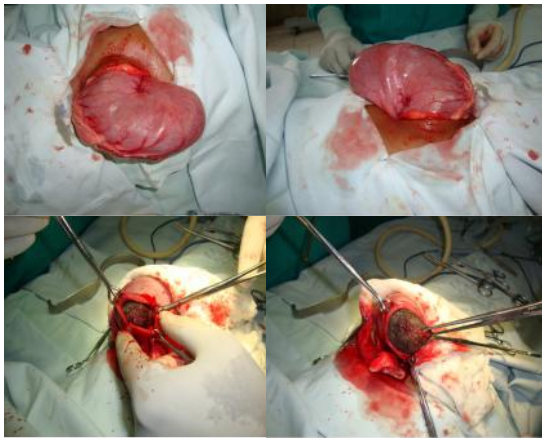

Fig 5 operative findings.

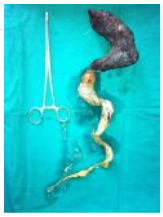

Fig 6. Specimen of trichobezoar.

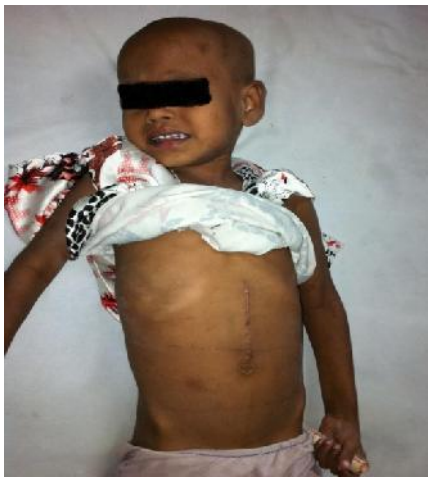

Fig 7.Postoperative photograph.

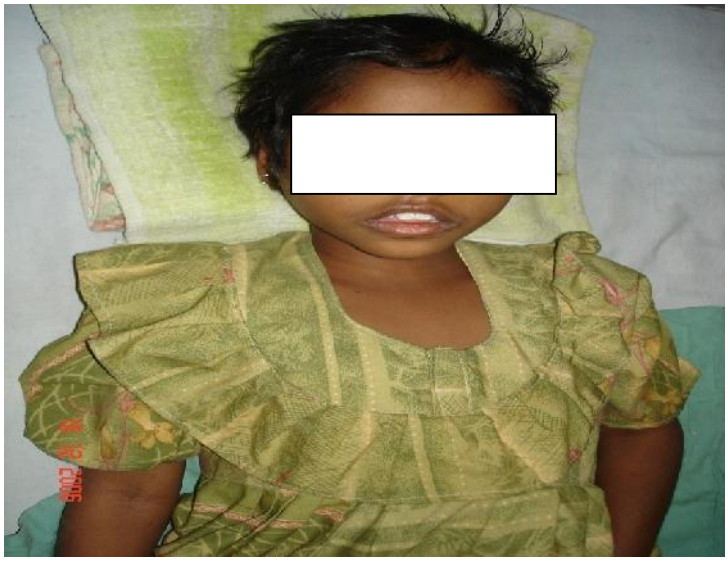

Fig 8. Child showing hair sparseness over the scalp.

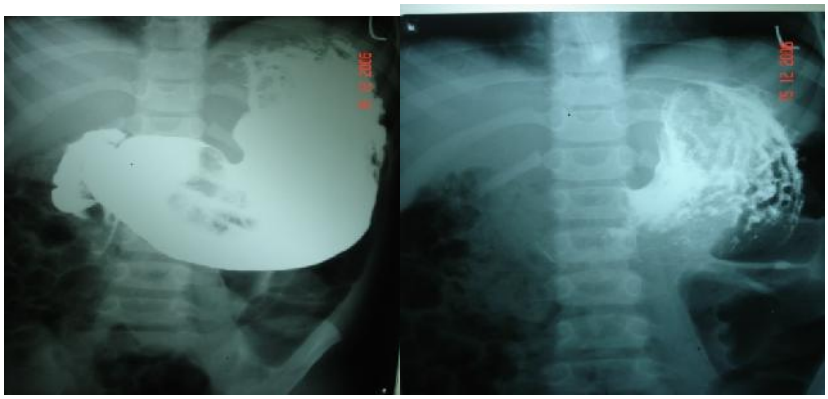

FIG 9. Barium study showing gastric filling defect.

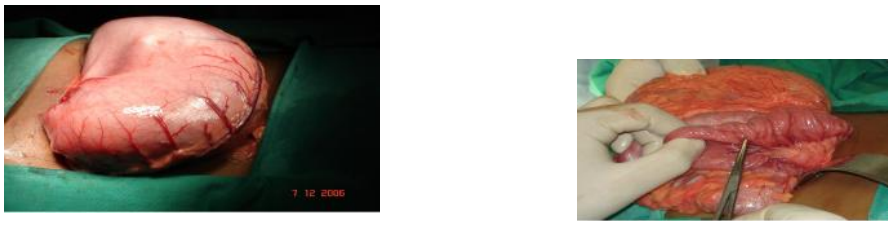

Fig 10. Operative findings –Gastric trichobezoar with tail extending into thesmall bowel.

Fig 11. Specimen extraction of trichobezoar with tail.

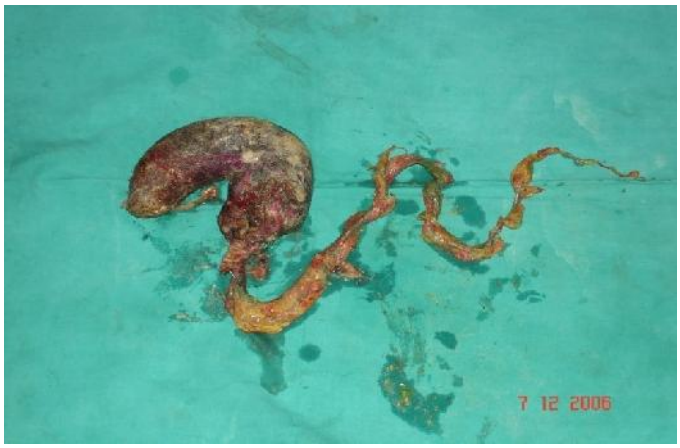

Fig 12. Specimen of gastric trichobezoar with tail.

Vaughan et al. in 1968 first described Rapunzel Syndrome, a rare presentation of trichobezoar,involving strands of swallowed hair extending as a tail through the duodenum, beyond the stomach.(4)

- It is named after a tale written in 1812 by the Brothers Grimm about a young maiden , Rapunzel, with a long hair who lowered her hair on to the ground from a castle , which was a prison tower to permit her young prince to climb up to her window and rescue her.(5) The common presentation of trichobezoar is in young females usually with an underlying psychiatric disorder. In patients with Trichotillomania (a psychological condition that involves strong urge to pull hair), around 30% will engage in trichophagia, and of these, only 1% will go on to eat their

hair to the extent requiring surgical removal. Less than half of the patients give a history of trichophagia.

- They generally cause non-specific symptoms. Palpable abdominal mass, Pain abdomen ,
- dyspepsia. , Nausea & vomiting, Post-prandial fullness. Failure to thrive. GI bleed. Intestinal obstruction and perforation.
- COMPLICATIONS include Anaemia., Gastric ulceration, Intestinal obstruction, perforation and peritonitis, Internal fistulae, Intussusception, Obstructive jaundice, Pancreatitis, Malabsorption, Protein losing enteropathy and Short bowel syndrome.
- Personal history, Psychiatric disorders in family, previous bezoars as wellas physical examination of palpable mass, halitosis and patchy hair loss aids in the diagnosis.
- Presentation of trichobezoar is usually late,due to low index of suspicion by the clinician.
- A palpable abdominal mass is present in 87.7%,abdominal pain in 70.2%,nausea and vomiting in 64.9%,weakness and weightloss in 38.1%,constipation or diarrhea in 32% and heamatemesis in 6.2% (5).
- Abdominal mass is usually well defined and mobile in 90% cases and may be indentable (Lamerton's sign).
- Various imaging modalities help in detection of bezoars(6,7).
- On conventional radiography,a mass of opaque soft tissue is visible in a swollen stomach (6,8).
- Edge of the bezoar may be seen as calcification (6,8).
- Ultrasonography has a limited role in diagnosis,it shows a typical curvilinear trichobezoar with bright echogenic band but high echogenicity of hair and the presence of multiple acoustic interfaces created by trapped air and food limit the ultrasonography of the trichobezoar (6,9).
- Both contrast radiography and upper GI endoscopy are the diagnostic procedures of choice(6,9).
- The upper GI contrast radiography confirms the existence of trichobezoar and also detects other complications such as gastric ulcers.Furthermore,upper GI endoscopy is also used for retrieval of proximal minor trichobezoars (6).
- The computed tomography is the most useful diagnostic tool because it reveals the localization of the bowel obstruction.A mottled gas pattern in the mass is reported characterizing the bezoar, and it is supposed to be the air bubbles retained within the bezoar(9,10).
- Recently ,Magnetic Resonance Imaging(MRI) has been recommended for the small intestinal disease ,MRI visualization of bezoars can be improved by breath holding with fast imaging techniques.MRI picture of bezoar is a mass in the small bowel containing mottled and confluent low signal intensities on both T1- and T2- weighted images(10).

- Treatment : Treatment in the early stages by endoscopic removal used only for small trichobezoars and there is risk of bowel perforation.(10)
  Other treatments like chemical dissolution,mechanical fragmentation,laser ignited mini explosive techniques (11) and endoscopics biopsy devices may be successful only for small bezoars.So surgery is the treatment of choice for large trichobezoar extending into the small bowel.(12)

Psychiatric evaluation is a must for all cases.For prevention of recurrence and excellent long term prognosis,parental counseling ,behavioural therapy and psychiatric followup is necessary.

CONCLUSION:

Trichbezoar is a rare condition especially in small age group of children and moreover Rapunzelsyndrome is still more rare. A Female child with history of trichotillomania and trichophagia along with features of chronic abdominal pain with clinical examination showing sparse hair over the scalp and abdominal mass in the epigastric region one should suspect the diagnosis of Rapunzel syndrome. And Psychiatric evaluation and treatment is must for prevention of recurrences.

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