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A rare case of Rapunzel syndrome in young boy: A case report

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Abstract
Trichobezoars are seen usually in adolescent girls and exploratory laparotomy for removal of trichobezoar is still management of choice though recently endoscopic removal, laparoscopic assisted and laparoscopic removal have been reported in adults and older children, with not much success. Here we report a 7-year-old boy who underwent exploratory laparotomy for removal of gastric trichobezoar, with uneventful recovery. The case is highlighted here for its rarity.

Keywords: trichotillomania, rapunzel syndrome, trichobezoar, exploratory laparotomy

1. Introduction
Trichobezoar is a mass of swallowed hair forming a ball in the proximal gastrointestinal tract, is a rare condition almost exclusively seen in adolescent females [1-3] with a history of psychological disorder. Human hair is resistant to digestion as well as peristalsis due to its smooth surface and accumulates between the gastric mucosal folds. With time, continuous swallowing of hair leads to the impaction of hair along with other gastric contents like mucus and food, causing the formation of a trichobezoar. In 1968 Vaughan *et al.* described that trichobezoar is usually confined within the stomach; sometimes it may extend through the pylorus, duodenum into jejunum, ileum or even colon and this condition is known as Rapunzel syndrome. Most trichobezoars lack symptoms in the early stages and may not be recognized due to their nonspecific presentation and attain large size before becoming symptomatic. Most commonly, trichobezoars produce obstructive symptoms, but they may cause ulceration and bleeding [4].They generally cannot be removed endoscopically and are often removed surgically.
Psychiatric disorders, like mental disorders, abuse, obsessive compulsive disorder, depression and anorexia nervosa may also be associated with trichobezoar, in young females with psychiatric comorbidity. Usually it is the result of the urge to pull out one's own hair (trichotillomania) and swallow it (trichophagia). Unrecognized cases of trichobezoar grow in size and weight due to the continued ingestion of hair. Sometimes a part of the tail of trichobezoar may break and migrate to the small intestine leading to acute intestinal obstruction [5, 6]. Trichobezoar may cause severe complications such as gastric mucosal erosion, ulceration and even perforation of the stomach or the small intestine, intussusception, obstructive jaundice, protein-losing enteropathy, pancreatitis. In severe cases death may occur.

2. Case Report
A 7 year-oldSikh (a community in which the male members also keep their hairs long) boy was detected to have a mass abdomen by a private practitioner when he was being evaluated for breathlessness. He was put on empirical treatment but no response. There were no symptoms pertaining to the mass. On examination he had an 18 cm × 9 cm firm to hard mass in the epigastrium and left hypochondrium. Plain xray abdomen revealed distended loops of gut along with globular mass in the stomach (See figure1).
Ultrasound of the abdomen showed a solid mass occupying the entire stomach, possibly a trichobezoar. This was followed by barium meal to confirm the diagnosis (See figure 2). On further probing it was revealed that parents had noticed that since last four years, he used to pull out and chew his hair and sometimes even others' along with threads, rubber bands etc. They said the problem resolved spontaneously after 6 months of treatment given by psychologist. But he continued eating hairs. For continuous low weight and anemia, he took treatment from local practitioners who prescribed him iron and calcium supplements along with anti-helminthes.
He was admitted in the hospital with complaint of mass in epigastric region, decreased appetite, severe anemia and fatigue. He was advised exploratory laparotomy and removal of the trichobezoar. Under general anesthesia and through themidline incision, supraumbilical incision laparotomy was conducted. On inspection of stomach, duodenum and proximal jejunum, stomach revealed the presence of foreign body in its entire segment. An incision was given along the anterior wall of stomach in its middle part, stomach opened in layers and trichobezoar was identified and with the hook of index finger, the antral part of the trichobezoar was delivered. Distal part of trichobezoar was delivered through the gastrostomy wound in toto. Then the fundal part of the trichobezoar was delivered. (See figure 3 and 4).Stomach was closed in layers, 3-0 vicryl suture after putting a Ryles tube No 14Fr. And abdominal drain of 20Fr was placed. Abdominal wound was closed with 2-0vicryl suture. The procedure took 45 minutes (operative time). The child passed stools on day 3. Feeding with liquids was started on post-operative day 3 when bowel sounds appeared and by the day 5 semisolid food was allowed. Abdominal drain was removed on day 5th. The child had an uneventful recovery and was discharged on 7th day of operation.

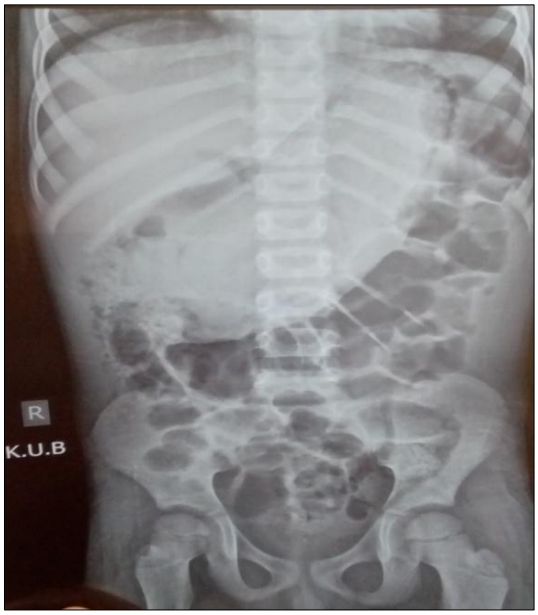

Fig 1: Plain radiograph showing distended gut loops along with globular mass in stomach.

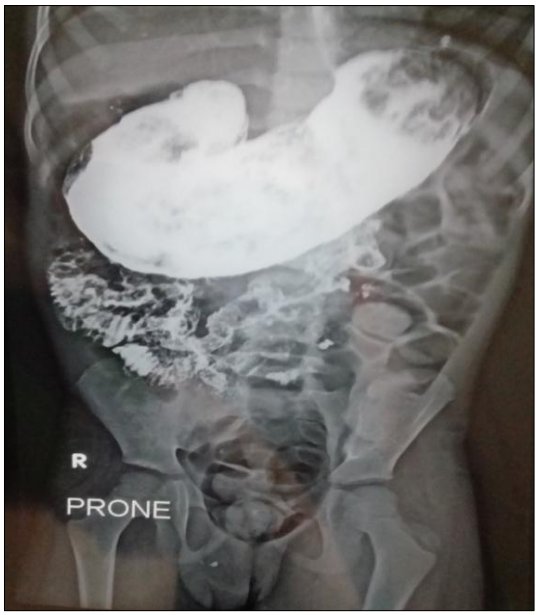

Fig 2: Barium meal showing gastric impaction with trichobezoar and its extension beyond pylorus into small gut

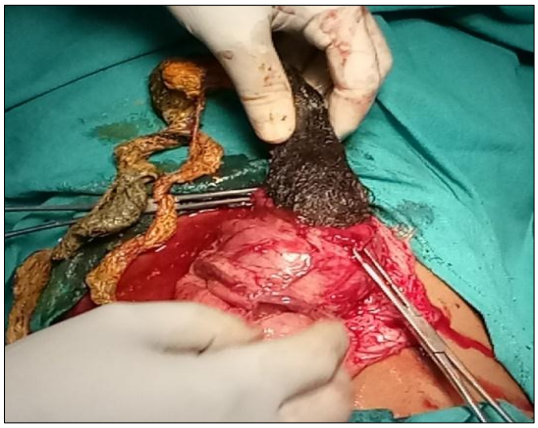

Fig 3: Proximal end (fundal) of trichobezoar being removed after removal of distal (pyloric) end and its extension into small intestine. (Rapunzel syndrome)

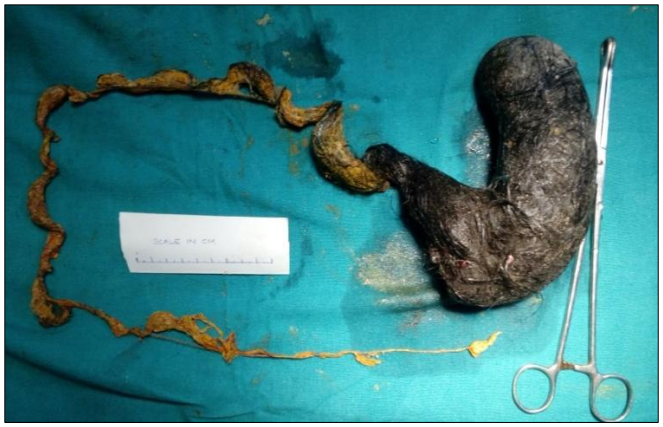

Fig 4: trichobezoar removed in our case was of about 100cm in length

3. Discussion

Trichobezoars often go undiagnosed during initial presentation and hence the much needed intervention is often delayed. Masses in the upper abdomen are usually suspected for malignant processes. Gastric Trichobezoars are mostly seen in young females along withpsychiatric illness leading to pulling out hair known as trichotillomania and ingestion of hair known astrichophagia [7]. But our patient is a 7 years old young boy, and had the same background of trichophagia. Trichobezoar should be considered as a differential diagnosis in adolescent females who present with symptoms like epigastric pain, fatigue, weight loss, epigastric mass and behavioral disorders. In 2009 Veena Gonuguntla *et al.*, found that the frequent location of these casts in the stomach is due to the holdup by the pylorus and the churning action of the stomach which helps to entangle new hair into the already formed cast [8]. Trichobezoars tend to form cast of stomach, with strands of hair having tend to form a cast of the stomach, with strands of hair having been observed as far as the transverse colon [9]. Decomposition and fermentation of fats give the bezoar, and the patient's breath, a putrid smell [8]. Being rare and often misdiagnosed, surgeons should be prepared to deal with trichobezoar, when presents in an emergency with obstructive enteropathy.

Treatment options include enzyme therapy (papain, cellulose, or acetylcystine), endoscopic disruption and removal, or surgical removal [4]. Recently in addition to endoscopy, other method of minimally invasive techniques that is laparoscopy [11-14] is being tried as compared to laparotomy. However these techniques are of limited usefulness, and larger trichobezoars require surgical removal [9]. Rates of successful endoscopic removal of Trichobezoars are very low. There are many case reports indicating unsuccessful attempts of endoscopic removal with or without fragmentation of trichobezoar [6, 11]. Nirasawa *et al.* were the first to report on laparoscopic removal of a trichobezoar. Since then very few reports of attempted laparoscopic removal were published [6, 12, 14].

Regarding laparotomy, it has 100% success rate, the relatively low complication rate, the low complexity, and the ability to carefully examine the entire gastrointestinal tract for satellites in a short period of time. Laparotomy is still considered the treatment of choice. Complications during removal of the trichobezoar likeperforation of the intestine, minor wound infection, pneumonia, paralytic ileus and fecal leakage through the lower part of the laparotomy wound have been reported [5].

However laparotomy was successful in majority of cases.
Exploratory laparotomy has been the most favored method in literature for removal of trichobezoar extending beyond jejunum (Rapunzel syndrome).Endoscopic removal should be avoided as the tail usually reaches jejunum and may break/dislodge causing acute obstruction in distal intestinal tract [5, 6]. Laparoscopy has been successful in Trichobezoars invading duodenum. But as per most literatures the Rapunzel tail at least reach into jejunum, hence these cannot be said to be Rapunzel syndrome [13, 15]. In case of breakage of tail or impending intestinal obstruction, multiple enterotomies may be needed.

4. Conclusion

Regarding treatment, we consider exploratory laparotomy is the treatment of choice in children with trichobezoar and should be the only treatment in children with Rapunzel syndrome. Trichobezoar should be considered in young females presenting with non-specific abdominal complaints. Along with surgical management, psychiatric consultation should be taken.

5. References

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