Rapunzel Syndrome Diagnosed on CT Scan in Adolescent Girl with Neuropsychiatric Disorder

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Abstract
Rapunzel syndrome is a rare type of trichobezoar with an extension of the hair ball into the small bowel. It is seen in adolescent girls with neuropsychiatric disorders. We present a 13 years old girl with rapunzel syndrome presenting with history of epigastric pain and vomiting containing hair for last 1 year. Rapunzel syndrome is a rare form of trichobezoar, with a tail extending into the small intestine. It has a varied presentation and is seen in females with psychiatric disturbance.

Key words: Neuropsychiatric disorders, Repunzel syndrome, Trichobezoar.

Introduction
Bezoar is defined as hard indigestible mass of material such as hair, plant fibers or seeds formed in the alimentary canal of animals and occasionally humans. It was formerly considered to be a useful medicine with magical properties and apparently still used for this purpose in some countries. According to the substance forming the ball, may be termed trichobezoar (hair ball), trichophytobezoar (hair and vegetable fiber mixed) or phytobezoar (Vegetable food ball). Other rare substances forming bezoars has also been described in literature.¹ Trichobezoar commonly occur in patients with psychiatric disturbances who chew and swallow their own hair called trichophagia. Repunzel syndrome is a rare form of trichobezoar in the stomach and its tail extending into the small bowel.² The lifetime prevalence is estimated to be between 0.6%-4% of the overall population.³ The standard imaging modality of choice to make a pre surgical diagnosis is CT scan of the abdomen.⁴

Case Presentation
A 13-year-old girl was referred to our radiology imaging department, with history of epigastric pain and vomiting with vomitus containing hair for almost one year. She was treated in different primary health care centres as a case of gastroenteritis and acid peptic disease. On examination an oblong mobile well defined mass was palpable at epigastrium. On questioning retrospectively her parents gave positive history of tricophagia. We performed CT abdomen and pelvis with contrast of this patient. CT scan showed distended stomach with well-defined air containing filling defects extending from stomach into duodenum and jejunal loops (Figure 1). There was also evidence of multiple spontaneous intussusceptions involving small bowel loops in the left upper quadrant (Figure 2). Prompted by these diagnostic findings and positive history of trichophagia patient was referred for surgical and psychiatric management.

Discussion
Trichobezoars are commonly seen in young females, often with an underlying psychiatric problem. Clinical presentation may be confusing. Patients are usually asymptomatic at beginning. A history of recurrent hospital visits with abdominal pain and vomiting is found in most cases. History of trichophagia is not always present either due to embarrassment or the unintentional nature of the problem. However prolonged intake of indigestible material can lead to gastric outlet obstruction which may present similarly to small bowel obstruction.

First case of trichobezoar was reported by Baudamant in 1779.¹ The postulated reason for formation in the stomach is that human hair especially long hair is resistant to digestion as well as peristalsis. So it tends to stay in stomach and over a period of time may form a large “hair ball” if hair eating (trichophagia) is continued. Most patients with trichobezoar have a co-morbidity of trichotillomania.⁷ Trichotillomania also known as trichotillosis is an irresistible desire to pull
Figure 1. Gastric bezoar extending into duodenum and jejunal loops

out one’s own hair. It was first described by Hallopeau in 1889. Repunzel syndrome is a rare form of trichobezoar located in the stomach and tail extending into the small bowel. Repunzel syndrome is named after a fairy tale written in 1812 by the Brothers Grimm about a young maiden, Repunzel, with long tresses. No fixed criteria have been described to define Repunzel syndrome. Some have defined it as a gastric trichobezoar with a tail extending up to ileocecal junction. Some have defined it simply as a trichobezoar with a long tail which may extend to the jejunum, ileum or ileocecal junction and still others have defined it as a trichobezoar of any size which presents in the form of an intestinal obstruction. Trichobezoar can also be found distally in the gastrointestinal tract without continuity with the stomach bezoar due to breakage and distal propulsion. Presentation ranges from nonspecific abdominal or epigastric pain to a range of complications including ulceration, perforation of stomach or small intestine, gastric outlet obstruction, intussusceptions, obstructive jaundice, protein losing enteropathy, pancreatitis and death. The hair appears black due to denaturing of the hair protein by the gastric acid. The most common tool used in literature for diagnosis of trichobezoar is a CT scan abdomen, which

shows a well-defined intraluminal ovoid heterogenous mass with interspersed gas.

Figure 2. Multiple spontaneous intussusceptions in left upper quadrant.

Conclusion

Trichobezoars should be considered as a differential diagnosis in young females with history of trichophagia and trichotillomania. Diagnosis can be easily made with the use of CT scan. Majority of these patients have an underlying psychiatric or social disorder, so a multidisciplinary approach is essential to prevent recurrence of the problem.

References