Rapunzel syndrome

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Abstract

Introduction. Trichobezoars are foreign bodies in gastrointestinal tract, composed of hair. They occur mainly in children and adolescents suffering from trichotillomania. They commonly occur in the stomach, but as they enlarge over time, they can extend through the pylorus into distal parts of the small intestine resembling a tail. This rare form of trichobezoar is named Rapunzel syndrome. Case report. We presented a 19-year-old female patient, who suffered from trichotillomania and trichotillophagia, which led to trichobezoar formation. Intra-abdominal tumour was suspected after initial clinical examination. Abdominal echosonography, endoscopy and abdominal computed tomography (CT scan) in the pre-operational period revealed trichobezoar formation. The patient was operated on and subjected to further psychiatric treatment. Conclusion. Trichobezoar should be differentially diagnostically taken into consideration in younger women with abdominal pain, nausea, vomiting, palpable abdominal mass and psychiatric disorders. Most trichobezoar cases require surgical treatment, whereas the patients need long-term psychiatric treatment and monitoring.

Key words: bezoars; trichotillomania; gastrointestinal neoplasms; digestive system surgical procedures; psychotherapy; treatment outcome.

Introduction

Trichobezoars are foreign bodies, concrements, composed of hair, which commonly occur in the stomach, but can extend through pylorus into duodenum and lower parts of the small intestine. They are almost always combined with trichotillomania and trichotillophagia or other psychiatric disorders. Trichotillomania belongs to the group of obsessive-compulsive disorders. It occurs almost exclusively in girls and younger women.

Human hair is resistant to digestion. Over time, continual ingestion of hair, together with food and mucus, leads to trichobezoar formation. The most common presenting symptoms of trichobezoar include palpable abdominal mass, vomiting and noticeable hair loss. It should be suspected in young women suffering from trichotillomania (pulling hair) and trichotillophagia (swallowing hair). Continuous ingestion of hair leads to the enlargement of trichobezoars with possible complications such as erosion, gastric ulcer, perforation of the stomach and small intestine.

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intussusceptions, enteropathy with protein loss, pancreatitis, and even lethal outcome.  

Rapunzel syndrome is an uncommon presentation of trichobezoar, involving strands of swallowed hair extending as a tail through the small intestine, beyond the stomach.

**Case report**

A 19-year-old female patient was admitted to the Clinic for Gastroenterology and Hepatology of the Clinical Centre Niš, with stomach pain, heartburn, nausea, weight loss and vomiting. These symptoms began two months prior to admission and worsened over time.

The patient had no history of prior diseases. Family medical history showed mother’s death from lung tumor.

The patient’s father described his daughter as moody after her mother’s death, with the habit of pulling hair.

After clinical inspection, hair loss and evident asymmetry in epigastric region of the abdomen were detected.

Abdominal palpation revealed a hard, tender, well-defined mass of approximately 10 × 5 cm in the epigastric region, extending into the right upper quadrant.

Laboratory findings revealed hypochromic anemia with hemoglobin levels of 116 g/L, hematocrit 34.7%, iron 1.5 μmol/L, reactive thrombocytosis (407 × 10⁹/L) and hypoproteinemia of 65 g/L. There was no electrolyte disturbance and liver functions, serum amylase, urea and creatinin were normal.

Plain abdominal radiography showed a certain amount of gas and intestinal content in the colon, whereas chest X-ray was normal.

Abdominal echosonography was done using a Siemens Acuson X 300, with multi frequency abdominal probe from 2 to 5 MHz.

Abdominal echosonography showed a homogeneous, echogenic mass, about 10 cm in diameter, below the pancreas. The ultrasonography of the liver, gallbladder, pancreas and spleen were normal.

Endoscopy was performed by using a Pentax video gastroscope. Endoscopy revealed a tumor mass in the stomach made up of hair, food residues and mucus completely filling the stomach, extending through the pylorus and blocking passage of the endoscope (Figure 1). Endoscopically, extraction was considered inappropriate due to the size of trichobezoar.

Computed tomography (CT) scan (Figure 2) showed a non-homogeneous, oval and large mass with a hazy outline that filled the entire stomach and the first part of duodenum. There were air pockets and the mass showed no contrast staining. It was surrounded by a thin rim of contrast material.

A psychiatrist was consulted and he claimed that a patient was apsychotic, with prominent depressive symptomatology, followed by a high level of anxiety, starting after her mother death. The psychiatrist recommended further psychiatric treatment.

The patient was transferred to the Surgical Clinic, where she was operated.

![Fig. 1 – Endoscopy revealed a tumor mass made up of hair in the stomach](image1)

![Fig. 2 – Computed tomography (CT) scan showed a non-homogenous oval and large mass with a hazy outline that filled the entire stomach and the first part of duodenum](image2)

An exploratory laparotomy through an upper midline abdominal incision was performed. Gastrotomy was done and trichobezoar was extracted (Figure 3). A trichobezoar completely filled the stomach and the first part of the duodenum.

![Fig. 3 – A gastrotomy was done and a trichobezoar was extracted](image3)
The patient was discharged with no recurrence of the symptoms, referred to the Psychiatry Department for psychiatric treatment.

Discussion

Bezoars are concrements composed of hair, animal or plant fibres, minerals, medicaments etc., accumulating in gastrointestinal tract, most often in the stomach and rarely in the small intestine. Bezoars composed of hair or hair-like fibres are called trichobezoars.

Trichobezoars, unlike other bezoar types, most frequently occur in patients with psychiatric disorders, trichotillomania and trichotillaphagia. They usually appear with signs and symptoms due to a mass in the stomach and may rarely extend to the small bowel as a tail (Rapunzel syndrome). Trichotillomania is a behavioral disorder which implies compulsive hair pulling. Hair is commonly pulled out from the scalp but also eyelashes, eyebrows, pubic region or some other body parts. Trichotillomania can lead to alopecia. This disorder begins in the earliest childhood or adolescence.

Five to 18% of patients who suffer from trichotillomania develop trichotillaphagia (swallowing hair), which leads to potentially serious complication, trichobezoar formation. Approximately 37.5% of patients suffering from trichotillaphagia will form trichobezoar.

Rapunzel syndrome is a rare form of trichobezoar, first described by Vaughan in 1968, as a tail-like extension of gastric trichobezoar into the small intestine. In 1999, Dahshaug reported 11 cases of Rapunzel syndrome, whereas in a recent literature review in 2007 only 24 cases were found.

For a long time trichobezoar can be asymptomatic or be manifested by epigastric anxiety (80%), abdominal pain (70%), nausea and vomiting (38%), asthenia with weight loss (33%). The first manifestations of bezoars are sometimes gastrointestinal complications such as: ulceration bleeding, mechanic intestinal obstruction, perforation of the stomach or small intestine with peritonitis or subphrenic abscess, digestive fistula, acute pancreatitis or cholestasis due to obstruction of Vater’s ampulla in Rapunzel syndrome. The most common complication is intestinal obstruction. Perforation and peritonitis result in death in 30% of cases.

An abdominal mass in the epigastrium is the most common sign. In our case, the size of abdominal mass led to asymmetry of the abdomen and was observed during examination. Laboratory findings confirm hypoproteinaemia and hypochromic anaemia as the most common manifestations of malabsorption syndrome.

Various imaging modalities have been recommended for detection of trichobezoars. Plain abdominal radiography is helpful in the diagnosis of intestine obstruction, but contributes little to the confirmation of trichobezoars. Echophonographic trichobezoar looks like a hyperechogenic mass, with wavy edges. The presence of multiple acoustic interfaces created by trapped air and food limits the ultrasonography of the trichobezoars.

Esophagogastroduodenoscopy is a method of choice for diagnosing trichobezoars. The typical colour of trichobezoar at endoscopy is black. It allows the clinician to distinguish between phytobezoars and trichobezoars. This is very important because treatment depends on the nature of a bezoar. CT scan is also very useful in diagnosing trichobezoars. Trichobezoar is presented on CT as well-defined ovoid intraluminal heterogeneous mass, occupying almost the entire lumen. CT scan has shown a hypodense lesion in the stomach with a mesh-like pattern. Oral contrast is sparse within the mesh, though prominent around the margins. The presence of a tail in the small intestine is reflected by small areas of hypodensity.

Small trichobezoar can be removed endoscopically from the stomach, but they are rare. Previous to removal, trichobezoar first must be fragmented by the biotic device or by using bezotomes. Most cases fragmentation is not possible due to the size and content, as in the presented patient. Fragmented large trichobezoars can migrate through the pylorus, causing obstruction of distal parts of the small intestine. Extraction of fragmented parts involves a large number of repeated insertions of endoscope, which can lead to esophagitis, ulcerations, even perforation of esophagus.

Surgical treatment is indicated in most trichobezoar cases, mostly for their size, but also composition. Nirasawa et al. first described laparoscopic removal of trichobezoars. A successful laparoscopic removal requires significantly longer operation time as compared to conventional laparotomy.

Laparotomy is a therapy method of choice, with respect to its success, possibility of careful examination of the whole gastrointestinal tract (stomach and intestines), as well as low level of complications.

In prevention of recurrence of trichobezoar, psychiatric treatment of trichotillomania and monitoring of the patient is very important. Common treatment includes medicamental and cognitive behavioral therapy. Selective serotonin reuptake inhibitors are medications with the highest efficacy in the treatment of trichotillomania.

Habit-reversal training is an initial behavioral therapy.

Conclusion

Trichobezoar should be differentially diagnostically taken into consideration in younger women with abdominal pain, nausea, vomiting, palpable abdominal mass and psychiatric disorders.

Therapy modalities depend on trichobezoar size and possible complications, but most of the cases demand surgical treatment. Each patient has to be submitted to psychiatric treatment and regular control.

This rare case demonstrates the importance of understanding patients in the context of their life situation.

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