



კლინიკური შემთხვევის აღწერა: 15 წლის გოგონა „რაპუნცელის სინდრომით“

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აბსტრაქტი

რაპუნცელის სინდრომი არის ტრიქობეზოარის უჩვეულო ფორმა, რომელიც გვხვდება პაციენტებში ფსიქიატრიული დარღვევების, ტრიქოტილომანიისა და ტრიქოფაგიის ანამნეზით, რაც იწვევს კუჭის ბეზოარის განვითარებას. ის ყველაზე ხშირად გვხვდება ბავშვებსა და მოზარდებში. დიაგნოზი ჩვეულებრივ კეთდება მხოლოდ ტრიქობეზოარის გამოვლენის შემდეგ. მკურნალობა მოიცავს ტრიქობეზოარის ქირურგიულ მოცილებასა და ფსიქიატრიული დაავადების მკურნალობას ფსიქოთერაპიით.

კლინიკური შემთხვევა: 15 წლის გოგონა უჩიოდა ზოგად სისუსტეს, მადის დაკარგვას, გულისრევას, პირღებინებას, კუჭში სიმძიმის შეგრძნებას. ჩივილები რამდენიმე კვირა გაგრძელდა, მათი ინტენსივობის მკვეთრი მატების გამო გოგონამ მიმართა კლინიკას. ჩაუტარდა გასტროსკოპული გამოკვლევა. გამოკვლევამ აჩვენა კუჭში დიდი უცხო სხეული (ტრიქობეზოარი), რომელიც ორგანოს დიდ ნაწილს იკავებდა. უცხო სხეულის ზომის გამო მისი ენდოსკოპიური მოცილება იყო შეუძლებელი.

მუცლის კედლის წინა-შუა ლაპაროტომიის შემდეგ მოხდა კუჭის წინა კედლის მობილიზება და გასტროტომია. კუჭიდან ამოიღეს დიდი 70-80 სმ სიგრძის ტრიქობეზოარი, რომელიც გადავიდა თორმეტგოჯა ნაწლავში და წვრილ ნაწლავში. ჰისტოპათოლოგიური გამოკვლევით გამოვლინდა შავი და ყვითელი ფერის თმა.

საკვანძო სიტყვები: „რაპუნცელის სინდრომი“, მიზეზები, მკურნალობა.

A Case Report: “Rapunzel syndrome” in a 15-year-old girl

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Abstract

Rapunzel syndrome is an unusual form of trichobezoar that occurs in patients with a history of psychiatric disorders, trichotillomania and trichophagia, which leads to the development of gastric bezoar. It is most common in children and adolescents. The diagnosis is usually made only after the trichobezoar is detected. Treatment includes surgical removal of the trichobezoar and treatment of the underlying psychiatric illness with psychotherapy.

Case report: A 15-year-old girl complained of general weakness, loss of appetite, nausea, vomiting, feeling of heaviness in the stomach. The complaints lasted for several weeks, due to a sharp increase in their intensity, the girl turned to the clinic. A gastroscopic examination was performed. Examination showed a large foreign body (trichobezoar) in the stomach, occupying a large part of the organ. Due to the size of the foreign body, it cannot be removed endoscopically.

After the anterior-middle laparotomy of the abdominal wall, the anterior wall of the stomach was mobilized and a gastrotomy was performed. A large 70-80 cm long trichobezoar was evacuated from the stomach, which had passed into the duodenum and small intestine. Histopathological examination revealed black- and yellow-colored hairs.

Keywords: “Rapunzel syndrome”, causes, treatment.

Introduction

Rapunzel syndrome is a rare condition in which trichobezoar – a large hair ball – is present in stomach and sometimes extends in small intestines. It is named after the Brothers Grimm’s fairy-tale character Rapunzel, who has long hair. Syndrome is caused by psychic disorders: trichotillomania – the irresistible desire to pull out one's own hair and trichophagia – the repeated ingestion of hair. There have been approximately 64 cases (fig 1) of Rapunzel syndrome worldwide since 1968. It is more common in female, because they usually have long hairs, but surprisingly first ever reported case was a teenager boy in 1979, who was eating his sister’s hair.

The typical age range affected from this syndrome is between 4 and 19 years of age.

What is a bezoar in general? Bezoar is a collection of undigested or partly digested materials, that eventually due to its size can cause blockage in the stomach or intestines. There are several types of bezoars, such as phytobezoar (indigestible food fibers like cellulose), Lactobezoar (concentrated milk formula), Pharmacobezoar (mostly medications which contains aluminum hydroxide gel and sucralfate), food bolus bezoar and most rare and interesting for us trichobezoar (concretions of hair). In adults, bezoar formation occurs more often due to gastroparesis, anatomical anomalies and gastric surgeries causing delay in stomach emptying and are more likely to accumulate foreign bodies. But in younger patients it is more likely to be

caused by some mental disorders. Like I discussed earlier, Trichobezoar is a result of a pathological ingestion of hair, in which remains undigested in stomach. Our stomach is unable to digest hair because of its smoothness. Insignificant amounts of hair would just pass right through your body, but when a person continuously ingests hair, over a period, it interacts with mucus and food particles and forms a mass which eventually gets bigger and in some cases extends through the pylorus into jejunum or even colon. The mucus covering the trichobezoar gives it a glistening shiny surface. Putrid smell is caused by decomposition and fermentation of fats in the interstices. The acidic contents of the stomach denature the hair protein and gives it its black color regardless of the original hair color.

Trichotillomania and trichophagia are psychiatric disorders, the exact causes are not entirely clear. It could be a way of dealing with stress or anxiety, a chemical imbalance in the brain, genetic predisposition, social environment or even changes in hormone levels during puberty. People with trich feel an intense urge to pull their hair out and they experience growing tension until they do. To feel a sense of relief they must pull their hair out. Most people pull out hair from their scalp, but sometimes they pull out from eyebrows, eyelashes, genital area. Affected person usually have bald patches, which have an unusual shape and is presented one side of the head more than other. The occurrence frequency of trichotillomania is estimated to be 0.52.0%, most recent studies found that 1.7% of people aged 16-69 suffer from it, with 30% of them suffering from trichophagia. Trichophagia without trich is hard to detect, until trichobezoar is formed and shows some symptoms.

Rapunzel syndrome has non-specific symptoms that may imitate other diseases and can be difficult to diagnose right away. Symptoms are usually caused by the blockage of trichobezoar. Patients can experience stomach pain, bloating, feeling full, because of that they tend to eat less and lose weight, nausea, acute epigastric pain, halitosis – bad breath, anorexia nervosa, vomiting after meals. These are also the signs of acute abdomen condition and intestinal obstruction. One of the reasons that complicates the diagnosis is that patients do not realize either how important it is for doctors to talk about eating hair or their psychiatric character of the disease is denied by them or their parents.

Case report

Here we present a case report of a 15-year-old girl with Rapunzel syndrome.

A 15-year-old girl complains of general weakness, loss of appetite, nausea, vomiting, feeling of heaviness in the stomach. The complaints lasted for several weeks, due to a sharp increase in their intensity, the girl turned to the clinic. A gastroscopic examination was performed - The esophagus is free of foreign bodies, mucous membrane pink with hyperemic areas. The cardia of the stomach does not close completely. A large foreign body (trichobezoar) is visualized in the lumen of the stomach (Fig 2), which occupies a large part of the lumen of the stomach. Pylorus is outgoing. Duodenal bulb mucosa with hyperemic areas, post bulbar part without pathology. Due to the size of the foreign body, it cannot be removed endoscopically. The patient was consulted by a surgeon, and considering her general condition, an operation was planned to remove the foreign body.

The patient was placed on her back on the operating table. General anesthesia was performed. The patient was approached from the anterior abdominal wall, the abdominal cavity was opened through an anterior median laparotomy in layers. After entering the abdominal cavity, a distended stomach with firm content was seen. To project the entire stomach, the anterior wall of the stomach was mobilized and a vertical gastrotomy was done using electrocautery. The stomach was distended, solid contents are palpated. A large trichobezoar was

evacuated from the stomach, which completely occupied the stomach and part of which was extended to the duodenum and small intestine. Its length was approximately 70-80 cm (about the length of a baseball bat). Suction and irrigation were done. The stomach was sutured in three layers, 1 drainage was placed near the stomach, from the contraperture in the left mesogastrium. After suction and irrigation, homeostasis was achieved and then the abdominal wall was closed. Staplers were used in surgery in place of sutures to close the skin. The patient tolerated the procedure, recovered smoothly and then was sent to the recovery room.

The removed foreign body (Fig3-4) was sent for histopathological examination, trichobezoar was confirmed with black- and yellow-colored hairs.



Figure 1 -2 Removed trichobezoar from a 15-year-old girl with Rapunzel Syndrome

Discussion

Like I already discussed Rapunzel syndrome sometimes does not present any symptoms. but when the symptoms are on hand there are different ways to diagnose it. The patient's history plays a significant role in differentiating Rapunzel syndrome from other diseases that have the same symptoms. The patient may also undergo laboratory tests, including X-ray, CT (Fig5-6), ultrasound. Bezoars in X-rays are seen as a shade or mass usually visible when they are in stomach. In CT-scans they appear as a round-shaped mass with air bubbles with obstruction of the gastrointestinal tract. Trichobezoars could initially be mistaken as a tumor or a cyst. That is why we should perform endoscopic examination for confirming diagnosis of bezoar. Moreover, during the endoscopic procedure, a sample can be taken to confirm the diagnosis by histopathological examination. It is possible to make the correct diagnosis only with a CT-scan and even an X-ray, but it depends on the type, size, and location of the mass. the first goal after confirming trichobezoar is to remove it from patient's body, because otherwise The foreign body will continue to grow in size and various complications will appear as a result, including Obstructive jaundice, Physical blockage in your belly or small intestine, Erosion of the mucus lining in your stomach and small intestine, small bowel perforation,

peritonitis, acute pancreatitis. trichobezoars can be removed via endoscopy, laparotomy, or laparoscopy. only small trichobezoars may respond to endoscopic fragmentation and vigorous lavage, 75% of attempted laparoscopic surgical extractions were successful. However, in our case due to trichobezoar size the best way to remove it from the patient body was laparotomy followed by gastrotomy.

But removing trichobezoar unfortunately does not mean that problem is solved. Rapunzel syndrome is a complication of psychic disorders, such as trichophagia and trichotillomania. So, when an emergency is handled patients should undergo psychotherapy. Trichotillomania (TTM) is classified as a disorder of the obsessive-compulsive group. It is one of body-focused repetitive behavior disorders and there is a noticeable relation between TTM and anxiety disorders. In patients with TTM hair-pulling and eating is a habit, therefore habit reversal training as part of behavioral therapy is extremely helpful. In children, hypnotherapy can be used, while in adults cognitive-behavioral therapy is the most common form of treatment. Alongside psychotherapy there are a few alternative forms of pharmacological approaches including SSRIs, tricyclic antidepressants, and atypical neuroleptics, however, data are not fully consistent. Surgery does not mean recovery. Unless the patient undergoes psychotherapy, the trichobezoar will form repeatedly until the psychological problem is resolved.



Figure 5 CT scan demonstrating trichobezoar occupying the extent of the stomach.

Figure 6 Barium meal showing filling defect due to trichobezoar

Conclusion

In conclusion, Rapunzel syndrome is a complication of rare mental disorders such as trichotillomania and trichophagia. trichobezoars can be life threatening and surgical removal must be done. But the main problem is not the complication but the disease. The patient should undergo psychotherapy to stop the bad habit of eating hair. Mental illnesses are not easy to manage, but with a good psychologist and the desire of the patient, everything is achievable.

