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A report of two cases

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Paediatric gastric trichobezoar: the Rapunzel syndrome. A report of two cases

Rapunzel syndrome is a rare case of bowel obstruction resulting from hair ingestion (Trichobezoar). The obstruction can occur in any level of intestinal tract, but usually the stomach is primary involved. This syndrome is usually reported in patients affected by Trichotillomania or Pica syndrome, an obsessive-compulsive disorder that are characterized by an irresistible need to eat body hairs or non-digestible substances [1]. When bowel obstruction occurs, it may be treated conservatively, but sometimes surgery is required. We reported two cases of Rapunzel Syndrome in two pediatric patients with different clinical presentation. Both patients were initially treated conservatively but eventually they underwent surgery.

KEY WORDS: Bowel obstruction, Rapunzel syndrome, Trichobezoar

Introduction

The term "*Bezoar*" means a foreign body that occurs in the digestive tract after the ingestion of non-digestible substances [2].
The trichobezoars occur in patients affected by mental illnesses. An example of these mental illnesses is Trichotillomania, where patients compulsively swallow their hairs without consciousness. Another eating behavioural disorder is the Pica disorder that is characterized by non-nutritive and non-digestible substances ingestion. The bezoars become clinically detectable when they prevent bowel emptying. The most frequent clinical presentations are abdominal pain, vomiting and constipation which can evolve to bowel perforation and peritonitis [2].
Possible treatments for Trichobezoars are the follows:
Chemolysis: cellulase is utilized on patients with soft symptoms. The dose is between 3 and 5mg in 300-500ml of water daily for 2-5 days. Metoclopramide could be administered to improve the bowel mobility (10mg daily) [3,4].
Endoscopic removal: advised for patient with insoluble bezoars. The bezoars are fragmented with pliers, loops, jet spray, argon plasma coagulation, or laser that could break the bezoars and let their extraction [5,6]. Surgery: it is necessary only if previous approaches have failed. [7]

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Case Report

Case n. 1

We report two cases of Rapunzel Syndrome with different clinical presentation but eventually both surgical treated.
In April 2019, the patient M.S., female, 13 years old (BMI 14.2), presented to Emergency Room complaining of abdominal pain, vomiting and sickness for 3 days. Her medical and surgical histories were unremarkable.
The mother of this young patient reported that the girl probably started to suffering from trichotillomania and Pica disorder when she was about 9-10 years old.
Physical examination showed a swelling abdomen with epigastric discomfort and tenderness under deep palpation. Blumberg sign was positive.
Blood test revealed leucocytosis (WBC 16.630/mmc with Neutrophils 86%, Lymphocytes 10%), negative CRP and Hb 12.5g/dl.
X-ray abdomen showed air-fluid levels and relevant colic faecal impaction (Fig. 1).
Abdominal CT scan was performed and revealed important gastrectasia, while gastric lumen contained different materials mixed with air. Same materials were founded in colic and jejunal tract (Fig. 2).
The patient underwent gastroscopy, performed in the operating room under general anaesthesia. During the gastric examination, a large cylindrical bezoar was found it consisting of hair, plastic and other inorganic material, occupying almost completely the gastric lumen. The bezoar was not removable endoscopically (Figs. 3, 4). So we decided for surgical approach. Midline mini laparotomy was performed and the bezoar was faster appreciated by gastric palpation.
We approached through vertical anterior gastrotomy and gradually removed the bezoar composed of hair, sponges and other plastic material of different shape and nature, with a total weight of about 2.5 kg (Figs. 5, 6).
Once the stomach was evacuated the duodenal patency was confirmed. Nasogastric-tube was placed under visual control and double layer sutures of the gastrotomy were performed in a double layer suture. A second one bezoar was founded in the middle- ileum and was treated by enterotomy and removing the foreign body (Figs. 7, 8). Transversal suture of the enterotomy was performed in two layers.

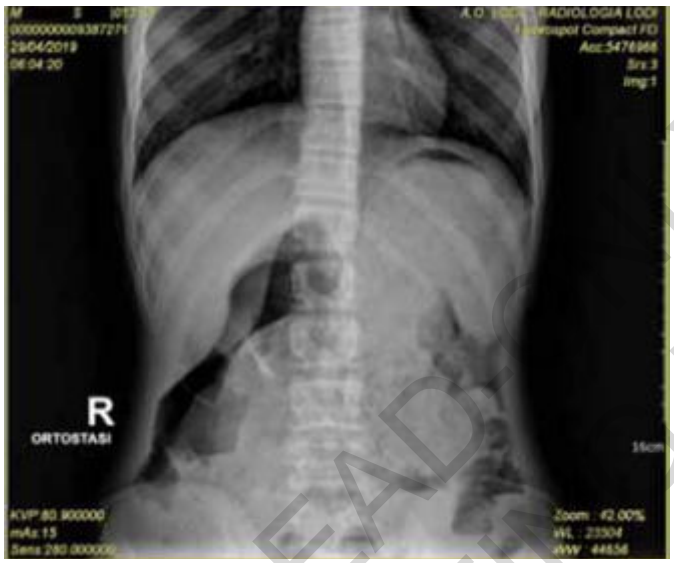

Fig. 1: Xray Abdomen.

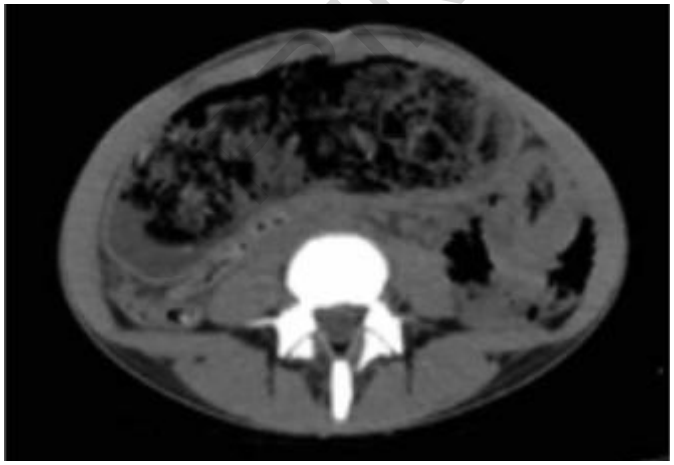

Fig. 2: TC upper Abdomen.

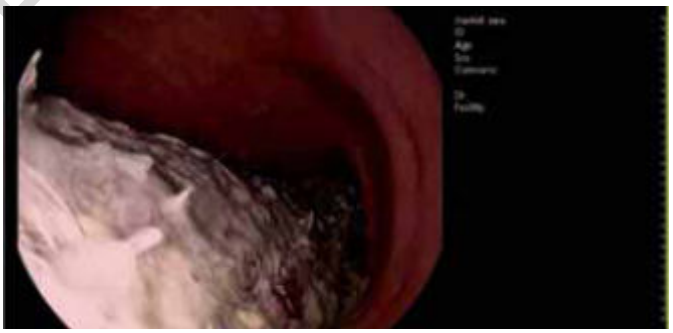

Fig. 3: Gastric bezoar at gastroscopy.

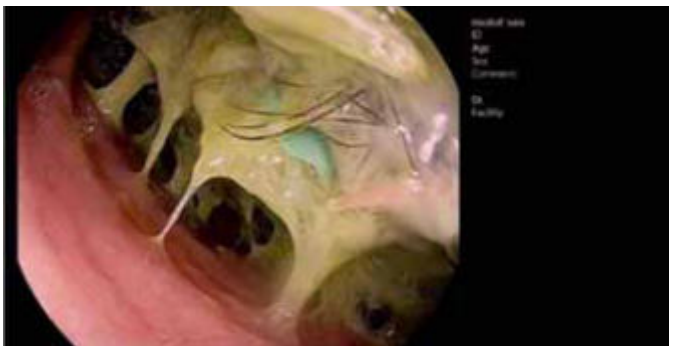

Fig. 4: Gastric bezoar at gastroscopy.

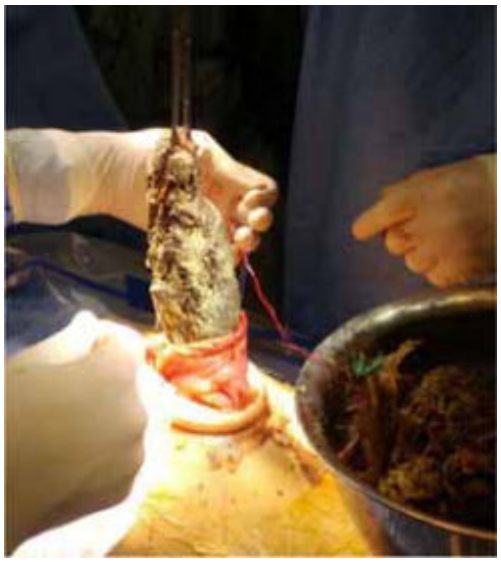

Fig. 5: Removing part of gastric Bezoar.

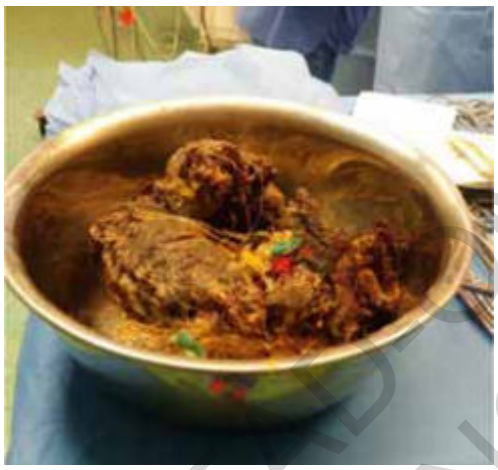

Fig. 6: Gastric bezoar.

During the hospitalization, the patient underwent a child neuropsychiatrist examination which confirmed the patient's psychological disorder and the reasons for her gestures.
The hospitalization was uneventful and the young patient was discharged after twelve days.

Case n. 2

The second patient, MVFG, was a young female, 15 years old, BMI 19.2, who in September 2019 was admitted for anemia probably due to hypermenorrhea she started treatment based on oral estroprogestinic, in October 2019 and she returned for check and blood test that showed a worsening of the anemization (Hb 8.4 g/dL). The young girl complaining also of abdominal pain and

Fig. 7: Removing ileal bezoar.

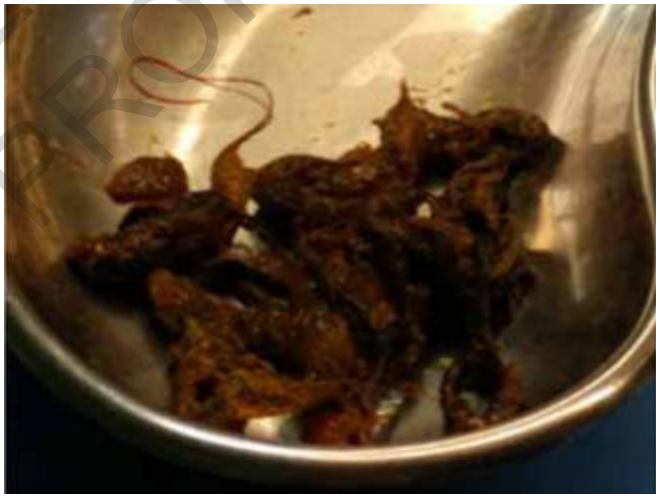

Fig. 8: Ileal bezoar.

dyspepsia. Stool test was performed and showed hair residues in the stool.
X-ray abdomen documented the presence of thin radiopaque material mixed with fine air bubbles in the stomach. We decided to perform gastroscopy that revealed the presence of a trichobezoar extended from the gastric body to the second portion of the duodenum. A child's neuropsychiatric examination was performed and confirmed the diagnosis of trichotillomania. We tried a second gastroscopy in the operating room to remove the trichobezoar, but after failure, we decided for surgical approach (Fig. 9).

We performed first a midline minilaparotomy, then an anterior gastrotomy and removed a large trichobezoar of about 700 g (Figs. 10, 11, 12, 13, 14). Another trichobezoar was found and removed in the ileum, starting to get complicated in bowel invagination.
The hospitalization was uneventful and this patient was discharged after ten days.

Discussion

Rapunzel syndrome is a rare case of bowel obstruction due to the presence of a trichobezoar [8-9] About 70% of people with this condition are women and under 20 years old. It is thought that this is due to the composition of the hair, longer than men are ones and above all often curly that can favour the aggregation of the hair at the level of the stomach thus forming the trichobezoar [1].

Fig. 9: Trichobezoar at gastroscopy.

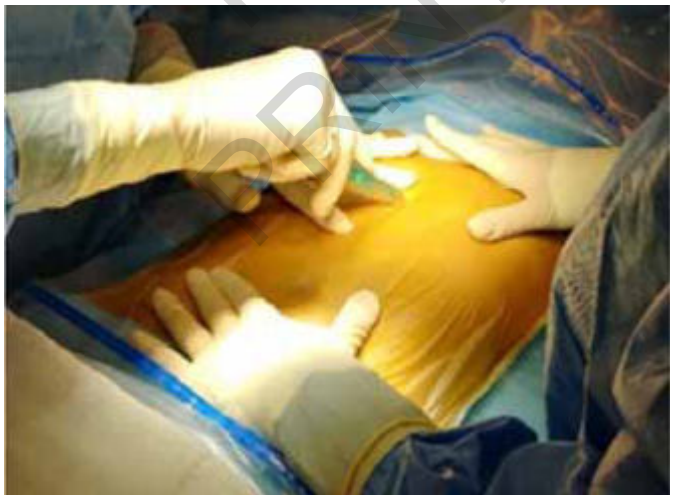

Fig. 10: Minilaparotomy anterior gastrotomy.

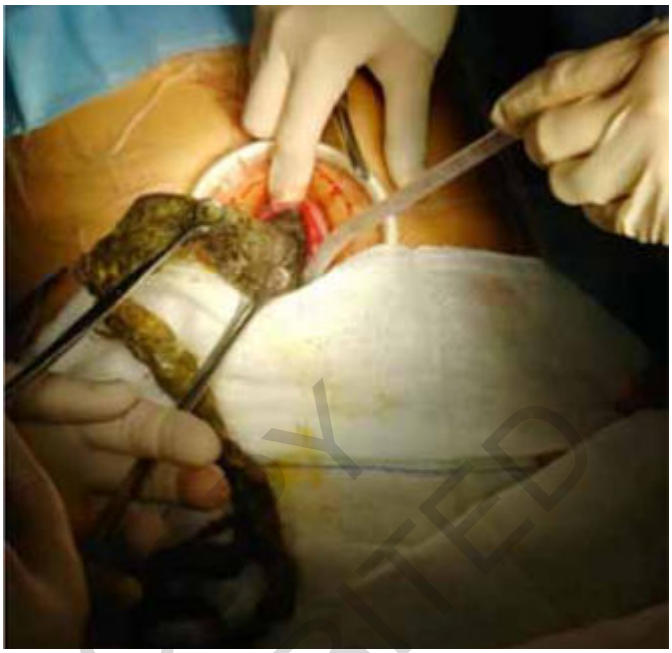

Fig. 11: Removing gastric trichobezoar.

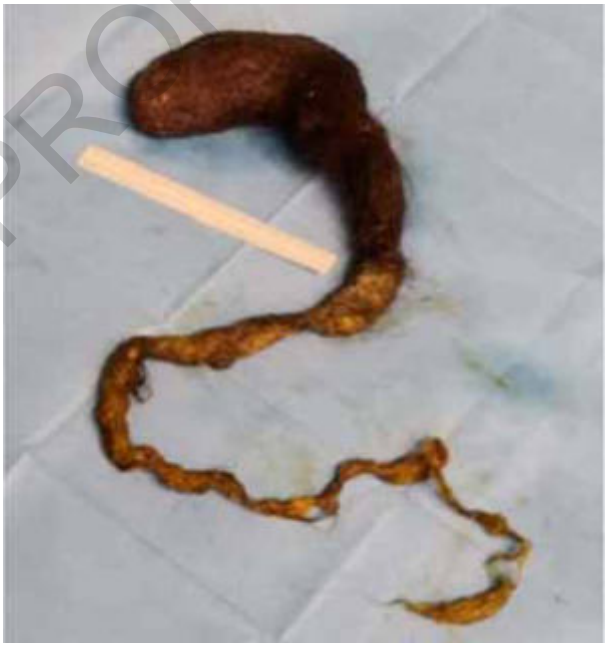

Fig. 12: Gastric/Duodenal trichobezoar.

Rapunzel syndrome is associated with disorders such as Trichotillomania and Pica disorder. Trichotillomania is a psychiatric disorder when patients used to treat their hair as an altered behaviour. Often subjects tend to "play" with torn hair, for example by nibbling the root or resting the hair on the lips, thus risking ingesting it [10]. One study showed that out of 24 people suffering from this disorder, 25% of them developed a trichobe-

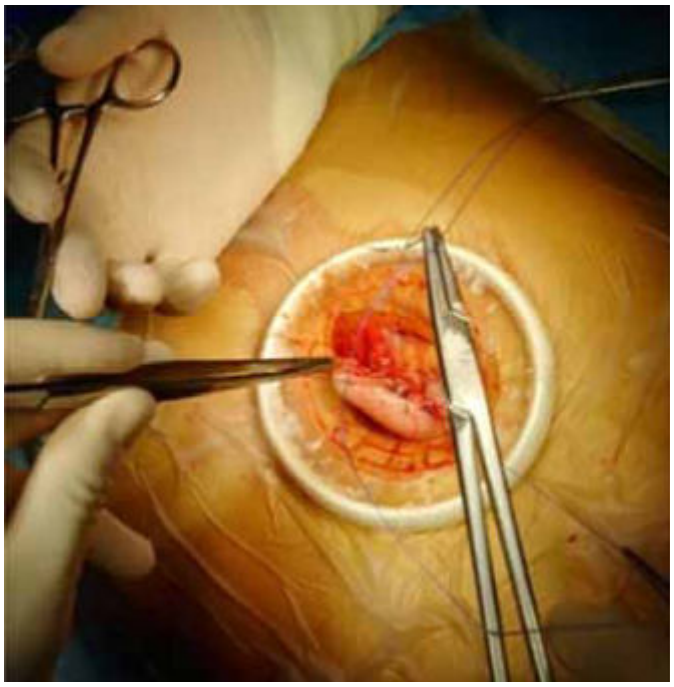

Fig. 13: Double layer sutures.

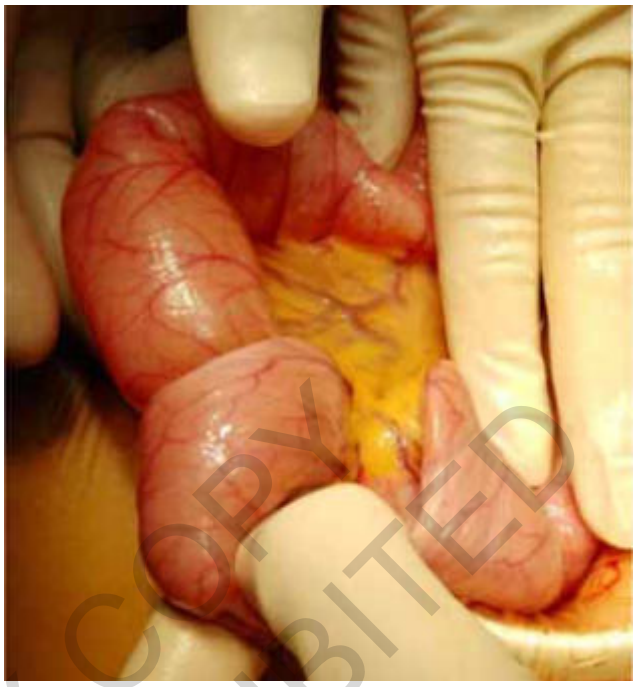

Fig. 15: Ileal invagination.

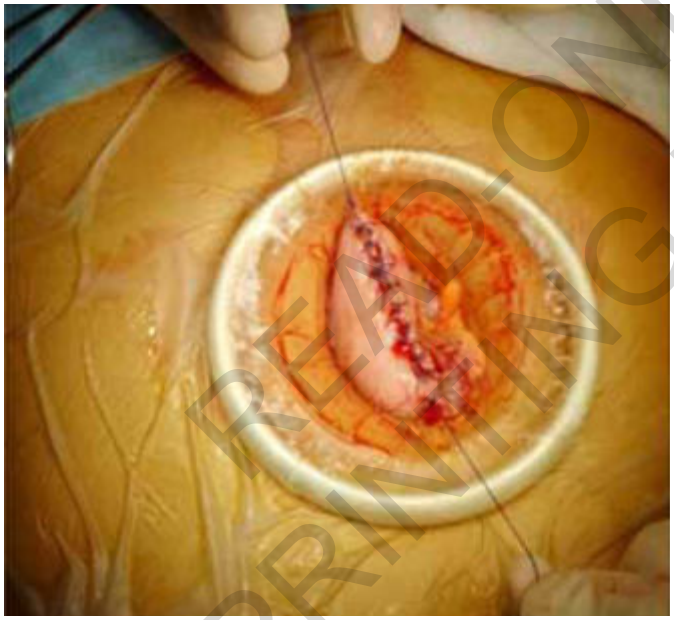

Fig. 14: Double layer sutures.

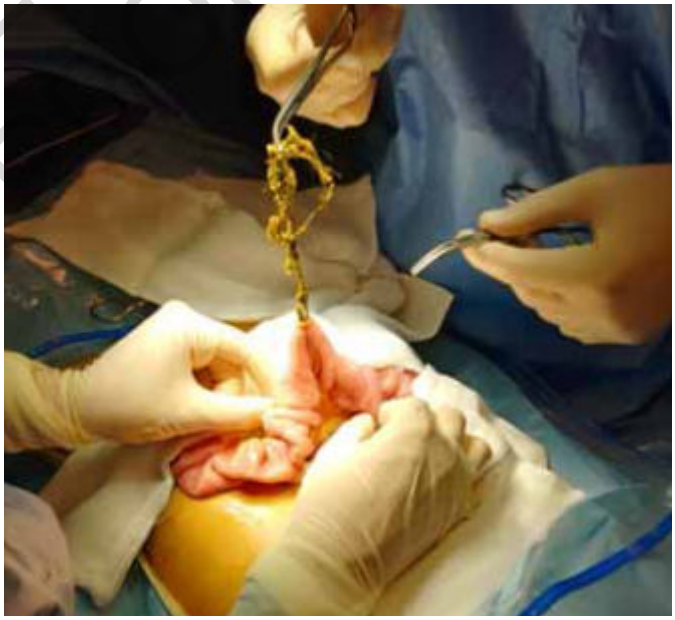

Fig. 16: Removing ileal trichobezoar.

zoar in their stomach. [1] Pica disorder, on the other hand, means a disorder of eating behaviour characterized by the ingestion of non-nutritive substances, which are indigestible by gastric juices and which can lead to an occlusive condition [11].

Rapunzel syndrome shows signs and symptoms of an obstruction bowel picture such as pain, abdominal distension, nausea and sickness, closed alveus to faeces and air.

The diagnosis is based on:

Clinical objectivity: distension of the abdomen, resistance of the abdominal wall, alternation of areas of tympanism (presence of air for dilated loops) and areas of dullness (presence of liquid), presence or absence of peristalsis and its characteristics [12].

Blood tests: complete blood count, hematocrit, electrolytes measurement, acid-base balance study.

Instrumental exams: endoscopic and radiological, such a RX and CT [13].

Although the presence of trichobezoar can be treated by medical-chemical or endoscopic ways, for Rapunzel Syndrome, it is almost exclusively suggested a surgical treatment that consists of a laparotomy with removal of the trichobezoars present at the gastric and bowel level which have led to the occlusive condition [7].

A study was carried out which investigated and selected 88 cases of this syndrome. According to this review, the most common symptoms of this syndrome were weight loss (26%), anemia (17%) and bowel obstruction (32%). About 40% of patients are under the age of 10, 39% are under 20 and 17% are aged between 20 and 30, while only 3% are over 30 years old [1].

Conditions associated with this syndrome are malnutrition leading to weight loss (BMI <18.5), Vitami B12 deficiency, Vitamin D deficiency and other minerals, with evidence of hypochromic microcytic anemia from iron deficiency [16,17].

Both clinical cases came from an epidemiological point of view in the most represented percentages, being both females aged between 10 and 20 years [1]. Both patients had psychiatric disorders such as trichotillomania which is the cause of Rapunzel Syndrome. The difference between the two cases is the clinical presentation: the first clinical case provided a clear and typical situation of bowel obstruction, associated with malnutrition (BMI 14.2 - hypoalbuminemia) and disorders of the hydroelectrolyte balance (initial hyponatremia). The second clinical case instead was a Rapunzel Syndrome linked to the malabsorption anemization related to the presence of trichobezoar which over the time could have led to bowel obstruction, also due to the presence of ileal invaginations due to the presence of bezoar itself.

The treatment depends on the patient's appearance and the size of the bezoar. In the most complicated cases, with a large bezoar, surgery is required, especially in patients who do not respond to conservative treatments who must be operated. During surgical removal, the gastrointestinal tract must be fully examined to look for additional bezoars [7].

Along with surgical treatment, psychiatric treatment is important for treating psychiatric disorders that are the causes of Rapunzel syndrome. The psychiatric treatment is based on behavioural and pharmacological therapies such as selective serotonin reuptake inhibitors, antipsychotics, tricylic antidepressant and stimulants [18].

For both patients, the removal of trichobezoar occurred through gastrotomy and enterotomy, demonstrating that the treatment of Rapunzel Syndrome is almost exclusively surgical, due to the considerable size of the trichobezoars that extend from the stomach to the small bowel.

Conclusions

Rapunzel Syndrome is a rare condition of intestinal obstruction characterized by the presence of a trichobezoar at the gastric level that extends to the small bowel. The disease is found prevalently in patients who present psychiatric comorbidities (obsessive-compulsive syndrome such as trichotillomania and pica disorder).

As demonstrated in literature, the treatment of this pathology is almost exclusively surgical. These complicated cases require an adequate clinical diagnosis and a multidisciplinary approach involving dieticians, nutritionists, pediatricians, psychiatrists, psychologists and general surgeons, to investigate the underlying causes and improve patient's outcomes.

Riassunto

La Sindrome di Raperonzolo è una rara causa di occlusione intestinale correlata alla presenza di un tricobezoar che si localizza a livello gastrico, il quale si estende anche a livello intestinale. I tricobezoar sono corpi estranei che si formano nell'apparato digerente a seguito dell'ingestione di capelli.

È una patologia che si riscontra in particolar modo in pazienti affetti da disturbi psichiatrici quali la tricotillomania, disturbo ossessivo compulsivo in cui i pazienti tendono ad ingerire i propri capelli, e il picacismo, disturbo dell'alimentazione in cui i soggetti tendono a ingerire sostanze inorganiche-indigeribili.

Il quadro occlusivo dovuto alla presenza di tricobezoar può essere trattato in modo conservativo, ma in casi gravi è necessario l'intervento chirurgico per la rimozione del corpo estraneo.

In questo Case Report si discuterà della Sindrome di Raperonzolo e in particolare si discuterà di due casi clinici correlati a tale sindrome. I casi clinici riguardano due giovani pazienti con Sindrome di Raperonzolo, giunte alla nostra osservazione presentando quadri clinici diversi: un quadro di anemia sideropenica da malassorbimento e un quadro franco di occlusione intestinale da tricobezoar.

Per gli scarsi risultati del trattamento conservativo, entrambe le pazienti sono state sottoposte ad intervento chirurgico di gastrotomia ed enterotomia in regime d'urgenza per la rimozione dei tricobezoar causanti i diversi quadri clinici.

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