Rapunzel Syndrome: An Infrequent Cause of Paediatric Mechanical Occlusion

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Abstract

Despite numerous cases of trichobezoars reported in the literature, few Rapunzel syndromes have been described. Rapunzel syndrome is a rare case of bowel obstruction resulting from hair ingestion (trichobezoar). The obstruction can occur in any level of the intestinal tract. This syndrome is usually reported in patients affected by trichotillomania and trichophagia. We reported a case of Rapunzel syndrome in a 5-year-old girl diagnosed on an abdominal computed tomography scan and confirmed during surgery.

Keywords: Bowel obstruction, Rapunzel syndrome, trichobezoar

Introduction

The bezoar is defined as any ingested but undigested substance accumulating as a solid mass in the digestive lumen, most often gastric.[1,2] The extension in the small intestine is quite exceptional, and represents a source of threatening complications, thus realising Rapunzel syndrome,[3] thus representing 2%–3% of all the aetiologies of small intestinal obstruction.[4] Its diagnosis is mainly based on imaging.[5] We report the case of a gastric and small-bowel trichobezoar complicated by intestinal obstruction in a 5-year-old child.

Case Report

We report the case of a 5-year-old girl, with no previous medical, surgical or psychiatric history, admitted to the emergency department with pain in the abdomen, nausea, vomiting and absence of stools for approximately 3 days. On physical examination, she was conscious, well oriented, afebrile, with normal vital signs. Her abdomen was distended and tender. Bowel sounds were absent. X-ray findings air-fluid levels in the small bowel. An abdominal ultrasound was performed [Figure 1]; it demonstrated an endoluminal hyperechoic arch in a small sub-umbilical loop generating a posterior shadow cone with distension of the upstream hailstones. Slightly abundant fluid effusion between the curls was also found. We completed the assessment by an abdomino-pelvic computed tomography (CT) scan before and after injection of iodinated contrast product (PDC I) [Figure 2]; she objectified a flat colon, a distension of the loops

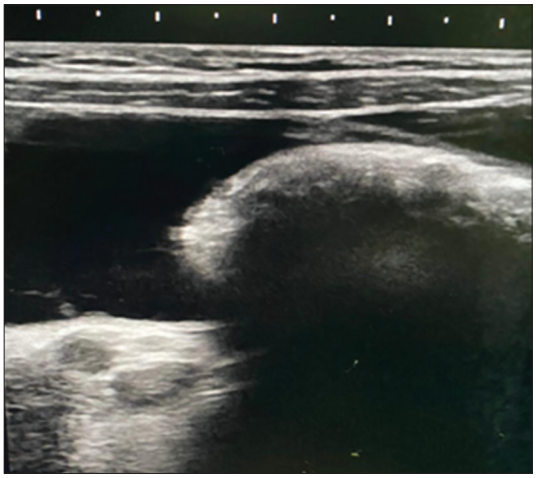


Figure 1: Abdominal ultrasound: Intraluminal material of an ileal loop in the form of a hyperechoic crescent with posterior shadow cone

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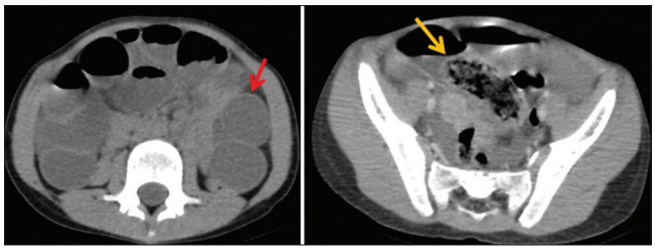


Figure 2: Abdomino-pelvic computed tomography scan: Distension of the small-bowel (red arrow) upstream of a hairless, oval endoluminal mass, of heterogeneous density, surrounded by a dense border closing containing air bubbles, unchanged after contrast in relation to the trichobezoar (yellow arrow)



Figure 4: The consequence of hair pulling in the frontal region of the scalp hair

measuring 30 mm in diameter upstream of a transition zone corresponding to an intraluminal formation in a pelvic ileal loop, oval, well delimited by a spontaneously dense border, containing trapped air bubbles. This material did not exhibit parietal attachment and did not enhance after injection of PDC I. A similar picture was identified in the stomach [Figure 3]. Analysis of the remainder of the abdominal cavity revealed a low abundance of peritoneal fluid effusion without signs of ischaemia in the intestinal wall or pneumoperitoneum. Rapunzel syndrome was suspected. The re-interrogation has rediscovered the notion of trichotillomania [Figure 4] and trichophagia comforting the diagnosis of trichobezoar. The patient was urgently operated; the procedure consisted of removing the small intestine obstacle and the gastric trichobezoar by gastrotomy [Figure 5]. A child psychiatric consultation has been planned.

Discussion

Rapunzel syndrome was first reported by Vaughan in 1968 and takes its name from the Brothers Grimm's tale 'Rapunzel' written in 1812, which tells the story of a princess with long blonde hair imprisoned in a tower.[6] It is defined by a gastric trichobezoar extending into the small bowel or even the transverse colon through the pylorus and duodenum.[3] A classification into three grades according to intestinal obstruction has been reported: the first grade corresponds to partial obstruction of the intestine, the second to complete obstruction and the third to complete obstruction with

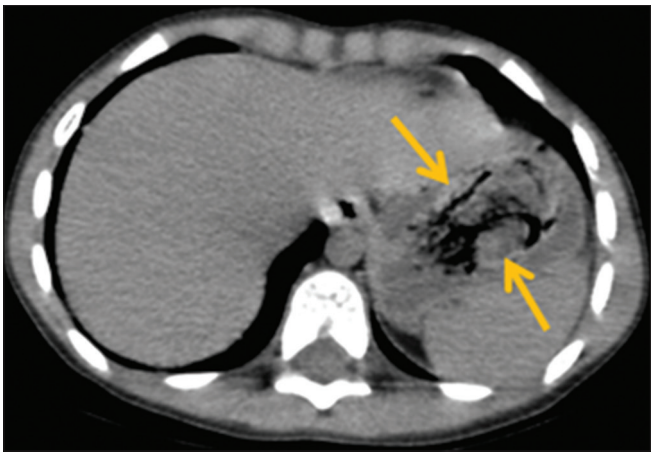


Figure 3: Abdomino-pelvic computed tomography scan: Gastric trichobezoar (yellow arrows)



Figure 5: Gastrotomy and the surgical specimen of the gastric trichobezoar

neurosis, perforation and peritonitis.[7] The female sex is the most affected (90% of cases), and the age of onset of trichobezoar is 80% <30 years with a peak of frequency between 10 and 19 years.[2,5] Psychological disorders are sometimes found. It is most often a young girl who chews and swallows her own hair or fibres taken from carpets, clothes or plush (trichotillomania with trichophagia), but in paediatrics, only 9% of children with trichobezoar would have real psychiatric problems.[6] Revealing clinical symptomatology of bezoars is varied and non-specific: abdominal pain, appetite disturbances, nausea, vomiting, asthenia with weight loss, transit disorders such as diarrhoea or constipation and even an unbearable smell of breath.[2-7] However, physical examination and questioning can guide the diagnosis. Thus, a range of alopecia can reveal an eating disorder. The interrogation must therefore be careful and can sometimes reveal, as in our case, trichophagia.[7] In extreme cases, it can be diagnosed following serious complications, the main ones being upper gastrointestinal bleeding on ulceration, peritonitis by digestive perforation with peritonitis or fistula, intestinal obstruction, acute intussusception, acute pancreatitis or cholestasis by obstruction of the ampulla of Vater.[1,2] In our observation, the mode of revelation was a small-bowel occlusion. Ultrasound shows the bezoar as an arcuate hyperechoic endoluminal formation with a posterior shadow cone.[1]

CT scan contributes the most to a diagnosis of small-bowel obstruction. It allows a positive, topographic and aetiological diagnosis and assesses the degree of intestinal ischaemia. It also contributes to therapeutic decision-making and the choice of surgical approach.[4] It shows in the case of bezoar a round or ovoid formation of heterogeneous density containing gas, not enhanced after injection of iodine contrast, filling the digestive lumen and a calibre disparity just downstream. Bezoars have a higher incidence of lesion shell, similar image in the stomach and fat-density floating fibers (FFF) sign that correspond to the visualisation of fatty density fibres within the intestinal lumen.[6,8]

The purpose of treatment is to empty the intestinal lumen, treat the complications and avoid its recurrence.[4] The gesture consists of fragmenting and pushing it into the cecum by antegrade washout. An enterotomy is indicated only if the bezoar cannot be fragmented and mobilised. Bowel resection is rarely indicated and should be reserved for cases of intestinal necrosis or if the bezoar is intimately encrusted within the intestinal wall.[4-6]

Extraction with a fibrescope is most often impossible due to the size of the bezoars, which prevent the crossing of the cardia. However, even in the case of small bezoars, it must remain exceptional because of the risk of oesophageal lesions linked to the passage of the fibrescope. It can be proposed in the case of contraindication to surgery after enzymatic dissolution.[7] In the event of associated psychopathological disorders: obsessive behaviour, compulsion, hyperkinetic syndrome, depressive syndrome, mental retardation and behavioural disorders, the child must be seen by a child psychiatrist. The approach should be based primarily on behaviour therapy.[3]

Conclusion

Rapunzel syndrome is an exceptional complication of trichobezoar. Typical CT imaging features play a diagnostic and prognostic role. Exploratory laparotomy is the only valid treatment. This syndrome is usually reported in patients affected by psychologic disorder that requires strong cooperation between the physician, patient and parents to prevent recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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