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Asperger syndrome in a patient with intestinal obstruction secondary to ileal trichobezoar - Case report

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ABSTRACT

The bezoar is a conglomerate of indigestible or partially digestible foreign material that stagnates in the digestive tract. Cases of primary intestinal trichobezoar, without associated gastric trichobezoar, are rare. Trichobezoar occurs most frequently in patients with mental disorders

We present the case of a patient aged 12 years and 5 months, who was referred to our hospital with the suspicion of intestinal obstruction, the clinical symptomatology consisting of uncontrollable vomiting, cramping abdominal pain and the absence of intestinal transit. Imaging investigations were inconclusive as to the cause of the bowel obstruction. Surgery was performed for mechanical intestinal obstruction and during the exploratory laparotomy an intraluminal formation was revealed in the ileum which after enterotomy was identified as a trichobezoar and other formations in the colon which were manually manipulated towards the distal colon and later evacuated manually by digital rectal palpation. Postoperative pediatric psychiatric evaluation revealed symptoms suggestive of Asperger Syndrome.

Our paper, explores the association between tricophagy and pervasive developmental disorders. Although many cases of autistic children ingesting inedible items are described in the literature, there are very rare situations in which pica causes obstruction of the small intestine, requiring surgical intervention.

Keywords: trichobezoar, intestinal obstruction, tricophagy, Asperger syndrome

INTRODUCTION

Bezoars are collections of partially or completely indigestible foreign material, retained in the gastrointestinal tract, most commonly in the stomach [1-4]. Several types of bezoars have been described depending on their composition: phytobezoars (plant material), trichobezoars (hair), lactobezoars (milk protein), lithobezoars (stones), plasticobezoars (plastic material), pharmacobezoars (tablet) [2,3,5,6]. Trichobezoars, (compact clumps of hair and other hair-like fibers chewed and swallowed) [2,5] account for less than 6% of all cases of bezoars found in humans [2,3]. They were first described in humans in 1779 by Baudamant who reported an autopsy in which he found a gastric trichobezoar leading to death by gastric perforation and peritonitis in a 16-year-old boy [1].

Up to 90% of cases have been reported in girls between 13 and 19 years of age with trichotillomania (compulsive pulling of one's own hair) and trichophagia (ingestion of hair that may be their own or other family members' or even doll hair) [2,6,7]. They are extremely rare in young children [2,8]. Trichobezoars develop and usually stay in the stomach, although they can occasionally go into the small intestine. For example, in Rapunzel syndrome, the gastric trichobezoar extends like a tail through the pylorus and into the small intestine, into the colon [5,6,9,10] or as an isolated or satellite bezoar in the small intestine [2,6,11]. The disease is manifested by nonspecific symptoms: abdominal pain, nausea, vomiting, constipation, weight loss, the presence of a palpable abdominal mass, anemia, malnutrition [3,8,9]. Sometimes it manifests itself through important complications such as digestive hemorrhage, intestinal occlusion, perforation and peritonitis [2-4,8,9,11].

Eating disorders, selectivity and poor oral intake are up to five times more likely to be encountered in Autism Spectrum Disorder (ASD) pediatric population than in neurodevelopmentally typical children [12]. In autistic children, food intake is very often conditioned by food category, colors or texture aversion, usually being preferred carbohydrates and processed edibles. 70% of children with ASD have at least one comorbid psychiatric disorder [13]. Children who associate autism spectrum symptoms with anxiety have an increased risk of feeding disorders, because they have an increased reactivity to stress, and multiple studies have correlated those comorbidities with higher post-stress cortisol levels [14]. Through their enhanced stress response, these children resort to aberrant eating behaviors.

Asperger syndrome (AS) is a neurodevelopmental disorder included in the autism spectrum in the DSM-IV (Diagnostic and Statistical Manual of Mental Disorders), characterized by difficulties in social interaction, restricted interests and repetitive behaviors. Unlike other autism spectrum disorders, AS does not typically affect intellectual development or language, although individuals may use a formal or literal speaking style. Since 2013, DSM-5 has brought AS together under Autism Spectrum Disorder (ASD) to reflect a continuum and support uniform diagnosis and treatment [15].

The comorbidity between Asperger's Syndrome and trichotillomania (the compulsive need to pull out hair) and trichophagia (hair ingestion) is complex, being associated with both neurobiological and genetic factors, but also with distinct behavioral manifestations. People with Asperger's syndrome frequently present a repetitive behavioral pattern, such as trichotillomania, which may be due to deficits in the cortico-striato-thalamo-cortical neurobiological circuits. These circuits play an important role in the control of impulses and the formation of habits, thus contributing to the appearance of repetitive behaviors found in both Autism Spectrum Disorder (ASD) and trichotillomania [16].

In addition to neurobiological factors, sensory processing problems often found in autism spectrum disorder can exacerbate trichotillomania-like behaviors because people with ASD may use them as self-soothing mechanisms in response to sensory overload. In addition, stress and anxiety, common in ASD, can intensify the urge to pull hair, creating a cycle of repetitive behaviors that are difficult to control [17].

CASE PRESENTATION

We will present the case of a 12-year-old patient who presented to "Grigore Alexandrescu" Emergency Clinical Hospital for Children, Bucharest, with symptoms suggestive of intestinal obstruction for surgical evaluation. The onset of the symptomatology was 6 days before the presentation with incoercible vomiting and cramping abdominal pain that were partially remitted during the hospitalization in another health facility but reappeared 3 days before, being accompanied by the absence of intestinal transit for 72 hours. The history didn't reveal certain known psychiatric conditions or manifestations suggestive for trichophagia.

At admission, patient's general condition was mediocre, subfebrile (37.7*C), she presented moderately distended abdomen, diffusely sensitive to palpation, no

palpable masses, no signs of peritoneal irritation. Digital anorectal examination reveals an empty rectal ampulla except for a small 3/2 cm conglomerate of fecal matter that has been digitally removed. About 1.3 l of food content is evacuated on the nasogastric suction tube.

Paraclinically, microcytic hypochromic anemia, elevated C-reactive protein (17.84 mg%) , hypochloremia (85 mmol/l), hypocalcemia (8.4 mg/dl), hyposideremia (24 microg/dl) and a lipase of 64U/l are detected, the rest of the blood tests being within normal limits. Abdominal X-ray showed no pathological elements. Abdominal ultrasound visualizes a solid mass with a hyperechoic wall and intense posterior attenuation located in the left paramedian supravesical area, with a diameter of 75 mm (fecaloma? adnexal mass?), distended hypoperistaltic intestinal loops, containing fluid, intraperitoneal fluid (50 mm) visible in the lower abdomen (supravesical).

Contrast enhanced computed tomography revealed distended jejunum and ileum, with loop diameter up to 35-40 mm, with fluid content and iodophilic thin walls, hypodense, inhomogeneous ovoid solid formations with hyperdense lyserium, located intraluminally in the distal ileum (70X40 mm), in relation to a hypodense intraluminal structure with dimensions of 25/10 mm (Meckel?) and in the ascending colon (80X35 mm) at the level of the hepatic flexure, downstream of which the transverse, descending and sigmoid colon are without content and liquid intraperitoneally in large quantity (65 mm in the lower abdomen).

An exploratory laparotomy was performed under general anesthesia and a firm intraluminal formation was revealed at the level of the proximal ileum, which could not be mobilized distally, upstream of which the jejunoileal loops were much dilated. A longitudinal enterotomy of about 4 cm was performed on the antimesosthenic edge of the supporting ileal loop, with the extraction of a foreign body of about 10/9 cm (macroscopically consisting of hair and food residues) and enteroraphy in two layers. (Figure 1). The control of the other viscera reveals intraluminal formations at the level of the ascending and transverse colon that were manually mobilized towards the sigmoid colon and rectum and later partially evacuated digitally by rectal palpation, having the same macroscopic appearance. No trichobezoars were seen in the stomach or small intestine proximal or distal to that previously described. Postoperative evolution was favorable, without complications.

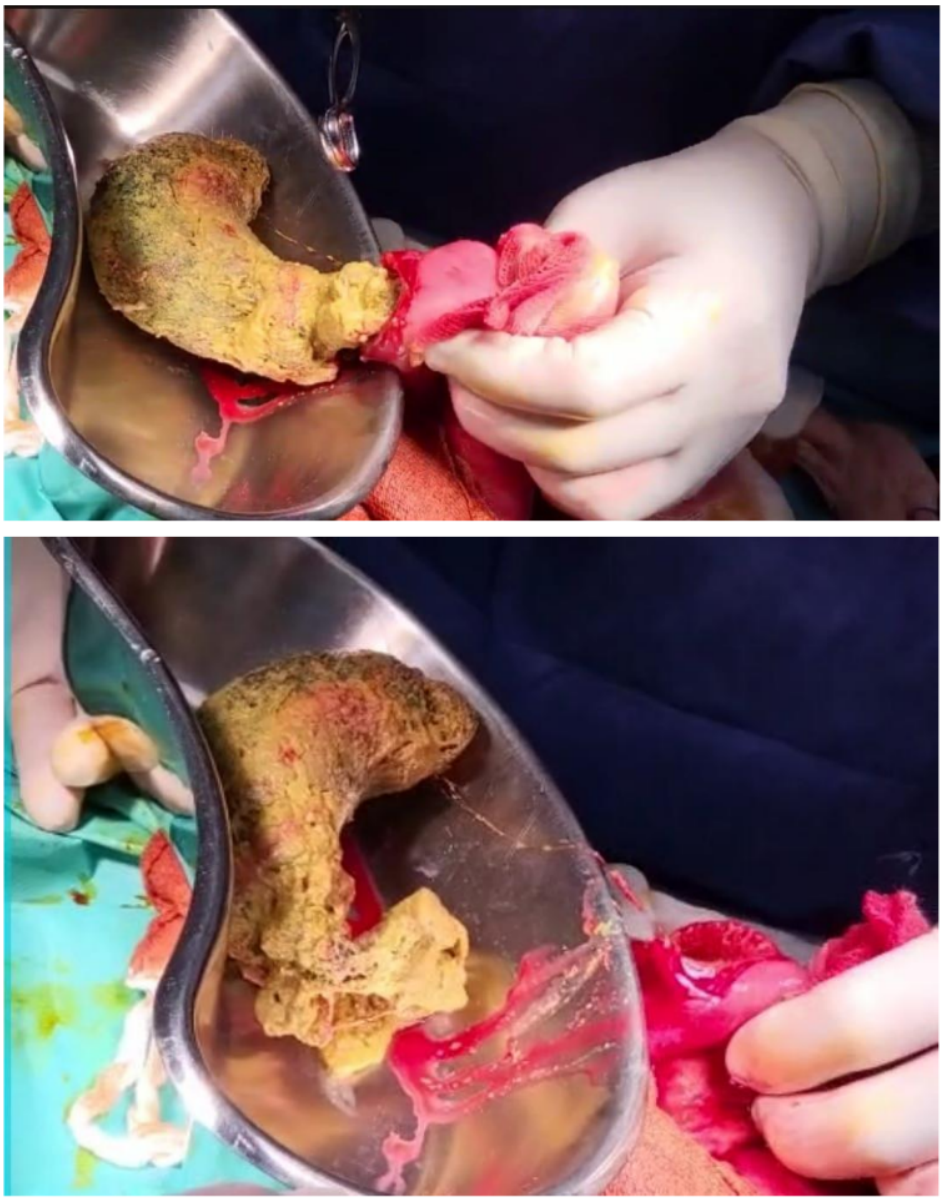

Figure 1. Intraoperative macroscopic aspect of the trichobezoar

Postoperative histopathological examination confirmed the diagnosis of trichobezoar.

The pediatric psychiatric evaluation performed the day after surgery revealed chronic PICA-type behaviors from a young age, lack of eye contact, chronic difficulties in understanding social norms, social isolation, motor stereotypes, restricted interests,

cognitive rigidity, abstract thinking. The parents confirmed the presence of aberrant eating behaviors that started from the age of integration into the school community (ingestion of nonnutritive items like erasers, pencils, hair ties elastics). Hair ingestion occurred around age 11 amid concerns about school performance/adjustment. After starting the fifth grade, the patient confirms the awareness of social inadequacy and multiple anxious states related to the maladjustment in the group of classmates and the inability to make friends.

At the age of 4, the patient was evaluated in a child and adolescent psychiatry department, suspicions were raised regarding the presence of a pervasive developmental disorder, but the evaluation procedures were not completed. The patient did not benefit from psychotherapeutic intervention for socialization difficulties, basal anxiety or pica-type behaviors prior to this hospitalization. Considering the urgent need to control hair ingestion behaviors to prevent postoperative complications during hospitalization, treatment with risperidone was initiated in progressively increased doses up to 1mg per day. When treating trichotillomania, antipsychotic medication have been shown to be beneficial either alone or in combination with selective serotonin reuptake inhibitors (SSRIs) for their augmentive potential [18].

During the hospitalization, the patient's tolerance to the psychotropic medication was very good, she had no adverse reactions and the trichophagia behaviors registered a complete remission. The patient was discharged on the ninth postoperative day, with good general condition, painless supple abdomen, good digestive tolerance, intestinal transit present, the surgical wound was in the process of scarring.

After 3 weeks from the surgical intervention, the patient presents for psychiatric reassessment, accompanied by her parents. Clinical psychiatric evaluation of the patient confirms the grammatically correct language but doubled by a particular intonation and bizarre prosody. During the interview the patient cannot maintain visual contact, and presents multiple stereotyped movements (such as walking on tiptoes) and mannerisms. The interview with the parents confirm the chronic inability to understand social rules or cues, the tendency to isolate herself, the lack of friendships, special interests (drawing, comic books characters), great inability of affective reasoning, of expressing emotional reciprocity and empathic communication.

The psychiatric evaluation was doubled by a psychological assessment using ADOS (Autism Diagnostic Observation Schedule) which revealed suggestive scores for an Autism Spectrum Disorder. The patient received recommendations to continue the

initiated treatment in the same doses and to be integrated into a cognitive behavioral and group therapy program and to be monitored monthly in the territorial pediatric psychiatry service.

DISCUSSION

Approximately 6% of all bezoars are intestinal trichobezoars, which are extremely uncommon [2,3]. Trichobezoars occur mainly (90% of cases) in young girls (13-19 years) who have mental health disorders and compulsive behaviors of chewing and ingesting hair [2,6,7,19]. About 1% of people with trichotillomania will develop trichobezoar that needs to be surgically removed, and only 30% of patients will also have trichophagia [2]. Sometimes parents hesitate to give information about behavioral disorders suggestive of trichotillomania and/or trichophagia in their children [4,20]. These behavioral problems may be due to psychiatric conditions, intellectual disability or factors such as hormonal changes, school pressure, child neglect or any event with an emotional impact [4,6]. In the presented case, the information regarding trichophagia was obtained only postoperatively, after the cause of intestinal obstruction was presented to the parents and no signs of trichotillomania (affected areas on the scalp) were revealed.

The size and position of the trichobezoar determine the range of clinical manifestations that can occur, ranging from non-specific signs and symptoms (abdominal discomfort, nausea, vomiting, early satiety) to more particular ones [3,8,9,21,22] and complications such as digestive hemorrhage, gastric erosions, gastric or intestinal perforation with peritonitis [3,11], intestinal obstruction [2-4] or even intussusception, obstructive jaundice and pancreatitis [10,21,22]. Trichobezoars typically reside in the stomach, however they can infrequently enter the small intestine as satellite or isolated trichobezoars and result in secondary intestinal blockage [2,6,11]. A unusual form of trichobezoar known as Rapunzel syndrome occurs when the stomach trichobezoar extends like a tail past the pylorus and into the small intestine or even the colon [5,6,9,10,20]. In the presented case, the symptoms and investigations established the diagnosis of mechanical intestinal obstruction and the etiological diagnosis was established intraoperatively, being an obstructive ileal trichobezoar, possibly primary (as a gastric trichobezoar was not identified in this case to suspect the detachment and

migration of a fragment of it in the small intestine) or migrated entirely from the stomach.

Trichobezoars can be diagnosed, depending on the location, by upper digestive endoscopy, the gold standard in the case of gastric bezoars [4,5], respectively contrast enhanced computed tomography in the case of intestinal trichobezoars, even if the changes are non-specific. Various authors have suggested that computed tomography is superior to other radiological investigations for the diagnosis of bezoars in cases of intestinal obstruction [3]. Demonstration of a well-defined intraluminal, oval, hypodense, inhomogeneous, air-bubbled mass with proximally dilated small bowel and distally collapsed loops is highly suggestive of a bezoar [3,6,23,24]. In the presented case, the computed tomography revealed intestinal occlusion but was not highly suggestive of its etiology.

The treatment in the case of intestinal trichobezoars with obstruction consists in their surgical removal, by laparotomy or laparoscopy. For small and soft bezoars, it is possible to attempt to manipulate them distally to the large intestine, and if the bezoar is large, hard and fixed in the intestinal lumen, enterotomy is performed with the extraction of the bezoar or, in case of complications (stricture of the loop, perforation), segmental resection of the loop [19,25]. It is important to ensure that there are no remaining trichobezoars in the stomach or small intestine. The laparotomy surgical approch is considered the gold standard treatment because of its high success an low complication rates, and because of the possibility to examine the whole digestive tract [2,6]. In the presented case, the preoperative diagnosis being mechanical intestinal obstruction without a clear etiology, it was decided to perform exploratory laparotomy and later to extract the intestinal trichobezoar through enterotomy followed by enterorrhaphy and the distal mobilization of the trichobezoar from the ascending colon.

Important in the management of trichobezoars is the prevention of relapses, through the evaluation and treatment of possible psychiatric conditions and behavioral disorders [4]. We intent to highlight the importance of evaluating a possible neurodevelopmental disorder in children and adolescents who display pica or trichotillomania behaviors. Cohort studies have reported high prevalence rates (12-13/%) of pica comorbid with ASD in children and adolescent subjects [26,27]. Investigating atypical developmental profiles (language development, presence of stereotypes, chronic food restriction habits and social interaction deficits) could help to understand vulnerability factors and the biologycal connection between

neurodevelpmental disorders and compulsive behaviors like pica, trichotillomania or tricophagia.

CONCLUSION

The preoperative diagnosis of intestinal occlusion by trichobezoar can be difficult, sometimes even the results of abdominal ultrasound and tomography not being conclusive. History suggestive of trichotillomania, trichophagia, or already diagnosed psychiatric conditions would be helpful in raising the suspicion of trichobezoar. Treatment in such cases of intestinal obstruction without specified etiology is surgical to identify and remove the cause of the mechanical obstruction and is followed by specialized psychiatric treatment to prevent relapses.

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