CASE REPORT

Trichobezoar in a patient with sickle cell anemia: A pain crisis is not always to blame

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Abstract  Trichobezoar, a ball of hair in the stomach, is an under-diagnosed problem that should be considered in patients with an upper abdominal pain and mass, weight loss, and bowel obstruction especially in young girls with underlying psychiatric disorder or mental retardation. Patients with sickle cell disease are susceptible to experiencing pica including, in rare occasions, trichophagia that could lead to formation of a trichobezoar, resulting in serious abdominal complications which are likely to be confused with ordinary vaso-occlusive pain crises. Here we present the rare case of gastric trichobezoar in a 9 year old girl with sickle cell anemia.

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1. Introduction

A bezoar is an accumulation of exogenous matter in the stomach or intestine (1). The word “bezoar” comes from the Arabic word “bedzehr” or the Persian word “padzhar”, meaning “protecting against a poison”. Historically, bezoars from animal guts were used as precious stones, antidotes to poisons, and today as part of traditional Chinese medicine (2). Bezoars are classified according to their composition into trichobezoars (hair), phytobezoars (vegetable), and lacto bezoars (milk/curd), and miscellaneous (fungus, sand, paper, etc.) (3). The most frequent type of bezoar in adults is phytobezoar, while trichobezoars are more often found in children and teenage girls (4). Trichobezoar is the formation of a hair ball in the stomach after compulsive eating of hair (trichophagia) (5). It is a rare condition that is reported most often in females, which may be attributed to their traditionally long hair (2). Trichophagia is considered a subtype of pica. The latter is a more...
general term defined as “the persistent eating of non-nutritive substances” (6).

The term “trichotillomania” was coined late in the 19th century to describe the recurrent pulling out of one’s own hair (7) and (8). In trichotillomania, there is an irresistible desire to pull out the hair from the scalp, eyelashes, eyebrows and other parts of the body, resulting in an instant release of tension, a sense of relief and security (3). It is postulated that the swallowed hair strands, which are too resistant to be propelled by peristalsis, are initially retained in altered mucosal valves of the stomach and, over time, impaction of hairs together with mucus and food occurs, leading to the formation of a trichobezoar (9). Trichobezoars are usually black from denaturation of protein by acid, glistening from retained mucus, and foul smelling from degradation of food residue trapped within it. The more severe manifestations are complete gastric outlet obstruction. (10).

Large trichobezoars may extend through the duodenum, causing small bowel obstruction, a condition known as “Rapunzel syndrome” that was first described in 1968 by Vaughan et al. (11). In that syndrome, gastric trichobezoars could have a long tail that can extend to ileocecal valve (12). Detached fragments of the bezoar may be detected as “satellite masses” within the small bowel and could lead to small bowel obstruction (10).

Common presenting symptoms of trichobezoars are abdominal pain, nausea, vomiting, weight loss, malnutrition, hematemesis, diarrhea, constipation, gastric ulcers, perforation, pancreatitis, obstructive jaundice, and malabsorption. On physical examination, epigastric mass may be palpated. Alopecia may also be present due to trichotillomania (13) and (10). Constant irritation of the gastrointestinal mucosa by the stomach cast and the tail has been suggested as the cause of the gastric or intestinal ulceration and protein losing enteropathy (14) and (15). Trichobezoars are usually associated with underlying psychiatric disorders, such as depression, obsessive–compulsive disorder, body dysmorphic disorder and, particularly, trichotillomania. However, their prevalence and co-morbidity is unclear (8).

Children with sickle cell disease frequently present to the emergency room with abdominal pain. The most frequent cause is a vaso-occlusive pain crisis. However, other etiologies exist and can be a source of great morbidity and mortality. One of the unusual causes of abdominal pain in these children is trichobezoar which is a consequence of pica. In general, pica appears to have a high prevalence among children with sickle cell disease reaching about 34% in one series (16).

2. Case report

A nine year old female patient with sickle cell anemia presented to our emergency room with severe acute abdominal pain, vomiting, fever, anorexia, and irritability. This acute condition was the culmination of a relatively milder illness that started about 2 weeks earlier, involving intermittent low grade fever, anorexia, moderate upper abdominal pain, vomiting, and foul smelling breath. During this earlier stage, the patient underwent some laboratory and radiological investigations, including abdominal ultrasonography, which was all non-revealing, apart from mild splenomegaly. Thus she was presumed to have an ordinary vaso-occlusive pain crisis and was treated accordingly and reassurance was given to the parents but the condition deteriorated. The parents were especially scared at the idea that their ever-wasted daughter lost further 2.7 kg from her weight over the last 4 months.

The past medical history of the patient was, for the most part, not unusual for a patient with sickle cell disease. She was diagnosed, at the age of 6 months, as having sickle cell anemia (HbS disease: HbS 82% and HbF 18%) and had multiple episodes of pain crises in the back, limbs, and chest but never in the abdomen. She had a history of blood transfusion twice and was receiving oral penicillin prophylaxis, folic acid, and other vitamins. She had also received the vaccines against capsulated bacteria. Six month ago, the patient had an episode of encephalopathy for several days and a definite diagnosis was not given to the parents. The patient also had a family history for sickle cell disease including the mother and an uncle.

What seemed unusual was to know that the girl started to show a bizarre behavior from the age of five years when she was found to like eating hair, either her own or from the others’ combs. The girl also used to eat other objects like small stones but to a much less extent than eating hair. She refused all kind of advice and continued to eat hair in secret. She was described by the parents as introvert, depressed, selfish, and with intelligence level less than her siblings but without school failures. She also exhibited features suggestive of obsessive–compulsive disorder like excessive hand washing. On psychiatric consultation, the parents were reassured that the hair eating is a behavior completely expected from an anemic child. Only multivitamins and minerals were prescribed but no psychotropic medications were given.

On clinical examination, the patient was cachectic, irritable, restless, lying still, pale, feverish (390c), tachypnic, and with the characteristic mongoloid features of patients with chronic hemolytic anemia. No obvious hair loss was noted. The girl’s weight, stature, and head circumference were all below the 3rd percentile for age (14.5 kg, 121 cm, 47 cm respectively). Abdominal examination revealed distension, rigidity, markedly tender left hypochondrial hard mass, and exaggerated (later decreased) intestinal sounds. Otherwise, the patient appeared normal.

Laboratory tests showed an Hb level of 10 g/dl, marked leukocytosis (34,000/ml) with absolute neutrophilia, normal liver and kidney function tests, normal serum ferritin, and normal serology for hepatitis B and C as well as for HIV. Blood and urine cultures were withdrawn and empirical antibiotics were given. A surgical emergency was strongly suspected and plain X ray chest and abdomen (erect) was ordered which showed diffuse dilatation of the stomach and proximal duodenum that were filled with mottled radiopaque contents. There was also left sided air under diaphragm (pneumoperitoneum).

Abdominal ultrasound showed an irregular mass with hyperechoic arc-like surface seen in epigastric region with marked posterior acoustic shadowing and marked hardness under probe, mild amount of fluid collection at the lesser sac and free intra peritoneal, in addition to mild splenomegaly. Abdominal CT without contrast was revealed marked dilatation of the stomach with mottled hyperdense structures within its lumen extending to the first part of the duodenum, mostly of collection or concretion of indigestible materials that accumulate and coalesce in the stomach (Fig. 1). By correlating the radiological findings with the patient’s history of trichophagia,
a gastric trichobezoar with early gastric perforation was suggested.

Exploratory laparotomy was performed through midline incision. A minimal purulent exudate was found at the lesser curvature and Morrison pouch with one cm perforation at the anterior gastric wall close to the lesser curve. The lesser sac was opened to fully explore the stomach. The stomach was massively distended by a hard mass with intact serosa. Anterior gastrotomy was done with the extraction of a hard, bile-stained mass 18 × 9 cm, 1.3 kg, formed of amalgamated hair and extending into the duodenum (Fig. 2). The gastric mucosa was found intact, apart from the perforation. Closure of gastrotomy and the perforation, using omental patch, was done. Cholecystectomy was done for a gall bladder stone felt at the operation. The postoperative course was uneventful and the patient was discharged on the 4th day. Blood transfusion was given after surgery to elevate Hb level to 10 g/dl.

3. Discussion

Patients with sickle cell anemia typically present with recurrent vaso-occlusive pain crises. When a patient with SCD presents with abdominal pain, physicians are inclined to attribute it to such vaso-occlusive events. However, this is not always the case and serious underlying surgical or medical problems may be missed such as vertebral marrow hyperplasia or infarction with compression of nerve roots, mesenteric and retroperitoneal lymphadenitis with infarction, and occlusion of the blood supply to abdominal organs with infarction of the spleen, liver, and less commonly the intestine. Other causes of abdominal pain in patients with sickle cell anemia include viral hepatitis, acute cholecystitis, cholelithiasis, choledochocholitis, pancreatitis, and peptic ulcer disease (17) and (18).

Our patient is a typical example of a patient with SCD who had a surgical abdomen that was erroneously diagnosed as an ordinary pain crisis. In fact, some investigators tried to differentiate between both conditions. It was found that pain in vaso-occlusive crises tends to be diffuse and unrelated to meals or bowel movements. It also tends to be associated with remote pain such as in the limbs and chest. In addition, a precipitating event is frequently found in vaso-occlusive crises such as upper respiratory infections, dehydration, acidosis, cold exposure, or physical exertion. Likewise, the pain of a vaso-occlusive crisis usually simulates that of prior crises and deviation from a characteristic pattern should raise suspicion of an alternative diagnosis. Also, in vaso-occlusive crises, the

![Radiological findings in our patient: (A) CT scan showing the gastric trichobezoar (right and left arrows) with a mottled hyperdense appearance, a hypodense area (middle arrow) denoting mild amount of fluid collection at the lesser sac from gastric perforation. (B) Plain scout CT of the abdomen showing dilated distended stomach with hyperdense shadow within its lumen (lower arrow) in addition to air under diaphragm on the left side (upper arrow).](image-url)
abdominal pain is often out of proportion to the physical findings found on abdominal examination. It is particularly important to note that pain localized solely to the abdomen, especially with tenderness to palpation or peritoneal signs, without joint or bone pain, is rarely due to sickle cell crisis. The presence and persistence of bowel sounds are not consistent with diffuse peritonitis and usually support the diagnosis of a crisis. Finally, vaso-occlusive pain crises often resolve rapidly within 48 h of medical treatment, in contrast to the pain in surgical conditions. It must be stressed that though extremely useful, these general rules have exceptions. For example, a precipitating event may not be found in vaso-occlusive crises.

Also, frank ileus or even silent abdomen may accompany a pain crisis (19) and (20).

Regarding our patient, what made an ordinary vaso-occlusive crisis unlikely was the presence of high grade fever, paucity of body motions (lying still), toxic appearance, irritability, restlessness, persistent vomiting, presence of a markedly tender abdominal mass, abdominal distension, rigidity, decreased intestinal sounds, and markedly elevated leukocytic count. In addition, abdominal pain had not been the target of previous vaso-occlusive episodes which were usually experienced in the limbs, back, and chest. Moreover, the course of pain was much more prolonged than expected from an ordinary pain crisis.
Trichobezoar in a patient with sickle cell anemia

3.1. Now, we must ask: what is the etiology of the problem?

Pica is the persistent eating of non-nutritive substances which is inappropriate to the developmental level (the normal mouthing of objects in infants and toddlers) and not part of a culturally sanctioned practice (30). Several types fall into this spectrum, including geophagia (eating dirt), pagophagia (eating ice), and trichophagia (eating hair). The prevalence of pica among patients with sickle cell anemia homozygous for HbS is about 35–54% and significantly lower in compound heterozygous patients (16) and (31). Though pica, in general, is highly prevalent among children with SCD, trichobezoars are very rarely reported. As far as we know, only two cases have been previously reported. Stein-Wexler et al. reported the unexpected occurrence, in a boy, of a gastric trichobezoar extending into duodenum and causing pancreatitis. On the other hand, Sciarretta and Bond reported a gastric trichobezoar in a young, 3 year old girl suspected to have sickle cell splenic sequestration. Our patient represents a further example in this regard.

The etiology of pica in SCD is not definitely known but it has been linked to developmental delay and brain damage resulting from silent infarction which occurs in up to one forth of children with SCD, often in the frontal lobes and associated with cognitive deficits and therefore potentially with unusual behavior. However, this association has not been confirmed (31). Our patient did not have any history of stroke. Notwithstanding, about 6 months ago she had an episode of encephalopathy and convulsions for several days during which neuroimaging proved normal. Likewise, trichophagia had been existent long before this episode of encephalopathy and continued to occur in the same way after it.

Iron deficiency had classically been blamed as an underlying etiology of pica (6). Although iron deficiency has been described in SCD (32), it is uncommon due to repeated blood transfusion and increased iron absorption. The RBC indices of our patient were not microcytic or hypochromic and thus not suggestive of iron deficiency anemia. Also, serum ferritin measured after recovery was normal, excluding iron deficiency as a cause of pica in this girl. Other mineral deficiencies were also implicated in pica such as calcium and zinc. It was also found that renal tubular reabsorption of zinc is decreased in patients with sickle cell anemia, leading to hyperzincuria (33). Our patient had normal serum calcium but serum zinc level was not measured and the girl was already on minerals and multivitamin supplementation for several months before presentation.

Trichobezoar are usually associated with underlying psychiatric disorders such as depression and obsessive compulsive disorder. Meanwhile, children with SCD do appear to be at greater risk for developing depression than those with other comparable disorders such as cancer, cystic fibrosis, and diabetes (34). Consequently, psychiatric disorders like depression could be a link associating SCD with trichobezoar. Our patient also showed symptoms of depression which could account for the trichophagia and trichobezoar (8).
4. Conclusion

It can be concluded that children with sickle cell anemia are liable to develop psychiatric symptoms including trichotillomania, leading to trichophagia and trichobezoar that can produce abdominal pain which is easily confused with a usual pain crisis. It is thus imperative to search for surgical causes of pain in patients presenting with a non-classic clinical picture.

Conflict of interest

None.

References