A rare outcome of iron deficiency and pica: Rapunzel syndrome in a 5-year-old child

Ali İslek1, Ersin Sayar1, Aygen Yılmaz1, Cem Boneva2, Reha Artan1

1Department of Pediatric Gastroenterology, Akdeniz University Faculty of Medicine, Antalya, Turkey
2Department of Pediatric Surgery, Akdeniz University Faculty of Medicine, Antalya, Turkey

ABSTRACT

Bezoar is defined as the accumulation of organic or nonbiological substances inside the gastrointestinal system. Trichobezoars are the most frequently detected ones and are mostly present in patients with neuropsychiatric disorders. The continuance of the trichobezoar tail-shaped extension over the duodenum and jejunum is described in Rapunzel syndrome. Both conditions are rarely reported in children. The present case submitted here is related to a 5-year-old girl referred with an abdominal mass and anemia, diagnosed with Rapunzel syndrome and developing trichobezoar due to iron deficiency and pica.

Keywords: Rapunzel syndrome, iron deficiency, pica

INTRODUCTION

Bezoar is the accumulation and mass formation of indigestible materials inside the gastrointestinal tract on account of several factors. Bezoars caused by hair particles, vegetable fibers, and semi-fluid medications or pills are called trichobezoar, fitobezoar, and pharmacobezoar, respectively (1). Trichobezoar has been widely reported in young women with psychiatric and neurological disorders (2). The clinical condition in which a bezoar trapped in the stomach extends over the duodenum and jejunum has been likened to long-haired Rapunzel in the Grimm Brothers and is thus termed as Rapunzel syndrome (3). In the present paper, a 5-year-old case with no psychiatric or neurological disorder but developing Rapunzel syndrome as an outcome of pica disorder is reported.

CASE PRESENTATION

A 5-year-old girl with vomiting complaints was referred upon detecting a mass formation in her abdomen to our hospital for further examination and treatment from the first center to which she applied. It has been reported that the patient has vomited a total of 3 times within two days and had no abdominal pain. Her height is 104 cm (25 p) with a body weight of 15 kilograms (3-10 p). Physical examination revealed noticeable pallor and a solid mass in the left hypochondriac area, and the epigastrium has been palpated. Laboratory results gave the following figures: hemoglobin: 4.5 g/dL, white blood cell: 11,700/mm3, platelet: 670,000/µL, MCV: 58 fl, RDW: 27%, ferritin: 2.3 ng/mL (normal range: 30-400 ng/mL), and iron: 12 µ/dL (normal range: 33-193 µ/dL). In the peripheral blood smear, anisocytosis, polychromasia, and poikilocytosis have been reported. Abdominal X-ray presented a dilated stomach and a mass formation with multiple small air bubbles inside (Figure 1). Despite the denials of the parents claiming that their child took no non-food materials, it was still agreed that an upper endoscopic examination be made, since the abdominal X-ray findings referred to bezoar. This examination has shown a trichobezoar filling almost all of the stomach and extending to the jejunum (Figure 2). The bezoar—too big to be removed with an endoscopic method—has been removed via surgical process (Figure 3). For all these initiatives and this notice, informed consent was obtained from the parents.
DISCUSSION

Bezoar is a rare condition and refers to masses that are composed of undigested food, fiber, or hair inside the gastrointestinal system (1). In a good number of cases reported, the primary cause is trichobezoar. Trichobezoars are formed by indigestible materials, such as hair, nylon, and wool. The incidence is greater in young female patients with psychiatric disorders and mental retardation (2).

It has been emphasized in relevant literature that the main cause underlying the formation of trichobezoars is trichotillomania, and these patients are, though not always, diagnosed with partial alopecia due to tearing out the hair (4). The present case, however, had no trichotillomania and no neuropsychiatric disorder. Endoscopic and surgical findings have shown that the patient was in the habit of eating hair, small hair, wool, and thread in clothes. Echoing the earlier findings reported in the literature, despite the great size of the removed material, the parents claimed that the practice of eating foreign material was too rare to notice in their child.

It was also found that paleness in the patient was noticed for a long time, but no examination or treatment was received. Iron deficiency anemia, which is widely common in patients with trichobezoar, is mostly a direct outcome of some gastric ulcers caused by the pressure effect of mass (5). Likewise, Larsson et al. (6) have reported that a patient with celiac disease who had trichophagia and trichobezoar in childhood that was an outcome of pica-originated celiac disease. In the examinations to detect reasons of the iron deficiency anemia, there was no hematuria, and occult blood positivity in stool, celiac autododies, and H. pylori antigen in the stool were negative. It has been concluded that iron deficiency of the patient was related to wrong feeding habits and was an outcome of nutritional deficiency that was bound to the growing mass effect and that the bezoar has developed as an outcome of pica, which comes secondary to iron deficiency.

Trichobezoars can be in small sizes or so large so as to fill up the whole stomach and extend to the duodenum, even as far as the jejunum. This case indicates the presence of Rapunzel syndrome (3). The most noticeable finding in the physical examination is the presence of a well-defined and sometimes moving mass generally trapped inside the epigastrium. If the history fails to provide sufficient findings revealing trichophagy, this mass is at times mistakenly likened to a tumor (7). In the present case as well, a big, solid mass has been palped inside the left hypochon-
The recommended treatment in huge trichobezoars and Rapunzel syndrome is removing the mass via surgical method (2). Upper endoscopy has been applied on our case, and bezoar has been shown. Yet, since the bezoar was too big to be removed via endoscopy, it has been surgically removed via gastrotomy.

To sum up, Rapunzel syndrome is quite rarely reported in children, and in most cases, there is no medical history that supports the presence of trichophagy. It should be kept in mind that there is the possibility of the existence of bezoar in children who have anemia and abdominal mass but no neuropsychiatric disorder.

Ethics Committee Approval: N/A.

Informed Consent: Written informed consent was taken from the patient’s parents who participated in this case.

Peer-review: Externally peer-reviewed.


Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES