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CASE REPORT

A Hairy Tail not a Fairy Tale – Rapunzel Syndrome

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Abstract Rapunzel syndrome is an unusual and rare type of trichobezoar. Bezoars can be classified according to the primary constituent, as trichobezoar (hair), phytobezoar (plant material) or miscellaneous (pharmacobezoar, lactobezoar, fungal agglomeration and foreign bodies). When a long tail of hair strands extends from the main mass in the stomach along the small intestine and beyond it is known as Rapunzel syndrome. Here we are reporting a case of Rapunzel syndrome with a very long tail who was managed successfully. And reviewing the literature on the pathophysiology and management of these patients. These patients commonly present with obstructive symptoms and needs a high index of suspicion especially, in young female patients who have alopecia circumscripta and underlying psychiatric disorders. Early diagnosis and treatment is required to prevent complications due to this condition. Currently surgical management of this condition is the treatment of choice.

Keywords Bezoars/complications · Bezoars/diagnosis · Bezoars/psychology · Bezoars/surgery · Female · Laparotomy

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Introduction

For centuries, it has been known that bezoars occur in the form of undigested material in the intestine of animals and humans. It can be classified according to the primary constituent, namely, trichobezoars (hair), phytobezoars (plant material) or miscellaneous (pharmacobezoars, lactobezoars, fungal agglomeration and foreign bodies). When a long tail of hair strands extends from the main mass in the stomach along the small intestine and beyond, it is known as Rapunzel syndrome, a rare presentation of a trichobezoar.

Case Report

A 28-year-old female presented with the symptoms of vomiting and epigastric pain for three months. There was no history of haemetmesis or melaena. She reported a history of having undergone a caesarian section two years ago. On further questioning, she also admitted to have a history of habitually ingesting hair, plucked from her scalp. On examination, a 6×4 cm epigastric mass was found which moved well with respiration. An ultrasound examination of the abdomen showed a linear hyperechoic area with shadowing in the epigastric region. A barium meal revealed a markedly dilated stomach with a bezoar (Fig. 1).

Laparotomy showed a grossly distended stomach, with the serosa appearing normal. The bezoar was occupying the entire stomach, with a tail extending up to the distal jejunum. A transverse gastrotomy was performed and the bezoar was removed, including its very long tail (Fig. 2).

She made an uneventful recovery. She was referred to the psychiatry department for further treatment.

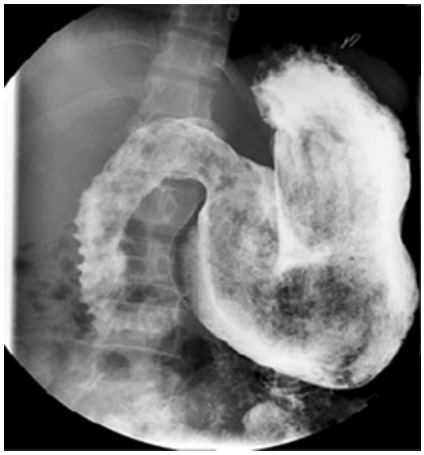

Fig. 1 Barium meal showing filling defect with mottling suggestive of Trichobezoar

Discussion

The term *bezoars* is derived from the Arabic word *badzehar* or Persian word *panzehr*, which means protection from poison. In 1968, Vaughan et al. who coined the term Rapunzel syndrome first described it [1]. There is no particular definition of Rapunzel syndrome; it is generally agreed that (a) the presence of a trichobezoar with a tail, (b) the extension of the tail to at least the jejunum and (c) the symptoms suggestive of obstruction would qualify to be termed as Rapunzel syndrome [1].

The most common presenting complaints are abdominal pain, vomiting and signs of gastrointestinal obstruction or perforation. Weight loss and malnutrition have also been described as its symptoms [2]. Rarely patients can also present with intussusception, bleeding, obstructive jaundice, pancreatitis or protein losing enteropathy [2, 3, 4]. If left untreated, it can lead to high mortality due to other complications (viz, perforation and bleeding).

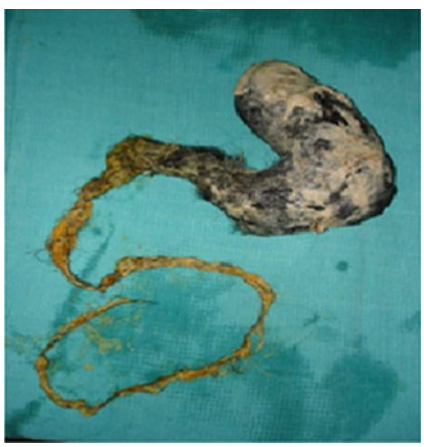

Fig. 2 Specimen of trichobezoar removed from the stomach with its long tail

Patients also exhibit symptoms of trichotillomania (an irresistible urge to pull one's hair) and trichophagia (morbid habit of eating hair). Only 30% of patients with trichotillomania will engage in trichophagia, and only 1% will ingest enough hair so as to warrant surgical removal [5]. Trichobezoars can rarely occur in patients without any psychiatric disorders.

On ultrasound examination, a curvilinear bright echogenic band with posterior shadowing is observed [6]. But this is not pathognomonic and can sometimes be misleading [2]. With the help of a barium study, the hair ball gets evenly coated with a small amount of contrast and becomes visible. It also helps detect extension, dislodgment (satellite masses) or synchronism. Trichobezoars in the stomach have a characteristic appearance, of a concentric inhomogeneous mass with entrapped air surrounded by contrast material, on CT scan. The presence of tail is reflected by small round hypodensities, though it sometimes can imitate traces of small bowel faeces seen in high-grade obstruction [7]. Contrast-enhanced CT or barium study would be the investigation of choice to confirm the diagnosis. Endoscopy may also be useful to confirm the diagnosis.

Surgical removal is the treatment of choice. Successful removal by laparoscopic surgery has also been reported [8]. Endoscopic methods have been reported successful, but mostly for small bezoars [9].

Recurrence of Rapunzel syndrome has been reported and long-term psychiatric follow-up is advised. Although it is a rare occurrence, awareness is required to diagnose and manage this condition.

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