

Rapunzel syndrome: a tail too long to tell!

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DESCRIPTION

Trichobezoar leading to Rapunzel syndrome (RS) is an extremely rare entity with about 90 cases reported in literature.¹ RS derives its name from the fairy tale 'Rapunzel', where the German princess let her long golden hair down from her tower to facilitate a tryst with her lover akin to the long and shiny tail of hair seen in RS.² Hair being slippery gets trapped in gastric mucosal folds, eluding peristalsis. More and more hair conglomerate to form a stomach-shaped mass coated with mucus called trichobezoar. This provides it a shiny glistening surface, and the acid secreted in stomach denatures the hair protein which gives it the typical black colour.³ When the tail of hair extends beyond the stomach into the small intestine, it is called RS.

A 6-year-old girl presented to our emergency department with history of abdominal pain and recurrent vomiting for 3–4 days. On examination she was dehydrated, had tachycardia and looked malnourished. Her abdomen was grossly distended without signs of peritonitis. An X-ray abdomen (erect) showed four-air-fluid levels suggestive of acute small bowel obstruction. With the provisional diagnosis of band obstruction, she was taken up for exploratory laparotomy after stabilisation.

On exploration, the stomach was grossly distended with a palpable semisolid mass inside it which seemed to continue into the duodenum, jejunum and up till the mid-ileum. Beyond this, the ileum and colon seemed normal in calibre. On gastrotomy, a giant trichobezoar was found in the stomach which was removed carefully (figure 1A,B). The long tail of the bezoar continued till mid-ileum and was removed

partly through the gastrotomy and partly through a separate enterotomy (figure 1C,D). The entire tail of hair was delivered without any free spill in the peritoneal cavity. Both the gastrotomy and enterotomy were repaired primarily in two layers. The hair ball weighed 2100 g, measured approximately 11 cm × 16 cm in size, and its tail extended about 110 cm in length distally into the jejunum and ileum (figure 2).

Retrospectively, parents gave history that their girl used to play with her hair and put them in her mouth since childhood, which she probably swallowed. However, they gave no history of any psychiatric illness. She had decreased appetite, early satiety and poor weight gain since childhood. On specific examination, her hair were sparse with the presence of flag sign, and she weighed below 50th percentile for her age. This history explained the sequence of events; trichotillomania followed by trichophagia, eventually leading to trichobezoar with its tail of hair extending beyond the stomach and causing acute intestinal obstruction. Hence, RS was diagnosed. The recovery was uneventful, and the child was discharged on day 7 after psychiatric evaluation. On follow-up after 1 month, she had no surgical complaints; however, behaviour therapy and counselling for trichotillomania, and a high protein diet for nutritional build-up were advised.

RS is commonly seen in young females like in our case; majority have some associated psychiatric disorders like trichotillomania. The most common symptoms and signs are abdominal pain (37%), nausea and vomiting (33.3%), obstruction (25.9%) and peritonitis (18.3%), and the less common ones are weight loss (7.4%), anorexia, haematemesis and intussusceptions (7.4%).² A few cases present with complications like obstructive jaundice, pancreatitis, gastric ulcer, appendicitis and gut perforation.¹

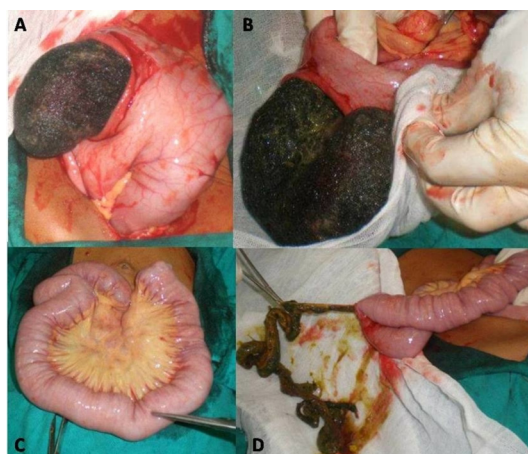


Figure 1 (A) Gastrotomy revealed large trichobezoar; (B) removal of trichobezoar through gastrotomy; (C) extended tail of trichobezoar causing distension of the small intestine up till mid ileum; (D) enterotomy and removal of the entire Rapunzel's tail of hair.



Figure 2 The entire specimen put together in toto; the long Rapunzel's tail of hair is seen clearly.



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Abdominal examination typically reveals an upper abdominal mass (Lamerton's sign). Radiological investigations like ultrasound, dye study and CT scan can clinch the diagnosis preoperatively. Endoscopy is both diagnostic and therapeutic. Laparoscopic removal is possible though not always feasible as the tail often extends into the jejunum. Hence, laparotomy is the preferred approach especially in emergent cases similar to ours.^{2,4}

A comprehensive and long-term psychiatric follow-up is needed in all cases as late relapse is possible. Apart from behaviour therapy, pharmacotherapy (clomipramine, quetiapine

or augmenting a selective serotonin reuptake inhibitor with an atypical antipsychotic effect) may be needed in recurrent or resistant cases.⁴

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Learning points

- ▶ Rapunzel syndrome is an extremely rare form of trichobezoar commonly seen in young females who are in a habit of eating hair.
- ▶ Acutely symptomatic, large trichobezoars, especially when accompanied by Rapunzel syndrome, must be removed surgically preferably by laparotomy or by laparoscopy.
- ▶ Comprehensive and long-term psychiatric follow-up is of utmost importance to prevent recurrence or late relapse of this condition.
- ▶ Patients presenting with signs or symptoms of intestinal obstruction should be examined thoroughly for psychiatric conditions and broken hair to rule out the rare possibility of trichotillomania.

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