

Case	(332) A hairy situation: rapunzel's syndrome
Authors	S.p. Mazzini Florindez, A.p. Corujo.
Centre	Hospital De La Sant Creu I Sant Pau.

CASE PRESENTATION

A 9 year old female patient brought by her parents to pediatric urgent care presenting with hyporexia, intermitent vomiting and abdominal pain for the past week. She had lost 4 kg in said week and her parents denied fever or any changes in stool or urine.

Physical examinaron found a distended and diffusely painful abdomen with no peritonism or visceromegaly. The abdominal standing x-ray showed distended small bowel loops with multiple airfluid levels, absence of gas in the colon and rectum and no pneumoeritoneum.

With the clinical suspicion of small bowel obstruction an abdominal CT was performed which showed distended small bowel loops with abundant intraluminal fluid and airfluid levels, a collapsed colon and two low-density intraluminal ovoid masses containing air bubbles with a typical mottled appearance, one occupying the whole gastric lumen and the other one at the distal ileum.

This radiological findings suggested a small bowel obstruction secondary to a gastric and ileal bezoar. The patient underwent surgery where the presence of a double trichobezoar was confirmed.

DISCUSSION

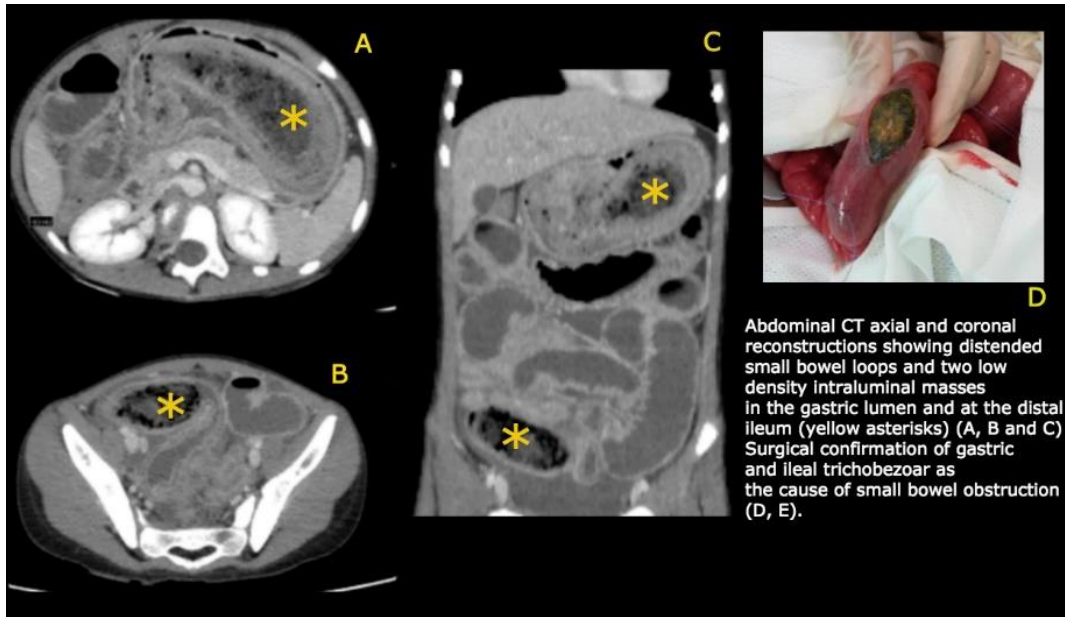
The presence of a dilated small bowel with a collapsed colon is highly suspicious of small bowel obstruction. The use of CT in the diagnosis of small bowel obstruction in children is a useful diagnostic procedure to confirm the obstruction, determine the cause and help pre-surgery planification. (1)

Trichobezoars consist of hairballs or hair-like fibers caused by chewing and swallowing hair or any other indigestible materials. They usually form in the gastric body and are found prepylorically. Some trichobezoars can fragment and pass through the pylorus into the small bowel causing a condition called Rapunzel syndrome, named after the princess in Grimm's Fairy Tales who let her hair down a tall tower for her prince. (2)

Patients who have Rapunzel syndrome usually exhibit trichotillomania and trichophagia. These disorders are often the result of psychological problems or mental retardation. In our patient's case, there was a prior history of trichophagia at 5 years old as well as a perfectionist personality. ?

CONCLUSION

In patients with clinical suspicion of small bowel obstruction and with consistent radiological findings, bezoars should be considered in the differential diagnosis. In young patients with psychiatric history or personality disorders it is particularly important to consider the possibility of fragmented trichobezoars (Rapunzel syndrome).



BIBLIOGRAPHY

- Jabra AA, Eng J, Zaleski CG, Abdenour Jr, Vuong HV, Aideyan UO et al. CT of Small-Bowel Obstruction in Children. American Journal of Roentgenology 2001; 177:2, 431-436
- Wang Z, Cao F, Liu D, Fang Y, and Li, F. The diagnosis and treatment of Rapunzel syndrome. Acta radiologica open 2016; 5:11, 1-4.