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Case Report

Rapunzel Syndrome in a Paediatric Patient

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Abstract

Rapunzel syndrome, or generically known as trichobezoar, is a rare condition. It usually happens among teenage population. We are presenting a case report of Rapunzel syndrome that happened in a 4-year-old child. She was initially investigated for nephrotic syndrome, as she had high blood pressure and hypoalbuminaemia. However, it was later found out to be a trichobezoar, indirectly causing both hypertension and malnutrition. This condition demanded a combination of surgical and psychiatric discipline for diagnosis and its treatment.

Keywords: Bezoar, foreign bodies, hair balls, nephrotic syndrome, trichotillomania

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Introduction

Rapunzel syndrome is first reported by Vaughan et al. in year 1968; influenced by a fairy-tale written by the Grimm's brothers (1). Rapunzel syndrome is a misnomer as it simply described hairball in the stomach, rather than a collective of symptoms.

It is a rare form of trichobezoar, which composed of human hairs in the stomach and usually extending into the oesophagus or the small intestine. It follows the shape of the stomach and has a long tail of hair, representing Rapunzel's hair. This condition is very rare and only less than 40 cases being reported so far (2). This is unique as it involves both surgical and psychiatric expertise.

Case Report

It was a 4-year-old girl who lived in the outskirts of Sabah (within the Borneo region of Malaysia) where she would require one-day journey to the nearest medical facility. She lived near a barber shop.

She was first presented to the Emergency Department with persistent high blood pressure for her age; in which she had systolic blood pressure ranging 130mmHg to 140mmHg; and diastolic pressure of 86mmHg to 90mmHg (more than 95th centile from her age and sex). She also had periorbital oedema and facial puffiness. She was hypovolaemic and unable to feed orally well. She didn't have any haematuria, or any preceding upper respiratory tract infection before. Thus she was admitted to investigate for nephrotic syndrome.

Blood investigations noted to have microcytic hypochromic anaemia with haemoglobin level of 6.5gm/dL. The renal profile was found to be deranged. The serum urea on admission was 12.1mg/dL, and creatinine of 124 μmol/L. Liver function test also had shown hypoalbuminaemia at 15gm/dL. Throat swab for

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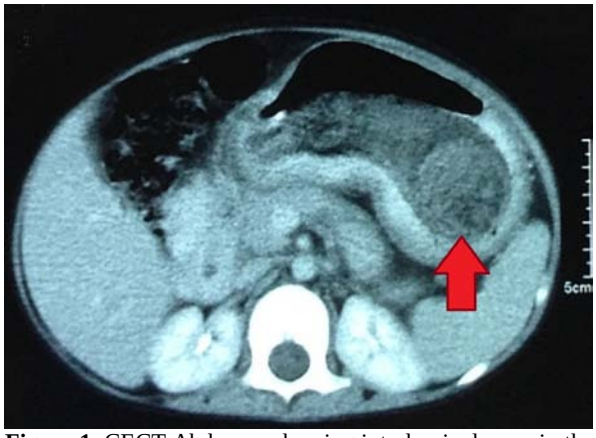

Figure 1: CECT Abdomen showing intraluminal mass in the stomach causing outlet obstruction (arrow)

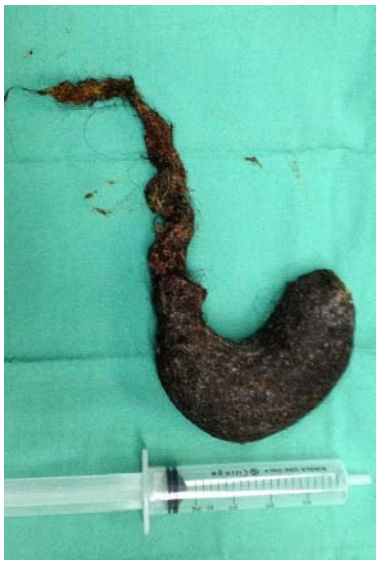

Figure 2: The trichobezoar removed en-bloc via gastrotomy through a mini-laparotomy

Streptococcus infection taken was negative. Biochemical test for urine also did not show any proteinuria.

In the ward, she was found to have a firm left upper abdominal mass, which was more to the left flank. Ultrasonography of the abdomen noted that the mass was solid and cystic at the left hypochondrium extending towards the midline. Both kidneys were normal in size and there was no hydronephrosis. Computed topography (CT) of the abdomen shown an intraluminal gastric mass. The mass was solid having air within encompassing the whole cavity of the stomach. The mass has extensions proximally into the oesophagus. It was concluded to be an intraluminal bezoar (Fig. 1).

An oesophagogastroduodenoscopy (OGDS) was done under general anaesthesia, anticipating open surgery for removal. During the scope, it was noted to have entangled hairball that filled up the whole gastric cavity and removing it via scope was unfeasible. Hence it was delivered via upper mini-laparotomy and gastrotomy (Fig 2). It was also noted that she had multiple Forrest III ulcers at the antrum and the duodenum.

Post-operatively she made an uneventful recovery. She was then referred to a psychiatrist. She had stopped eating hair from the barber shop after a few months of counselling and lifestyle changes.

Discussion

Rapunzel syndrome is associated with psychiatric disorder where there is a tendency to pluck own hairs (trichotillomania) and ingests it (trichophagia) (3,4). Majority of the reported cases comprise of teenage girls with long hairs (5). However we are reporting Rapunzel syndrome in a 4-year-old girl. She has no alopecia, but the child is known to be fond of playing at a nearby barber shop. This may suggests that she is only suffering from trichophagia.

Trichobezoar develops when indigestible and slippery human hair accumulates between the mucosal folds of the stomach, where it gets enmeshed. Over a period of continuous ingestion, propulsion and retropulsion nature of the stomach leads to the formation of hairballs. Trichobezoar can act as a foreign body in the cavity of the stomach or intestine, causing outlet obstruction. Besides, it can cause malabsorption as portrayed in the case reported.

In our case, the child initially masquerades as nephrotic syndrome. Her blood pressure was high on admission, could be attributed to the distension pain of the stomach as well as the multiple ulcers in the antrum and duodenum. Secondly, she had marked hypoalbuminaemia (15gm/dl), leading to periorbital oedema and facial puffiness. This is due to long standing malnutrition as she had an obstructed stomach. Malabsorption also causes iron deficiency anaemia.

Diagnosing Rapunzel syndrome requires high index of suspicions and also a detailed history (6). Imaging using computed tomography can diagnose foreign body in the stomach as well as to rule out any malignant condition in an upper abdominal mass. The management and treatment of a bezoar need to

encompass removal of the mass and prevention of recurrence by addressing the underlying emotional cause.

Upper endoscopy is the gold standard for diagnosing and removal of trichobezoar (7). In our case, endoscopic removal was abandoned as the trichobezoar was too huge and it cannot be shredded. Laparotomy is the cornerstone the management of trichobezoar with extension into the small bowel. Few of the research studies have shown laparoscopic-assisted removal of the hairballs in at least 2 cases reported (8).

Psychiatric evaluation and intervention is much needed in such cases. Intervention that could be done is aimed to prevent hair consumptions, thus recurrences. Medications such as quetiapine, hair-extension removal and also involvement of family members are crucial in treating the root of the problem (4,6).

Conclusion

Rapunzel syndrome has been described in the literature as having complications such as intussusception, protein-losing enteropathy, pancreatitis and gastric perforation (1). However, mortality from Rapunzel syndrome is rare. We would like to highlight that it can also masquerade as nephrotic syndrome, as it can share similar clinical features in long-standing cases. We present this interesting case as it poses a challenge in the diagnosis, however the treatment would be removal of the bezoar. There are numerous method of removing the offending foreign body though.

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