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Case Report

Trichobezoar masquerading as massive splenomegaly: Rapunzel's syndrome revisited ☆☆☆

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ABSTRACT

Trichobezoars are usually formed due to ingestion of hair or hair-like fibres and present with a wide spectrum of clinical manifestations. We report a case of Rapunzel's syndrome associated with trichotillomania in a 16-year-old girl who presented to our Haematology unit with complaints of fatigue, abdominal distention, and early satiety. Initial evaluation demonstrated anaemia, thrombocytosis, and a left hypochondrial mass suggestive of splenomegaly. However, ultrasound of the abdomen showed no hepatosplenomegaly and blood investigations were not suggestive of haematological malignancy. Not long after, the patient presented to the emergency department with suspected acute abdomen. Computed tomography of the abdomen revealed intraluminal gastric and jejunal masses causing small bowel obstruction. Emergency laparotomy confirmed gastric and jejunal trichobezoars, and subsequent psychiatric evaluation confirmed trichotillomania. Clinicians should consider trichobezoar in the differential diagnosis of abdominal pain and a non-tender 'spleen-like' abdominal mass.

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Introduction

First reported in 1968, Rapunzel syndrome is a rare form of gastrointestinal trichobezoar and is defined by trichobezoars

that pass beyond the gastric pylorus into the small or large intestines [1]. It carries a wide spectrum of clinical manifestations such as anorexia, abdominal pain, obstruction, intussusception, and peritonitis [2], and is often diagnosed in association with psychiatric disorders such as trichotillomania

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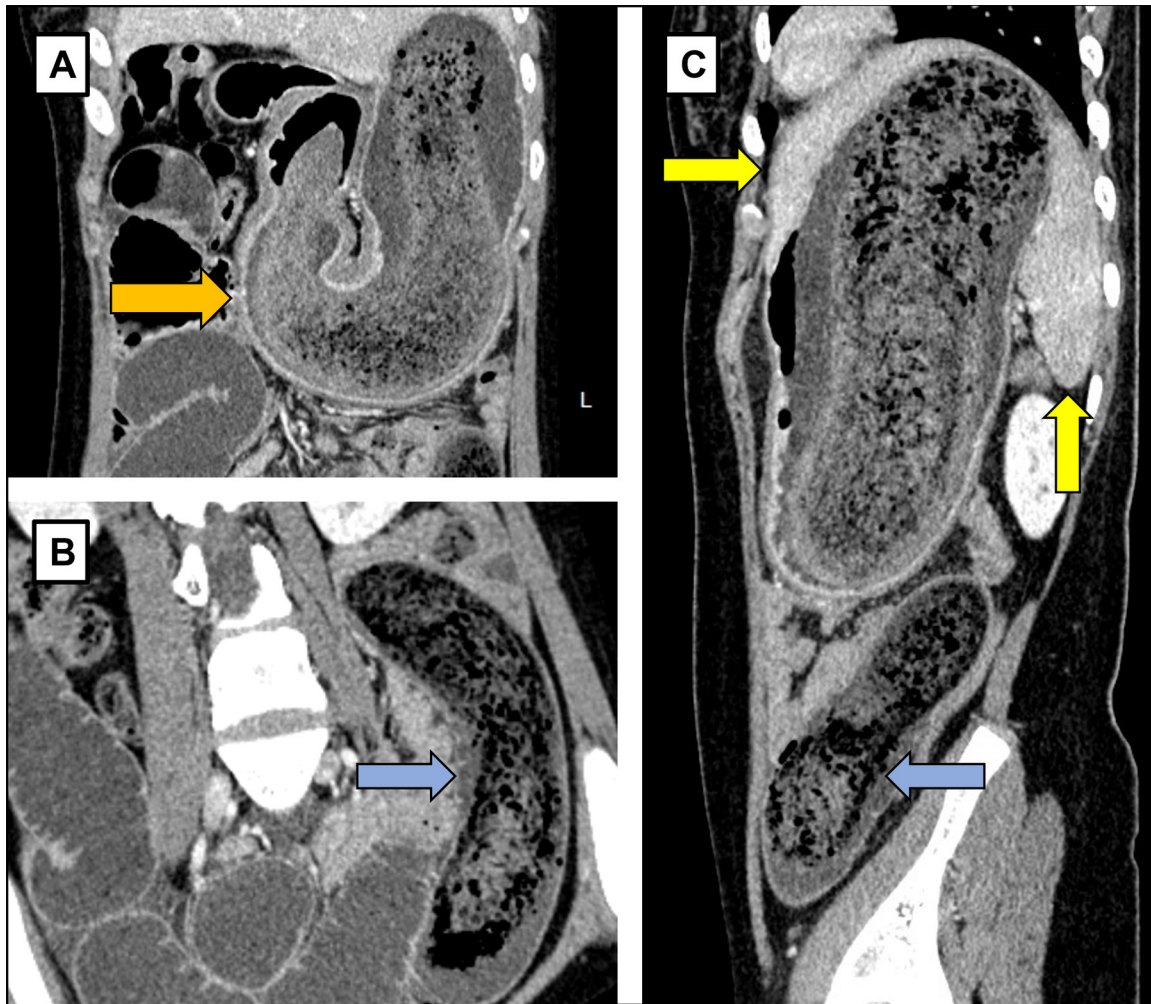


Fig. 1 – A. Computed tomography (CT) abdomen in coronal view showed a grossly distended stomach due to intraluminal trichobezoar (orange arrow). **B.** 3-dimensional multiplanar reconstruction of CT Abdomen demonstrates a trichobezoar in the jejunum (blue arrow). **C.** CT abdomen in sagittal view demonstrates displacement of the spleen (yellow arrows) by the grossly distended stomach (C). The jejunal trichobezoar is visualized inferiorly (blue arrow).

and trichophagia [3,4]. We report a case of trichotillomania in a 16-year-old girl that resulted in trichobezoars masquerading as massive ‘splenomegaly’.

Case report

A 16-year-old previously healthy indigenous Malaysian female presented to our Haematology unit with a 3-month history of fatigue, early satiety, and abdominal distention. She denied weight loss, fever, or night sweats. She attended a local school, was a non-smoker, and teetotaler. Physical examination revealed conjunctival pallor, distinct areas of hair loss over the scalp, and a large, palpable abdominal mass extending 20 centimetres below the left costal margin. The mass was firm and non-tender. There was no associated lymphadenopathy or hepatomegaly. Other systemic examinations were unremarkable.

Initial laboratory investigations revealed microcytic hypochromic anaemia (haemoglobin concentration of 10.4 g/dL, mean corpuscular volume of 77 fL, and mean corpuscular haemoglobin of 24 pg) attributed to iron deficiency (transferin saturation of 17% and ferritin of 4.8 ng/ml). Total white cell count was within normal range. Thrombocytosis was evident (platelet count of $683 \times 10^3/\mu\text{L}$), but peripheral blood film examination showed no leucoerythroblastic features or tear drop cells. Myeloproliferative neoplasm was initially suspected in view of clinically evident splenomegaly, anaemia, and thrombocytosis. However, screening for both JAK2V617F and calreticulin (CALR) were negative. Autoimmune screening was negative. Subsequent abdominal ultrasonography ruled out hepatosplenomegaly – however, the presence of intense posterior bowel shadowing was noted.

Two weeks later, she presented to the emergency department of another hospital with complaints of severe abdominal pain and constipation. Computed tomography of the abdomen revealed small bowel obstruction due to an intralu-



Fig. 2 – Photograph of two trichobezoars removed intraoperatively from the stomach and jejunum, measuring approximately 22 centimetres and 10 centimetres in length respectively.

minimal mass within the jejunum. Another similar mass was visualized in the stomach causing gross gastric distension (Fig. 1). There was no hepatosplenomegaly or intraabdominal lymphadenopathy. Emergency laparotomy and gastrojejunostomy was performed. Intra-operatively, gross distension of the stomach and mid-to-distal jejunum was noted. Two large masses composed almost entirely of hair were removed (Fig. 2). Her post-operative recovery was uneventful. Subsequent psychiatric evaluation confirmed that the patient had trichotillomania and had been intentionally ingesting her hair for several months, as the result of intolerable stress in her academic pursuits and complicated interpersonal relationships. She was referred to a clinical psychologist to assist in stress management, habit reversal and cognitive behavioural therapy.

Discussion

Trichobezoars occur due to irresistible hair-pulling, stroking, licking, chewing, and eating of hair by patients with trichotillomania – an ‘impulse control disorder’ [3,4]. Other types of bezoars include phytobezoars (food and vegetable matter), trichobezoars (hair and fur), lactobezoars (undigested milk and mucus usually found in neonates) and pharmacobezoars (medications) [5]. The incidence of trichobezoars has been re-

ported to be less than 1 percent, with approximately half of the cases being gastric trichobezoars [6]. Only about 30 cases of Rapunzel syndrome have been described in literature since its initial conception, with approximately a quarter of them presenting with intestinal obstruction [2], while nearly half of them may present as an abdominal mass [7].

Abdominal X-rays of a bezoar often reveal non-specific shadowing and may be mistaken for abscess or faeces in the colon [8]. Abdominal ultrasound may demonstrate a band of increased echogenicity [9]. In retrospect, our patient’s ultrasonography finding of posterior bowel shadowing is explained by compacted hair being a very echogenic material [9]. Computed tomography is more sensitive than ultrasonography in diagnosing trichobezoars and allows differentiation of trichobezoars from pseudocysts or tumours [6]. While imaging modalities may aid diagnosis, thorough history taking is necessary in confirming the diagnosis [7].

Surgical laparotomy or laparoscopy accompanied by psychiatric consultation remains the mainstay of management of Rapunzel syndrome [7]. Complete endoscopic removal of a trichobezoar is often impossible due to its large size relative to the endoscope’s forceps [10]. Furthermore, there is an added risk of endoscopic fragmentation leading to distal migration of the smaller pieces of the trichobezoar [10].

Trichobezoars presenting with iron deficiency anaemia and clinical “splenomegaly” are rarely reported and reflects the chronicity of the condition in our case. It is estimated that trichobezoars take approximately 6 months to become symptomatic [11], once again highlighting the importance of careful history taking.

Conclusion

The presence of a left hypochondrial mass, anaemia and thrombocytosis in a young individual often raises the alarm for myeloproliferative neoplasm and warrants further investigation. However, in the absence of positive genetic mutation screening (JAK2 or CALR) or radiological evidence of hepatosplenomegaly, the astute clinician should revisit the patient’s history with careful focus on psychosocial considerations.

Author contributions

All the authors were directly involved in the management of the patient. AZYK, SL, LEN and TLL acquired the clinical data. ASOT, AZYK and LEN prepared the final manuscript. All the authors approved the final manuscript.

Ethical approval

This article contains a study of a human participant and was registered via National Medical Research Register Malaysia with a Research ID of NMRR-21-192-58670.

Patient consent

Written informed consent was obtained from the patient and her parents for the anonymised information to be published in this article.

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