



Rapunzel Syndrome: Sonography and Computed Tomography of Trichobezoar

라푼젤 증후군: 모발위석의 초음파 및 전산화 단층촬영소견

Andre Tjie Wijaya, MD^{1*}, Budiawan Atmadja, MD²

¹Department of Radiology, Faculty of Medicine Udayana University, Sanglah General Hospital, Denpasar-Bali, Indonesia

²Department of Radiology, PGI Cikini Hospital, Jakarta, Indonesia

Trichobezoar is caused by hair ingestion that accumulates in the gastrointestinal tract. It is associated with psychiatric disorders. Occasionally, the hairs invade through pylorus and cause obstruction, which is known as Rapunzel Syndrome. A 9-year-old girl was admitted with signs of intestinal obstruction and a lump at her left side abdomen. Patient had history of trichotillomania and trichophagia. Abdominal ultrasonography and enhanced abdominal computed tomography were performed, which revealed a hair ball. Trichobezoar and Rapunzel Syndrome are bizarre and uncommon diseases. The case demonstrated that imaging modalities can accurately diagnose these diseases.

Index terms

Bezoar
Tomography, X-Ray Computed
Syndrome
Trichobezoar
Ultrasonography

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*Corresponding author: Andre Tjie Wijaya, MD
Department of Radiology, Faculty of Medicine Udayana University, Sanglah General Hospital, Denpasar-Bali 80114, Indonesia.
Tel. 62-0361-227911 ext 384 Fax. 62-0361-224206
E-mail: andre.tjie.wijaya@gmail.com

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INTRODUCTION

Bezoars are the accumulation of ingested materials. Different types of bezoars can occur in human and their name reflects the ingested material, such as vegetable or fruit fiber (phytobezoar) and milk curd (lactobezoar). The most common type of bezoar is trichobezoar, which is made of hairs (1, 2).

Trichobezoars are formed in the stomach but sometimes may pass through the pylorus into small intestines (Rapunzel Syndrome). The name came after Grimm Brothers' fairy tale in 1812 about a 12-year-old maiden who lowered her long hair from a tower to let her prince climbing up the tower and rescue her. The syndrome was discovered for the first time by Vaughan in 1968 and since then approximately 30 cases have been reported in the literature (1-4).

CASE REPORT

A 9-year-old girl complained about intermittent abdominal pain that subsided after vomiting since 1 year ago. She had a poor appetite and early satiety. There was also a lump in her abdominal. Patient had a history of Trichotillomania and Trichophagia since the age of 2 years and never received psychiatric treatments before.

Ultrasonography (US) was performed using 3.5 MHz transducer ultrasound (MyLabTMClassC; Esaote, Genoa, Italy). Abdominal US on the mass revealed an arc-like hyperechoic curvilinear with posterior acoustic shadow (Fig. 1A). Computed tomography (CT) examination was performed using a 64-channel multi detector CT (Somatom Definition AS; Siemens, Erlangen, Germany). On CT, there was a heterogenous intragas-

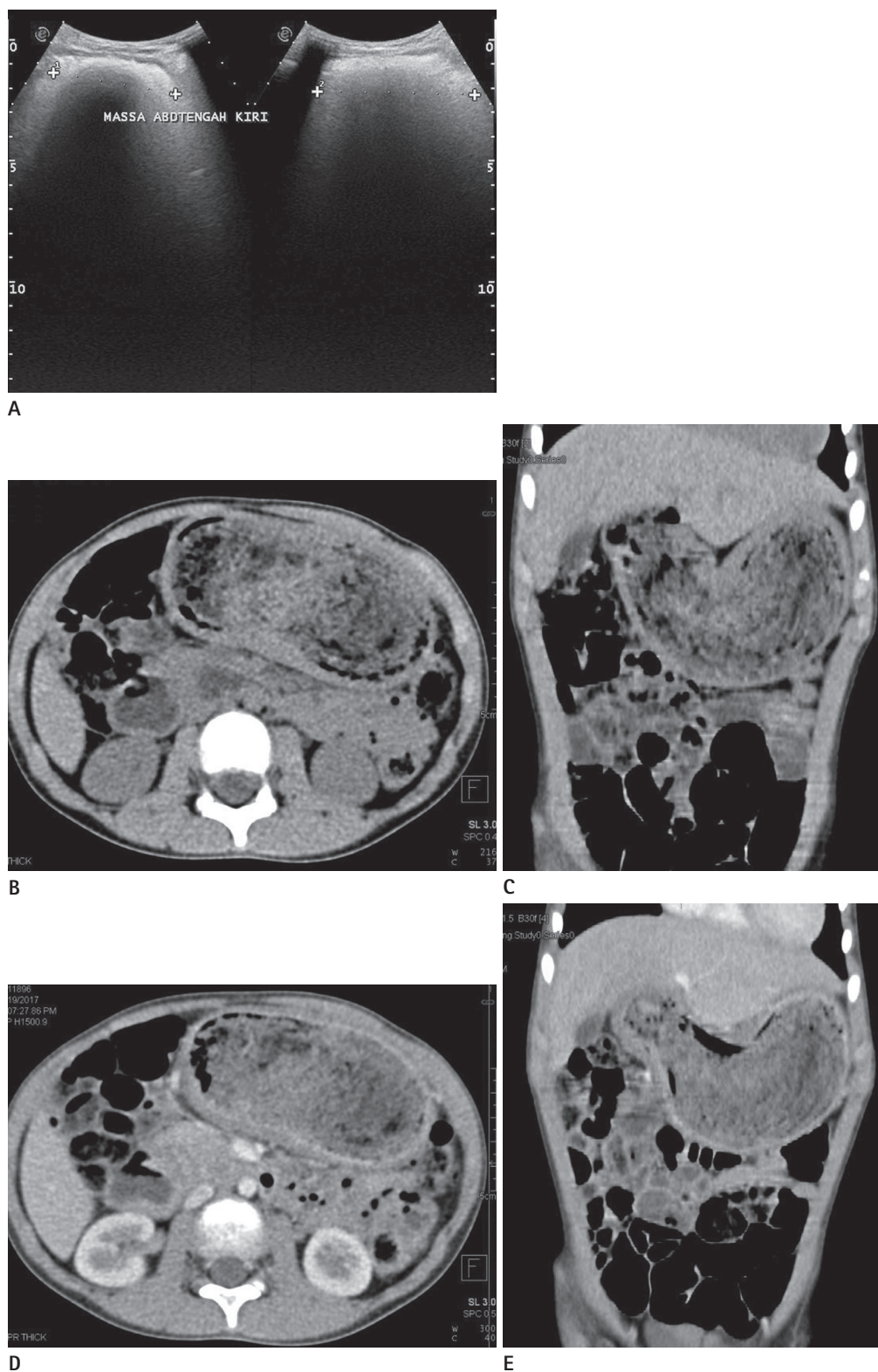


Fig. 1. A 9-year-old girl with signs of intestinal obstruction, a lump at her left side abdomen and history of trichotillomania and trichophagia. **A.** Sonogram of left upper abdomen shows hyperechoic curvilinear casting with clear posterior acoustic shadow. **B-D.** (B) and (C) Axial and coronal section of non-contrast CT show intragastric mass with heterogeneous density and air-bubble within it (D) and (E). On enhanced CT, there is no enhancement in the mass. CT = computed tomography



F

Fig. 1. A 9-year-old girl with signs of intestinal obstruction, a lump at her left side abdomen and history of trichotillomania and trichophagia.

F. The hair ball had 11 cm long of gastric part and 6 cm long of duodenum part. It has a perfect cast of the stomach, pylorus, and superior part of duodenum with weight about 1 kg.

tric mass that extended to superior part of duodenum with air bubbles within and there was no enhancement in the mass with intravenous contrast administration (Fig. 1B-E).

The patient underwent a laparotomy and a hair ball, which weighed about 1 kg, was removed. It had a perfect cast of the stomach, pylorus, and superior part of duodenum (Fig. 1F). After the surgery, psychiatric consultation was scheduled and the patient was discharged home 6 days after the surgery.

DISCUSSION

Trichobezoars are associated with psychiatric disorders including Trichotillomania and Trichophagia. Most of cases have been reported in young females and in countries where women traditionally have long hair (1, 4, 5).

The ingested hairs escape from peristaltic because of their slippery surface and detained in the gastric mucosa folds. Gastric peristalsis causes hairs to be enmeshed into a ball shape. The hairball becomes even more matted together and casts the shape of the stomach, as a solid mass. Because of the hold up by the pylorus and the churning action of the stomach, the hairball usually located in the stomach (1, 3, 5).

The gold standard for diagnosis trichobezoar is upper gastrointestinal endoscopy (2, 4). Even though upper gastrointestinal is the gold standard diagnosis for trichobezoar, it may not prove

the presence of a co-existing Rapunzel Syndrome.

The diagnosis of Rapunzel Syndrome depends on the use of CT and intraoperative finding both. However, there are various diagnostic criteria to describe Rapunzel Syndrome. First is a trichobezoar with a tail extend up to ileocecal junction; other criteria is a trichobezoar with a long tail extend up to jejunum or beyond; or as a trichobezoar of any size which can cause intestinal obstruction (1, 3).

The sonographic appearance of trichobezoar was a hyper-echoic curvilinear (arc-like) with acoustic shadow. However, there were difficulties in revealing multiple bezoars and unable to locate bezoars that far from the abdominal surface (6, 7).

CT is a superior to other radiologic tools for trichobezoars diagnosis and excludes differential diagnosis in patients with intestinal obstruction. Hairballs appear as non-enhanced, well-described, heterogenous, intraluminal mass with mottled appearance. The heterogenous and mottled appearances come from different densities of its material, including food debris and air-bubble (2, 7).

Trichobezoar and Rapunzel Syndrome are bizarre, uncommon diseases. Radiologists should be able to recognize and diagnose them precisely. Imaging plays an important role to make a diagnosis and exclude the differential diagnosis in young women with symptoms of intestinal obstruction. In order to avoid complications, an early and accurate diagnosis is important, which is able to be made by imaging.

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Andre Tjie Wijaya^{1*} · Budiawan Atmadja²

모발위석은 모발섭취로 인해 위장관에 축적되어 발생하며 정신질환과 관련있다. 섭취한 모발이 유문을 지나 장폐쇄를 발생시킨 이 희귀병은 라푼젤 증후군이라고도 불린다. 좌복부의 소괴와 장폐쇄 증상을 보인 9세 여자 소아 환자가 입원하였다. 환자는 발모광과 식모벽의 병력을 갖고 있었다. 복부 초음파와 조영전산화단층촬영을 시행하였으며 모발 덩어리가 발견되었다. 모발위석과 라푼젤 증후군은 희귀하며 일반적이지 않은 질병이다. 모발위석과 라푼젤 증후군을 영상으로 정확하게 진단하는 증례이다.

¹우다야나대학교 상글라종합병원 방사선과, ²PGI 치키니병원 방사선과