Rapunzel Syndrome: a case report

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Abstract

Background: Trichophagia secondary to trichotillomania can be a potentially life-threatening condition if left untreated. Trichobezoars consist of hairballs in the lumen of the digestive tract; when it extends past the duodenum it is called Rapunzel Syndrome. Although trichobezoars are not uncommon, they are rare in psychiatric literature. The authors present a clinical case of Rapunzel syndrome and discuss some relevant aspects concerning the identification and treatment of this entity.

Case Report: This report documents the case of a 21-year-old female patient with a trichobezoar submitted to surgical intervention. She had a history of hair pulling since the age of 4 with intermittent psychiatric follow-up.

Conclusions: Early diagnosis and intervention are crucial, but not always sufficient to avoid serious complications. Further research is needed to increase knowledge regarding the etiology and treatment of this psychiatric condition.

Keywords: Trichotillomania, Trichophagia, Trichobezoar, Rapunzel Syndrome.

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Introduction

Despite its long history in medical literature, trichotillomania and trichophagia have received little research attention[1].

The prevalence of trichotillomania ranges from 0.6%–1.6% when DSM-IV criteria are used [2]. Among those affected only 30% will engage in trichophagia, and of these, only 1% will require surgical removal [3]. Trichotillomania is more common in girls [4] and the age at onset is bimodal, either in early childhood or in adolescence [5].

While rare, the formation of trichobezoars (hairballs) is the most serious medical complication of trichotillomania and subsequent trichophagia [4].

Bezoars are foreign bodies occupying the gastric cavity [6] and can be classified in: phytobezoar, trichobezoar, lactobezoar and miscellaneous [7]. When the trichobezoar extends from the stomach to various lengths of the intestine is called “Rapunzel Syndrome”, as an allusion to the fairy tale [8].

In DSM-IV-TR, trichotillomania is classified as an impulse control disorder not otherwise specified [9]. However, some believe it should be more appropriately classified as an obsessive-compulsive spectrum disorder because both share similar patterns of comorbidity and a familial and/or genetic relationship [3].

Recent research suggests that trichotillomania is a heterogeneous disorder characterized by different pulling styles [10] and different clinical presentations. Patients usually pull hair from the scalp, but they can also pull it from eyelashes, eyebrows, legs, armpits, and pubic regions [4].

Clinical studies referred to these pulling styles as automatic (subjects pull their hair while engaged in other activities [10]) and focused pulling [10] (associated with negative affect, with tension before and relief after pulling [4], patient’s time and attention are directed specifically for the purpose of hair pulling), which may coexist in the same patient [4].

In the more common “automatic” type, subjects are unaware that they are pulling their hair at all or become aware only when upon discovering it in their hands [10].

The majority of patients with hair-pulling (70%) report the presence of other body-focused repetitive behaviors, most commonly skin-picking and nail-biting [11].

Although data on treatment efficacy is inconclusive, common modalities include cognitive behavior therapy and pharmacological treatment [3, 12]. Several pharmacological modalities have been used corresponding to distinct views on the etiology of trichotillomania, including impulse-control disorders, obsessive-compulsive disorders, behavioral problems and addiction[13].

Case report

A 21 year-old Caucasian woman reported clinical symptoms with four months evolution, consisting of epigastric pain, nausea and vomiting mostly at night, and weight loss.

After first evaluation an upper endoscopy and an abdominal ultrasound were performed, both with inconclusive results. Later a CT-scan reported a distended stomach, filled with an heterogeneous density material, probably a trichobezoar. She was referred to a surgery department of a general hospital. The treatment consisted of an anterior gastrotomy with removal of a large bezoar with a tail which extended through the pylorus. The patient was then referred for psychiatric evaluation by the liaison psychiatry team.

No previous history of a trichobezoar was reported. She is single, living with her parents, grandparents and a younger brother (18 years old). They are small farmers, and both siblings helped in the family business. Having completed the ninth degree, including a catering course, she had until then several jobs in restaurants since the age of eighteen. Now she works in a bakery.

There was no known familiar psychiatric history and she had an uneventful developmental growth. At the age of four, when her brother was born, she started pulling her own hair and eating it. She refers a tense jealous relationship. She describes nail-biting, in hands and feet, since then until the age of eighteen. Since the age of six years, she began attending a child psychiatrist as well a psychologist, and alternative medicines, but this behavior never went under control. She reported having satisfactory social and romantic relationships.

During psychiatric follow-up, the patient showed no depressive, obsessive, psychotic or other psychiatric symptoms. She mentioned that on most occasions, she did not remember pulling her hair, but she eventually saw it in bed and sometimes in feces.

The patient was introduced to fluvoxamine up to 100mg, showing improvement in hair pulling episodes. She is now wearing a wig as she had her hair shaved due to the alopecia spots.

Discussion

Case reports of trichobezoars are common in surgery practice but rare in psychiatric literature. Collaboration with liaison psychiatry is in this instance especially relevant to avoid delay in treatment.

Trichotillomania and trichophagia should be considered in patients with hair loss as early diagnosis and intervention are important to avoid possible fatal complications.

Interesting about this case was the birth of a younger brother acting as a psychological stressor triggering this behavior.
An accurate diagnosis is necessary but not always sufficient. In this case early age diagnosis did not alter the course of illness, leading up to surgery.

This particular patient started fluvoxamine according to the obsessive-compulsive spectrum hypothesis, however the correct classification and treatment approach for this syndrome remains controversial.

Further research is needed to guide the clinician's choice of treatment for this condition.

Competing interests
The authors declare no conflict on interest.

References