Multiple and synchronous squamous cell carcinoma of the esophagus in a young woman: An example of early and rapid carcinogenesis?

Carcinoma epidermoide de esófago múltiple y sincrónico en una mujer joven: ¿un ejemplo de carcinogénesis temprana y rápida?

Tumor multicentricity is occasionally observed in esophageal squamous cell carcinoma. It is linked to genetic instability of the p53 gene, added to the loss of heterozygosity, as early events of the carcinogenesis of multifocal or multiple squamous cell carcinoma of the esophagus, and to risk factors associated with the environment and lifestyles.

Case presentation

A 47-year-old woman, who was a heavy smoker for more than 20 years, sought medical attention for moderately intense, continuous retrosternal chest pain of 3-month progression that was exacerbated by food intake, and for dysphagia with solid foods, and weight loss. Physical examination revealed tachycardia, dyspnea, cachexia, and bad teeth. Upper gastrointestinal endoscopy study identified the presence of 2 lesions with a tumor-like aspect in the middle and lower third of the esophagus. They exhibited an elevated level of anular and stenosing plaque growth, respectively (Fig. 1). Afterwards, transhiatal esophagectomy through left cervicotomy was performed. The pathology study reported well differentiated multiple squamous cell carcinoma with anular stenosing growth infiltrating up to the periesophageal fibroadipose tissue with moderately differentiated squamous cell carcinoma foci at the level of the serosa and periesophageal tissue, regional lymph node metastasis, and multifocal lymphatic and blood vascular invasion (Fig. 2). The patient had satisfactory postoperative progression.

This is a very unusual case in a young woman, a heavy smoker, presenting with multifocal squamous cell carcinoma. It contrasts with the habitual esophageal squamous cell carcinoma predominantly observed in men above 60 years of age. The particular circumstance of this patient could be linked to the possible coexistence of genetic instability of the p53 gene, added to the marked and prolonged smoking habit. Mutations in the p53 gene, together with the loss of heterozygosity, constitute early events of carcinogenesis.

Figure 1  A) Tumor in the middle third, showing increased plaque growth. B) Tumor in the lower third showing elevated annular and stenosing growth.

multiple step esophageal carcinogenesis of multiple primary squamous cell carcinoma. In addition, the marked and prolonged smoking habit is also closely related to the events of multicentric carcinogenesis in the upper gastrointestinal tract, including the esophagus. Saeki et al. established the fact that cigarette smoking is clearly associated with an increased frequency in the accumulation of the p53 protein in cases of multiple primary squamous cell carcinoma. Consequently, this mixture could solidify early and rapid carcinogenesis, with the subsequent development of multifocal squamous cell carcinoma, as observed in the present case.

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Conflict of interest

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References

Rapunzel syndrome: A rare cause of abdominal pain

Síndrome de Rapunzel: una causa rara de dolor abdominal

A bezoar is an accumulation of organic substances within the gastrointestinal tract. Rapunzel syndrome is a rare and infrequent form of a trichobezoar with a tail that extends beyond the stomach into the small bowel.

We present herein the case of a 10-year-old girl referred to our center for abdominal pain. The patient had a past history of trichophagia of several-week progression, chronic abdominal pain, unmeasured weight loss, and predominantly postprandial nausea and gastric vomiting for the past 7 days.

Upon physical examination, the patient was dehydrated, with generalized pallor, abdominal distension, and a palpable and painful mass in the epigastrium.

Laboratory work-up reported hemoglobin of 8.7 g/dl (12-15 g/dl), hematocrit of 26.9% (36-46%), leukocytes 9,250/mm³ (5,000-10,000/mm³), and platelets 274 x 10³ mm³ (150-400/mm³). The rest of the laboratory tests were normal. Computed tomography scan identified an intragastric tumor extending into the duodenum that had no contrast medium enhancement (Fig. 1).

Exploratory laparotomy revealed a large intraluminal mass in the stomach. After gastroscopy, a large trichobezoar was found that extended into the small bowel, and it was removed. The patient did not present with any postoperative complications and was released on the 6th day after the procedure (Fig. 2).

Following the medical-surgical treatment, the patient was diagnosed with depressive disorder, anxiety syndrome, and poor self-esteem. She was initially treated at the medical psychology service and later referred to the pediatric psychiatry service at another center for her integrated treatment.

Rapunzel syndrome was first described by Vaughan in 1968. Since then, fewer than 100 cases have been described in the medical literature, with varying clinical characteristics. Different authors have used different criteria for reporting their cases of Rapunzel syndrome. Some have defined it as a gastric trichobezoar with a tail extending to the ileocecal valve, others have described it simply as a trichobezoar with a long tail that can extend into the jejunum, ileum, or ileocecal valve, and still others have defined it as a trichobezoar of any size that presents with bowel obstruction.

Trichobezoar, a hairball in the proximal part of the gastrointestinal tract, is a rare condition and is seen almost exclusively in young women. Human hair is resistant to digestion and peristalsis because of its smooth surface, and thus accumulates in the folds of the stomach mucosa. After a period of time, the continued ingestion of hair results in its impaction, together with mucus and food, causing the formation of a trichobezoar. In the majority of cases, it is confined to the stomach. Nevertheless, in some cases, the trichobezoar extends beyond the pylorus into the jejunum, ileum, and even the colon.

Patients with bezoars usually present with nonspecific gastrointestinal symptoms, such as abdominal pain, anorexia, and vomiting. Severe complications can occur, including obstruction, bleeding, or perforation.

The most common presentation characteristics are abdominal pain (37%), nausea and vomiting (33%), obstruction (25%), and peritonitis (18%). Although not very common, patients can also present with weight loss (7%), anorexia, hematemesis, and intussusception (7%). The majority of patients deny any past history of trichotillomania or trichophagia, even when specifically asked. Therefore, psychiatric evaluation and management is recommended.

Diagnostic studies include ultrasound, tomography, and panendoscopy. Computed axial tomography has a high accuracy rate, but ultrasound precision is not as high in these cases. Computed tomography reveals a well-circumscribed lesion in the gastric region composed of concentric spirals of different densities with air bubbles mixed up inside them. Oral contrast medium fills the most peripheral interstices of the lesion and a thin band of contrast medium circumscribes it. The absence of filling upon intravenous contrast medium-enhancement rules out a neoplastic lesion.

Endoscopy plays the most important role in gastric bezoar detection, as well as in treating some of them. Tomography is useful for detecting small gastric bezoars and those in the small bowel. A tomography scan is particularly valuable in patients that need surgical management of small bowel