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CASE REPORT

Adenocarcinoma of the esophagogastric junction presented as a vegetative tumor in the hiatal hernia sac

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Abstract

Introduction: The incidence of esophageal and esophagogastric junction adenocarcinomas has been increasing in developed countries, with primary risk factors including gastroesophageal reflux disease (GERD), smoking, and obesity. Chronic GERD can lead to Barrett's esophagus (BE), a premalignant condition that elevates the risk for esophageal adenocarcinoma.
Case Report:| We present a case of a 63-year-old woman with nausea and epigastric pain. Her history was notable for Sjögren's syndrome and a positive family history of prostate cancer, without smoking or alcohol use. Endoscopy revealed a distal hiatal hernia and long-segment Barrett's esophagus (LSBE) with a 3 cm ulcerated lesion at the gastroesophageal junction, located within the hernia sac. Biopsies confirmed well-differentiated adenocarcinoma in LSBE. CT imaging showed a tumor extending through the hiatal hernia with distal esophageal wall thickening and lymph node metastases but no distant metastases. The patient underwent a subtotal esophagectomy with lymphadenectomy and esophagogastric anastomosis using the Ivor Lewis procedure. Pathology confirmed adenocarcinoma with lymph node metastasis and a close circumferential resection margin. Six cycles of postoperative chemotherapy with platinum and fluoropyrimidine were administered. The patient was recurrence-free at a three-year follow-up.
Conclusion: This case underscores the association between hiatal hernia, Barrett's esophagus, and esophageal adenocarcinoma, particularly in the setting of disrupted antireflux mechanisms. The findings support the importance of early investigation and monitoring in patients with GERD and BE to mitigate cancer progression.

Keywords: **esophagogastric junction cancer, gastroesophageal reflux disease, hiatal hernia**

Adenokarcinom ezofagogastričnog spoja prezentovan kao vegetativni tumor u sakusu hijatusne hernije

Sažetak

Uvod: Incidenca adenokarcinoma jednjaka i ezofagogastričnog spoja raste u razvijenim zemljama, a glavni faktori rizika uključuju gastroezofagealnu refluksnu bolest (GERB), pušenje i gojaznost. Hronični GERB može dovesti do Barrettovog jednjaka (BJ), premalignog stanja koje povećava rizik za adenokarcinom jednjaka.
Prikaz slučaja: Predstavljamo slučaj 63-godišnje žene sa mučninom i epigastričnim bolom. U anamnezi se navodi Sjogrenov sindrom i pozitivna porodična istorija raka prostate, bez pušenja i konzumacije alkohola. Endoskopijom je otkrivena distalna hijatalna hernija i Barrettov jednjak dugog segmenta (BJDS) sa ulcerisanom lezijom prečnika 3 cm na ezofagogastričnom spoju, unutar hernijskog sakusa. Biopsije su potvrdile dobro diferentovani adenokarcinom u BJDS. CT snimak je pokazao tumor koji se širi kroz hijatalnu herniju, sa zadebljanjem distalnog zida jednjaka i

metastazama u limfnim čvorovima, bez udaljenih metastaza. Pacijentkinji je urađena subtotalna ezofagektomija sa limfadenektomijom i ezofagogastričnom anastomozom metodom Ivor Lewis. Patologija je potvrdila adenokarcinom sa metastazama u limfnim čvorovima i blizak cirkumferentni resekcioni margina. Nakon operacije primenjena je hemoterapija u šest ciklusa sa platinom i fluoropirimidinom. Na kontrolnom pregledu tri godine nakon operacije, pacijentkinja je bila bez recidiva.

Zaključak: Ovaj slučaj naglašava povezanost između hijatalne hernije, Barrettovog jednjaka i adenokarcinoma jednjaka, naročito u uslovima poremećenih antirefluksnih mehanizama. Nalazi podržavaju važnost ranog ispitivanja i praćenja pacijenata sa GERB-om i BJ kako bi se smanjio rizik od progresije raka

Klučne reči: **karcinom gastroezofagealnog prelaza, gastroezofagealna refluksna bolest, hijatus hernija**

Introduction

Esophageal and esophagogastric junction cancers are showing an increasing incidence in developed countries. Over the last few decades, there has been an increase in oesophageal and gastric cardia adenocarcinomas incidence, compared with stable or declining rates for oesophageal squamous cell carcinoma.[1,5].

Risk factors for esophageal adenocarcinoma are gastroesophageal reflux disease (GERD), tobacco smoking and excess body weight, whereas high dietary intake of fruit and vegetables may prevent the development of esophageal cancer of any histological type. Long-standing gastroesophageal reflux disease may induce Barrett's esophagus (BE), a premalignant condition presented by epithelial metaplasia of the distal part of the esophagus[1-5].

Patients that present with symptoms like dysphagia, weight loss, loss of appetite, gastrointestinal bleeding, recurrent aspiration or emesis should be further investigated and undergo an upper intestinal endoscopy [6].

We report a case of a 63-year-old woman, diagnosed as adenocarcinoma of the esophagogastric junction, presenting as a tumor in the hiatal hernia sac.

Case Report

A primary care physician refered a 63-year-old woman to a gastroenterologist regarding complaints of nausea and pain in the stomach. Her past medical history included Sjogren sy. There was no history of smoking, alcohol consumption, and previous surgery. She had the positive family history of a father diagnosed with prostate cancer.

Outpatient workup for persistent abdominal pain was inconclusive, leading to the recommendation for esophagogastroduodenoscopy (EGD), which showed the distal hiatal hernia at the gastroesophageal junction along with the long-segment Barrett's esophagus (LSBE) longitudinally measured 60 mm. In the hiatal hernia sac, there was the lesion of 3cm in diameter showing vegetative growth pattern and central ulceration (Figure 1).

Multiple biopsies were taken from these areas. The biopsy revealed LSBE with a well-differentiated adenocarcinoma.

Clinical examination and laboratory studies had unremarkable results. Computed tomography revealed gastric tumor through the hiatal hernia, as well as thickening of distal esophagus wall with prestenotic dilatation of oesophagus. Lymph node metastases were observed surrounding the stomach; however, liver metastases and peritoneal dissemination were not observed.

Following diagnosis of locally advanced resectable oesophageal cancer, subtotal esophagectomy with two-field lymphadenectomy was performed, and reconstruction was carried out with a gastric tube conduit with oesophagogastric anastomosis in the upper mediastinum (Ivor Lewis procedure). The postoperative pathological diagnosis was of well differentiated adenocarcinoma with lymph node metastasis and circumferential resection margin involvement of 0.4mm, pTNM8: T3 N1 (2/37) Mx L1 VO PN1, R1, stage IIIB. 6 cycles of postoperative ChT with a

platinum and fluoropyrimidine followed the surgery. The patient was found to be alive and recurrence-free three years after the surgery.

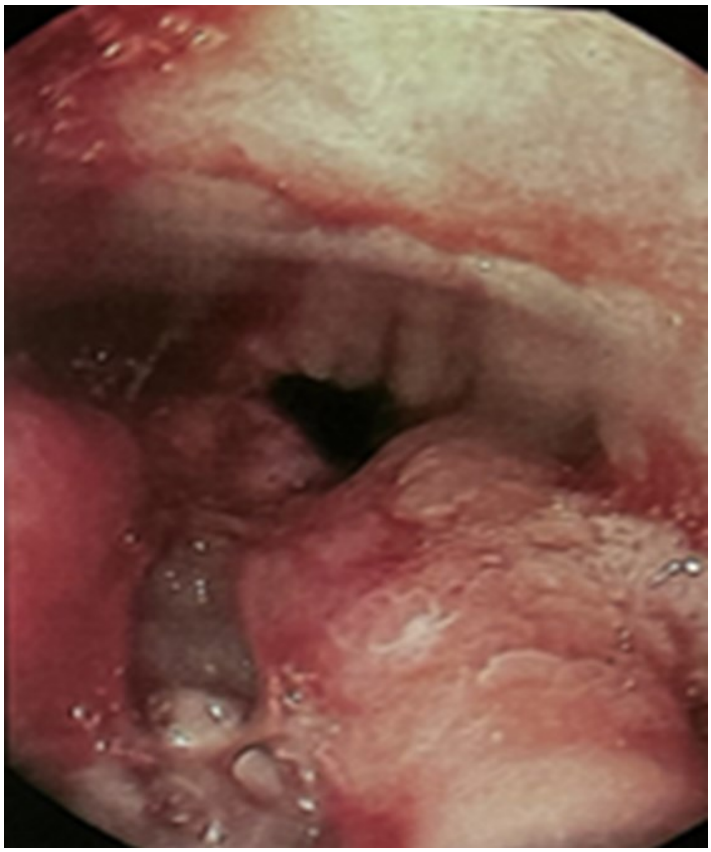

Figure 1: EGDS-verified tumor of the esophagogastric junction in a hiatal hernia

Discussion

According to the Siewert classification, adenocarcinoma of the esophagogastric junction (AEG) represent a tumor with an epicenter located within 5 cm of the esophagogastric junction, which is further classified into three subtypes according to the anatomic location of the tumor center.

In our case, the patient had Siewert type I, which is adenocarcinoma of the distal esophagus that usually arises from an area with specialized intestinal metaplasia of the esophagus - Barrett esophagus (BE), and may infiltrate the esophagogastric junction from above. The vast majority of the increase in incidence of adenocarcinoma of the esophagus is due to the rise of Barrett esophagus in young, otherwise healthy men - particularly in white people in the US and Western Europe[7,8].

EGD performed on our patient showed the distal hiatal hernia at the gastroesophageal junction along with the long segment BE longitudinally measured 60 mm. As studies have shown, there is strong relation between Barrett's esophagus and hiatal hernia, with hiatal hernia being present in a majority (72% to 96%) of patients with BE[9-11]. Moreover, it was shown that development of BE or progression to high-grade dysplasia or adenocarcinoma was related to the size of hiatal hernia. On the other hand, patients with hiatal hernia are significantly more likely to present with GERD symptoms than those without, and majority of patients with severe esophagitis have hiatal hernia[12-16].

These above mentioned associations between hiatal hernia and reflux symptoms, reflux esophagitis, Barrett's esophagus and esophageal adenocarcinoma are largely due to the disruption of many of the antireflux mechanisms that leads to increased esophageal acid exposure.

Conclusion

This case report highlights the complex interplay between gastroesophageal reflux disease, Barrett's esophagus, and the development of adenocarcinoma at the esophagogastric junction. The increasing incidence of esophageal adenocarcinomas, particularly in patients with long-standing reflux conditions and hiatal hernias, underscores the importance of vigilant surveillance and early intervention in at-risk populations. Our patient's case exemplifies the critical need for thorough investigation of gastrointestinal symptoms, particularly in individuals with predisposing factors such as Barrett's esophagus. Despite the challenges presented by her diagnosis, the successful surgical intervention and subsequent management illustrate the potential for favorable outcomes in cases of locally advanced, yet resectable esophageal cancer. Ongoing research into the mechanisms underlying these associations is essential to enhance early detection and refine treatment strategies, ultimately improving patient prognosis and quality of life.

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