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Management of a patient with primary adenocarcinoma of the proximal third of the esophagus: a case report

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Abstract

Background Cervical esophageal adenocarcinoma is a rare tumor often associated with Barrett's esophagus, and its prevalence is lower than distal esophageal adenocarcinoma. Upper esophageal malignancies are usually squamous cell carcinomata; about 5% of these cases are seen during upper gastrointestinal endoscopy.

Case presentation We present a patient (a 39-year-old Iranian man) with invasive adenocarcinoma of the proximal third of the esophagus, who was referred to the hospital with no endoscopic or histopathologic evidence of Barrett's esophagus. We discuss the probable cause of this tumor and treatment options and emphasize the importance of recognizing and managing this rare disease.

Conclusions This case highlights the rarity of this adenocarcinoma and emphasizes the potential of chemotherapy as a standalone treatment, as well as the need for personalized therapeutic strategies and further research for the management of esophageal cancer.

Keywords Primary adenocarcinoma, Barrett's esophagus, Esophagus cancer, Cancer, Case report

Introduction

Adenocarcinoma of the upper third of the esophagus is uncommon and has a dangerous prognosis. It may arise from the esophageal glands or ectopic gastric mucosa (EGM) [1]. Barrett's esophagus is a common risk factor [2]. The origin of primary adenocarcinoma in the cervical esophagus can be due to the "cardiac" glands of the mucosa, submucosa glands, or heterotopic gastric mucosa (HGM). However, studies have shown that little is known about the prognosis or ideal surgical treatments, chemotherapy, or radiotherapy [3]. EGM or gastric inlet patch occurs in approximately 2.5% of the population and rarely results from heterotopia of the stomach or submucosal glands. Some studies have reported only 58 cases of esophageal adenocarcinoma in EGM areas [4].

Although EGM is common, biopsies are rarely performed for evidence of dysplasia or adenocarcinoma. In the meantime, endoscopists need to be aware of its

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potential to have the right treatment approaches for its consequences by determining the exact type of adenocarcinoma. This report describes a 39-year-old man with invasive adenocarcinoma of the proximal third of the esophagus. The insights gained from this study can help us pay more attention to invasive adenocarcinoma patients and correctly identify the clinical features.

Case report

The case under study was a 39-year-old Iranian man without a significant risk factor in the past but with a complaint of progressive dysphagia in swallowing solid and liquid material for 3 months. He had no history of smoking or alcohol abuse. He had no complaints of hematemesis, melena, weight loss, altered defecation, abdominal pain, dyspnea, or cough. There was no significant family history or any other chronic disease such as diabetes, hypertension, or chronic obstructive pulmonary disease (COPD). His physical examination was normal. Blood biochemistry, including blood, liver, kidney function, CEA, and Ca19-9, were within normal limits (Table 1).

The workup included a barium swallow study (Fig. 1), which showed an irregularity, and partial stenosis in the lower part of the cervical esophagus. Endoscopy was recommended. Upper endoscopy showed ulcerated mucosa and mucosal irregularity in the upper third of the esophagus (15 cm from incisors) (Fig. 2). Pathological findings

Table 1 The laboratory findings of the patient

Variable (hematology test)		Normal reference range
WBC	5.13	4000–10,000 × 1000/cmm
RBC	6.06	4.50–6.30 mil/cmm
Hb	16	14–18 mmol/l
HCT	48	39–54%
PLAT	300	150–450 × 1000/cmm
FBS	98	70–115 mg/dl
BUN	42	19–44 mg/dl
CR	1	0.7–1.4 mg/dl
Total bilirubin	1	0.1–1.2 mg/dl
Direct bilirubin	0.4	Up to 0.3 mg/dl
AST	23	Up to 37 U/l
ALT	24	Up to 41 U/l
Serum iron	50	35–169 mμg/dl
TIBC	330	250–450 μg/dl
Ferritin	217.70	30–400 μg/ml

WBC white blood cell count, *RBC* red cell count, *Hb* hemoglobin, *HCT* hematocrit, *PLAT* platelet count, *FBS* fasting blood sugar, *BUN* blood urea nitrogen, *CR* creatinine, *AST* aspartate aminotransferase, *ALT* alanine aminotransferase, *TIBC* total iron binding capacity, *Cmm* cells per cubic millimeter, *mil/cmm* million cells/cells per cubic millimeter

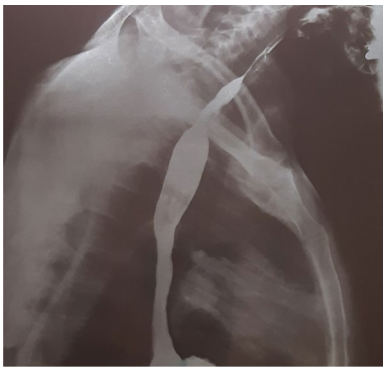

Fig. 1 Male patient's barium swallow, an irregularity, and partial stenosis in the lower part of the cervical esophagus

showed esophageal mucosa with an infiltrative neoplastic growth composed of malignant epithelial cells with large pleomorphic nuclei and gland formation (adenocarcinoma). In the endoscopic ultrasound (EUS) view, there was a heterogeneous hypoechoic lesion measuring 14.5 × 16 mm in diameter in the lateral aspect of the esophagus involving the mucosa and muscular mucosa and invading through muscularis propria into per esophageal connective tissue. Adjacent vasculature, including the carotid artery and jugular vein, adjacent muscles, prevertebral fascia, and thyroid, were not involved, and the interface was preserved (T3). No regional or extraregional lymph nodes were found as far as could be seen (N0; T3N0Mx). Cervical and thoracic computed tomography (CT) scans showed eccentric wall thickening of the lower cervical esophagus (Fig. 3). No evidence of cervical and mediastinal lymphadenopathy was seen. Contrast-enhanced CT (CECT) scan of the abdomen showed a normal liver, spleen, pancreas, and kidneys. No free fluid or lymphadenopathy was seen.

Chemoradiotherapy was delivered as oxaliplatin, 5-FU, and Taxol with intermittent radiotherapy. The patient's symptoms and signs were relieved over time, and the endoscopic image also improved (Fig. 4). With supportive treatment, oral intake and the general condition improved in the following 4 weeks. At the third monthly follow-up, the patient had an adequate semisolid and occasionally solid diet. His general condition also improved, with no additional complaints.

Discussion

Adenocarcinoma in the cervical esophagus is extremely rare and has been explained in the literature as based on ectopic gastric-type mucosa in the upper part of the esophagus. In contrast, adenocarcinoma of the distal esophagus is common. The relative prevalence has been increasing from 1.7% to 10% historically to more than

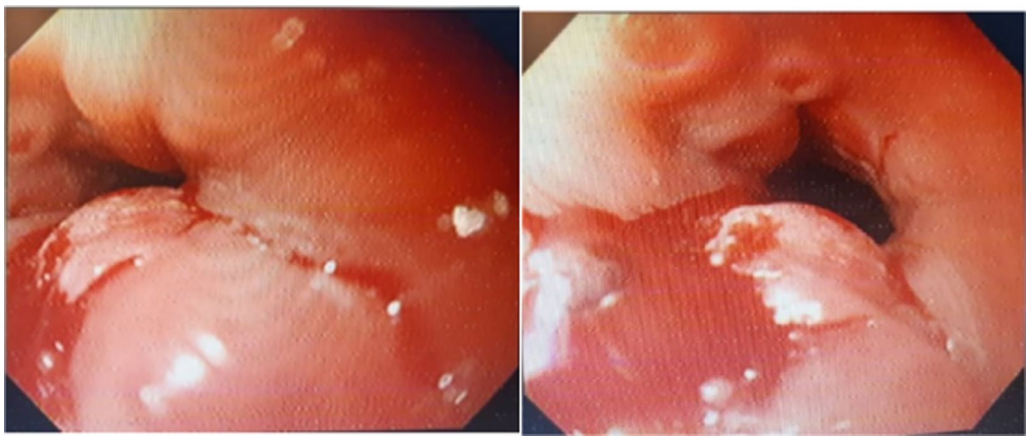

Fig. 2 Male patient's endoscopy findings: ulcerated mucosa with partial stenosis was seen in the upper third of the esophagus

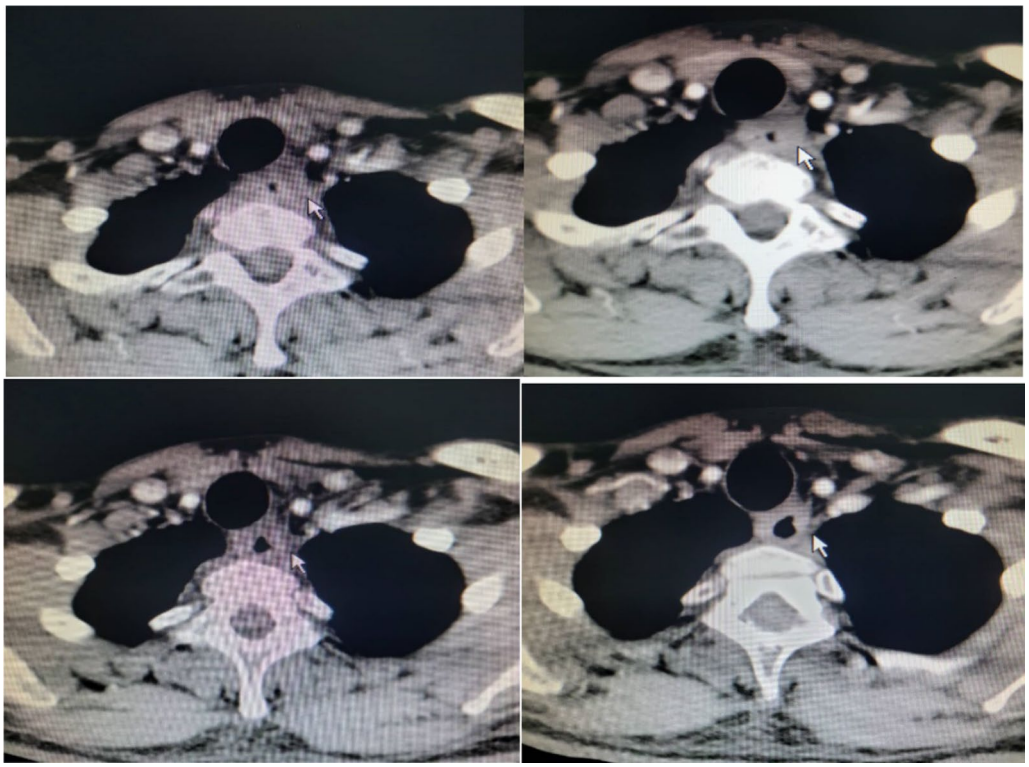

Fig. 3 Male patient's cervical and thorax CT scan findings: eccentric wall thickening of lower cervical esophagus

50% of all malignant tumors of the esophagus in more recent studies [5, 6]. The reasons for this shift in histology are not well established. Adenocarcinoma of the cervical esophagus can arise from mucosal "cardiac" glands, submucosal glands, and EGM [5, 7, 8]. HGM is thought to occur when embryologic bidirectional replacement of the esophageal columnar epithelium by squamous mucosa fails to be completed. HGM is most often asymptomatic and found incidentally during endoscopic procedures for unrelated symptoms in approximately 3.8–10% of the adult population [9, 10].

Riddiough *et al.*'s study suggested two different pathways for the pathogenesis of adenocarcinoma in EGM, including (1) induction of the metaplasia–dysplasia pathway via local secretory acid or reflex acid that leads to intestinal metaplasia and intestinal-type adenocarcinoma, and (2) intrinsic development of adenocarcinoma within gastric/foveolar cells in EGM leading to

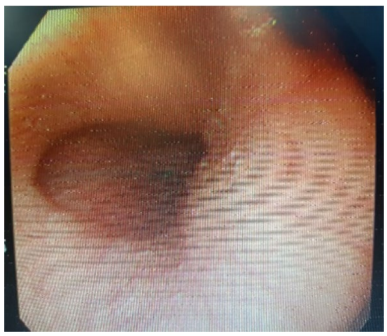

Fig. 4 Normal mucosa in the upper third of the esophagus

gastric type adenocarcinoma EGM [4]. EGM is common, while malignant progression is rare; nevertheless, biopsy is rarely performed for evidence of dysplasia or adenocarcinoma. Determining the exact type of adenocarcinoma on endoscopy may have implications for treatment approaches. Diagnosis of EGM on endoscopy may identify patients at higher risk for proximal esophageal adenocarcinoma; however, this relationship and the need for screening require further study [4]. The incidence of EGM is 4.0% in males and 2.9% in females [11].

No standard therapy exists for primary cervical esophageal adenocarcinoma [12]. Treatment strategies published in the few case reports range from endoscopic mucosal resection to laryngo pharyngo-esophagectomy with lymph node dissection with or without neoadjuvant or adjuvant chemoradiotherapy. In general, radical surgery is the mainstay of treatment, and radiotherapy is a viable alternative (especially when surgery is not possible). The treatment method should be considered on the basis of the disease, the patient's condition, and other parameters [13]. Due to the gradual increase in the number of cases of upper esophageal adenocarcinoma, it is necessary to standardize the treatment protocol [1]. However, the number of cases reported in the current literature is too small, and the details of their staging and treatment are inconsistent to postulate whether these tumors have a different prognosis than the more common distal esophageal adenocarcinomas [5].

Our patient presented with a relatively large, ultrasound-staged T3N0. His symptoms improved relatively with chemoradiotherapy, and his upper endoscopy (4 months later) showed erythema and mucosal healing without ulcer or mass lesion.

Conclusions

According to our information, this is the first report of an Iranian patient with cervical esophageal adenocarcinoma. Rapid diagnosis of these patients with clinical features to participate in treatment are important factors that lead to patient survival. Understanding the details of staging and treatment of distal esophageal adenocarcinomas should be considered properly. To improve patients, multidisciplinary management of this rare disease requires the cooperation of specialists in gastroenterology; the ear, nose, and throat (ENT); medical oncology; radiological oncology; and pathology. Due to the too-small number of reported cases, more studies are needed in this regard.

Abbreviations

CECT	Contrast-enhanced computed tomography scan
COPD	Chronic obstructive pulmonary disease
EGM	Ectopic gastric mucosa
EUS	Endoscopic ultrasound
HGM	Heterotopic gastric mucosa
ENT	Ear, nose, and throat
CT	Computed tomography

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Author contributions

ASP, AE, VM, and SS contributed to the treatment design and patient management. MAZ, SS, AHH, and SS conceived the presented idea and wrote the paper. All authors gave consent for publication. All authors read and approved the final manuscript.

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Availability of data and materials

Not applicable.

Declarations

Ethics approval and consent to participate

This research has a code of ethics (No. IR.RUMS.REC.1400.251) from Rafsanjan University of Medical Sciences.

Consent for publication

Written informed consent was obtained from the patient to publish this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare no competing interests.

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