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# RESPONSIBLE EDITORS FOR THIS ISSUE

Production Editor: Ying-Yi Yuan; Production Department Director: Xu Guo; Editorial Office Director: Jin-Lei Wang.

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**NAME OF JOURNAL**

*World Journal of Clinical Cases*

**ISSN**

ISSN 2307-8960 (online)

**LAUNCH DATE**

April 16, 2013

**FREQUENCY**

Thrice Monthly

**EDITORS-IN-CHIEF**

Bao-Gan Peng, Jerzy Tadeusz Chudek, George Kontogeorgos, Maurizio Serati, Ja Hyeon Ku

**EDITORIAL BOARD MEMBERS**

https://www.wjgnet.com/2307-8960/editorialboard.htm

**PUBLICATION DATE**

June 26, 2022

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https://www.wjgnet.com/bpg/gerinfo/239

**ONLINE SUBMISSION**

https://www.ffpublishing.com
Esophagogastric junctional neuroendocrine tumor with adenocarcinoma: A case report

Zhen-Zhen Kong, Lu Zhang

**Specialty type:** Medicine, research and experimental

**Provenance and peer review:** Unsolicited article; Externally peer reviewed.

**Peer-review model:** Single blind

**Abstract**

**BACKGROUND**

At present, cases of esophageal neuroendocrine tumors combined with cardia adenocarcinoma are extremely rare worldwide, and there are no clinical reports. Herein, we describe such a case for clinical reference.

**CASE SUMMARY**

The presence of cardia cancer and esophageal neuroendocrine tumors in a single patient has not yet been reported. The patient in this case underwent prompt endoscopic treatment and additional surgical resection. Pathology revealed the following: The distance between the cardia cancer and the esophageal neuroendocrine tumors was small, approximately 3 mm. Vascular invasion was observed. The esophageal neuroendocrine tumor was determined to be grade G3. According to the treatment guidelines, after the patient received an explanation of their condition, additional surgical procedures were provided in a timely manner. Early detection and early treatment can successfully prolong survival and improve the quality of life of patients.

**CONCLUSION**

Early detection and early treatment can successfully prolong survival and improve the quality of life of such patients.

**Key Words:** Esophageal neuroendocrine tumor; Cardia moderately differentiated adenocarcinoma; Endoscopic treatment; Surgery; Pathology; Case report
Core Tip: Cases of esophageal neuroendocrine tumors combined with moderately differentiated gastric cardia adenocarcinoma are very rare. Pathology is the gold standard for diagnosis. Endoscopy and additional surgical resection proved to be successful in our case. Early detection and early treatment are both of great significance to the life and health of patients. Considering the successful resection of this case, we provide this case report to serve as a clinical reference.

INTRODUCTION
At present, cases of esophageal neuroendocrine tumors (NETs) combined with cardia adenocarcinoma are extremely rare worldwide. The presence of cardia cancer and esophageal NETs (E-NETs) in a single patient has not yet been reported. Herein, we describe such a case for clinical reference.

CASE PRESENTATION
Chief complaints
A 76-year-old man was hospitalized due to the presence of a cardia mass.

History of present illness
Previous gastroscopy showed a 0-IIa-like cardia lesions and chronic atrophic gastritis with erosions. The pathological examination revealed the following: Tubular adenoma with high-grade intraepithelial neoplasia; mild chronic atrophic gastritis of the antrum; intestinal metaplasia; and *Helicobacter pylori* infection (Figure 1).

History of past illness
The patient’s medical history was unremarkable.

Personal and family history
The patient’s personal/family history was unremarkable.

Physical examination
No remarkable characteristics were found during the physical examination.

Laboratory examinations
The laboratory results were all normal.

Imaging examinations
Previous gastroscopy showed a 0-IIa-like lesion of the cardia and chronic atrophic gastritis with erosions. Enhanced computed tomography scan of the full abdomen was performed after hospitalization, which revealed that the local gastric wall of the gastric cardia was slightly thickened, no significantly enlarged lymph node shadow was seen around the cardia, and the rest of the region appeared unchanged (Figures 2-4).

FINAL DIAGNOSIS
The final diagnosis was differentiated cardia adenocarcinoma and E-NET (G3).

TREATMENT
Endoscopic submucosal dissection and surgical resection were performed.
OUTCOME AND FOLLOW-UP

The patient was in good general condition without obvious discomfort (Figures 5 and 6).

DISCUSSION

NENs are a group of highly heterogeneous tumors originating from neuroendocrine cells. They can occur in many parts of the body but are most often found in the digestive system, followed by the lungs.
E-NETs are very rare[1], accounting for only 1.4% of all gastrointestinal pancreatic tumors[2] and 0.15%-2.80% of all esophageal tumors[3]. This is due to the poor development of the neuroendocrine system in this area of the body[2]. The incidence rate varies across countries[4]; these tumors are more commonly found in Asian countries than in Western countries[5]. Studies have found that smoking (present in 49%) and drinking (present in 31%) may be a high-risk factor[6,7]. At present, cases of E-NETs combined with cardia adenocarcinoma are extremely rare worldwide, and there are no clinical reports.

Pathology is the gold standard for the diagnosis of NETs. The proliferation activity of tumor cells can be evaluated by the number of mitotic figures or the Ki-67 index. According to the 2019 WHO classification standards, NETs are divided into three grades: G1, G2, and G3. The classification criteria are as follows: G1 is defined as < 2 mitotic cells/10 high-power fields (HPFs), G2 as 2-20 cells/10 HPFs, and G3 as > 20 cells/10 HPFs. The Ki-67 index is classified as follows: G1, ≤ 2%; G2, 3%-20%; and G3, > 20%[8]. When the Ki-67 index is inconsistent with the mitotic cell classification, it can instead be classified as high or low. DAXX/ATRX and p53/RB mutations can be used to distinguish G3 NETs from neuroendocrine carcinomas (NECs). According to the guidelines, NETs are < 1 cm in size, are grade G1/G2, have a low metastasis rate (< 3%), and do not infiltrate into the muscularis propria (T1 stage). Thus, they are suitable for endoscopic treatment. For tumors or NECs more than 2 cm in diameter, the metastasis rate can reach as high as 60% to 80%, so radical resection is the first choice.
Figure 5 Immunohistochemical staining of esophageal neuroendocrine tumors. A: Syn; B: CD56; C: CgA; D: Ki-67 (25%+).

Figure 6 Representative pathology of excised gastric specimens.

CONCLUSION

A case of cardia adenocarcinoma combined with E-NETs has not yet been reported. In our patient, after timely endoscopic treatment, pathology revealed that the distance between the cardia cancer and the E-NETs was small, approximately 3 mm, vascular invasion was observed, and the E-NET was determined to be grade G3. According to the treatment guidelines, after the patient received an explanation of their condition, additional surgical procedures were provided in a timely manner. Complete resection of the lesion significantly improved the patient’s quality of life.

FOOTNOTES

Author contributions: Kong ZZ was involved in writing the article; Zhang L was involved in the conception of the study; all authors read and approved the final manuscript.

Supported by Zhejiang Provincial Department of Health Clinical Research Application Project, No. 2022KY924; and General Project of Zhejiang Provincial Department of Health, No. 2021KY835.

Informed consent statement: Informed written consent was obtained from the patient for publication of this report and any accompanying images.
Conflict-of-interest statement: The authors declare that they have no conflicts of interest to disclose.

CARE Checklist (2016) statement: The authors have read the CARE Checklist (2016), and the manuscript was prepared and revised according to the CARE Checklist (2016).

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S-Editor: Fan JR
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REFERENCES


