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**Esophagogastric junctional 'neuroendocrine tumor' (NET) with adenocarcinoma: A rare case**

**A rare case**

Lu Zhang, Zhen Zhen Kong, Shuo Zhang

## **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 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 was treated with prompt endoscopic treatment and additional surgical resection. The surgery was a success. Pathology showed negative margins. Early detection and early treatment can successfully prolong survival and improve the quality of life of patients. This case is reported as a clinical reference.

### **CONCLUSION**

Cardia cancer combined with esophageal neuroendocrine tumors in a single patient has not yet been reported. Timely endoscopic treatment was provided, and pathology revealed the following: The distance between the cardia cancer and the esophageal neuroendocrine tumors was small, approximately 3 mm. Vascular invasion was also 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. In our case, complete resection of the lesion significantly improved the patient's quality of life.

**Key Words:** Esophageal neuroendocrine tumor; Cardia moderately differentiated adenocarcinoma; Endoscopic treatment; Surgery; Pathology; Case report.

Zhang L, Kong ZZ, Zhang S. Esophagogastric junctional 'neuroendocrine tumor' (NET) with adenocarcinoma: A rare case.

**Core Tip:** Cases of esophageal neuroendocrine tumors combined with moderately differentiated gastric cardia adenocarcinoma are very rare. Pathology is the gold

standard for diagnosis. With endoscopy and additional surgical resection, treatment was successful. 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**

A 76-year-old man was hospitalized due to the presence of a cardia mass. Previous gastroscopy showed a 0-IIa-like lesion of the cardia and chronic atrophic gastritis with erosions. The pathological indications were: cardia: tubular adenoma with high-grade intraepithelial neoplasia; antrum: mild chronic atrophic gastritis; intestinalization+; and HP-. Enhanced Ct scan of the full abdomen was performed after hospitalization and revealed the following (Figure 1): 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 area appeared unchanged. After excluding the associated contraindications, endoscopic submucosal dissection (ESD) was performed, and a smooth bulge in the lower part of the esophagus was visualized intraoperatively (Figure 2). There was a flat, irregular bulge in the posterior wall of the cardia, the lesion surface was covered with white specks, M-NBI showed glandular duct disorder, and part of the glandular ducts had disappeared (Figure 3). Nodular blood vessels were also observed. A hooked knife was used to mark the lesion range. Submucosal injection was performed along the outer edge of the markings, and the lesion lifted well. The hooked knife was used to strip the lesion along the outside of the marker point, and the wound was cauterized to ensure hemostasis. Postoperative pathological findings revealed the following (Figure 4): The cardia specimens were confirmed to be differentiated cardia adenocarcinoma; the general type was 0-IIa+IIc type, the size was 2.1 cm×0.6 cm; the histological type was tub2>pap; and infiltration into the mucosal muscle layer (MM) was observed. The esophageal specimens were confirmed to be an esophageal neuroendocrine tumor (E-NET, G3); the general type was 0-IIa; the size was 0.4 cm×0.3 cm; infiltration into the mucosal muscle layer (MM) was observed; and vascular invasion (+) was noted. The immunohistochemical staining results of the cardia mass

were as follows: ⑤p53 (85%+), Ki-67 (30%+), Syn (focal +), CgA (individual+), CD56 (-); ⑦p53 (90%+), and Ki-67 (40%+). The immunohistochemical staining results of the esophageal masses are shown in Figure 5, which revealed the following: ⑤p53 (90%+), Ki-67 (25%+), Syn(+), and CD56 (partial+). (Note: ⑤and ⑦refer to the lesion sites of the 5th and 7th pathological slices, respectively.) According to the guidelines, surgical resection was recommended. The majority of laparoscopic gastric resections are followed by esophageal-gastric anastomosis. Thickening of the gastroesophageal junction was seen during the operation, and no obvious metastatic nodules were found in the liver, peritoneum, pelvis, or pancreas. The upper section of the stomach was removed from the abdominal cavity through the opening of the upper abdomen, the closure was vertically cut along the small curved side of the antrum to form a His angle of approximately 3 cm, and the gastric body was cut to the distal end at approximately 4 cm of the cardia next to the base of the stomach so that the residual stomach body was a tubular stomach. The width of the gastric cavity was approximately 4 cm, reaching to approximately 1 cm below the original vertical cutting site, and the specimen was removed. Twelve days after the operation, the patient was discharged from the hospital. Postoperative pathological manifestations are presented in Figure 6. No tumor tissue residue was found, and the examination was negative at both ends of the incision margin and the upper margin. Regarding the lymph nodes the results were as follows: cardia left, 0/7; cardia right, 0/6; and small curved side, 0/15.

## **CASE PRESENTATION**

### ***Chief complaints***

The chief complaint was the presence of a cardiac mass.

### ***History of present illness***

Previous gastroscopy showed a 0-IIa-like cardiac lesion and chronic atrophic gastritis with erosions. The pathological indications confirmed the cardia, revealing the following: tubular adenoma with high-grade intraepithelial neoplasia; antrum: mild chronic atrophic gastritis; intestinalization+; and HP-. Enhanced CT scan of the full abdomen was performed after hospitalization: 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.

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### *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. The pathological indications confirmed the cardia, revealing the following: tubular adenoma with high-grade intraepithelial neoplasia; antrum: mild chronic atrophic gastritis; intestinalization+; and HP-. Enhanced CT scan of the full abdomen was performed after hospitalization: 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.

## **FINAL DIAGNOSIS**

The final diagnosis was differentiated cardia adenocarcinoma and esophageal neuroendocrine tumor (E-NET, G3).

## **TREATMENT**

Endoscopic submucosal dissection and surgical resection was planned.

## OUTCOME AND FOLLOW-UP

The patient was in good general condition without obvious discomfort.

## DISCUSSION

<sup>2</sup> Neuroendocrine neoplasms (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. Esophageal neuroendocrine tumors (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 between 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 high-risk factors (6-7). At present, cases of esophageal neuroendocrine tumors combined with cardia adenocarcinoma are extremely rare worldwide, and there are no clinical reports.

Pathology is the gold standard for the diagnosis of neuroendocrine tumors. 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 system standards, neuroendocrine tumors are divided into 3 levels, G1, G2, and G3. The classification criteria are as follows: G1 is defined as <2 mitotic cells/10 HPFs, G2 is defined as <sup>3</sup> 2-20/10 HPFs, and G3 is defined as >20/10 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, neuroendocrine tumors 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 neuroendocrine carcinomas more than 2 cm in diameter, the metastasis rate can reach as high as 60% to 80%, so radical resection is the first choice.

## **CONCLUSION**

A case of cardia adenocarcinoma combined with esophageal neuroendocrine tumors has not yet been reported. After timely endoscopic treatment, pathology revealed that the distance between the cardia cancer and the esophageal neuroendocrine tumors was small, approximately 3 mm, vascular invasion was observed, and 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. In our case, complete resection of the lesion significantly improved the patient's quality of life.



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