Endoscopic mucosal resection of duodenal bulb adenocarcinoma with neuroendocrine features: An extremely rare case report

Ming-Yao Wen, Yu Wang, Xiao-Yan Meng, Hua-Ping Xie

Ming-Yao Wen, Department of Otolaryngology-Head and Neck Surgery, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, Hubei Province, China

Yu Wang, Department of Pathology, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, Hubei Province, China

Xiao-Yan Meng, Department of Radiology, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, Hubei Province, China

Hua-Ping Xie, Department of Gastroenterology, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, Hubei Province, China

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Correspondence to: Dr. Hua-Ping Xie, Department of

Gastroenterology, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, No. 1095 Jiefangdadao, Wuhan 430030, Hubei Province, China. hpxie@tjh.tjmu.edu.cn
Telephone: +86-27-83663333
Fax: +86-27-83662891

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Abstract

Duodenal adenocarcinoma, especially duodenal bulb with neuroendocrine features (NEF), is extremely rare. Here, we report one such case of duodenal bulb adenocarcinoma with neuroendocrine features. A 63-year-old Han Chinese woman was admitted to our department with the diagnosis of a duodenal bulb polyp and underwent an endoscopic mucosal resection. The pathological findings confirmed it as duodenal bulb adenocarcinoma with NEF. The patient remains curative after one and half a years of follow-up. Duodenal adenocarcinoma with NEF might be a low malignant neuroendocrine tumor rather than a conventional adenocarcinoma. Endoscopic treatment, including endoscopic mucosal resection, might be an ideal option for the adenocarcinomas with NEF.

Key words: Duodenal adenocarcinoma; Neuroendocrine features; Endoscopic mucosal resection; Duodenal bulb; Rarity

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Core tip: We report one extremely rare case of duodenal bulb adenocarcinoma with neuroendocrine features (NEF). Duodenal adenocarcinoma with NEF might be a low malignant neuroendocrine tumor rather than a conventional adenocarcinoma. Endoscopic treatment, including endoscopic mucosal resection, is plausibly an ideal option for adenocarcinomas with NEF.

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INTRODUCTION

Primary malignant tumors of the duodenum represent 0.3% of all gastrointestinal tract tumors.

Duodenal adenocarcinoma accounts for half of all small bowel adenocarcinomas^[1-3]. Histologically, 40% of small bowel cancer are adenocarcinoma and another 40% are neuroendocrine tumors. Recently, a rare case of duodenal adenocarcinoma with neuroendocrine features (NEF) was reported that was neither a conventional neuroendocrine tumor nor a typical duodenal adenocarcinoma^[4]. It remains unclear whether duodenal adenocarcinoma with NEF should be treated as adenocarcinoma or neuroendocrine tumors or both. Here we report one extremely rare case of duodenal bulb adenocarcinoma with NEF which was successfully removed by endoscopic mucosal resection (EMR) and enjoyed a good survival without recurrence or metastasis during one and half a years of follow-up.

CASE REPORT

A 63-year-old Han Chinese woman was admitted to our department with anorexia and epigastric fullness for over two years. She complained of aggravation after eating and remission after belching. There were no significant prior diseases in the medical history. Esophagogastroduodenoscopy from the local hospital revealed a protrusion in the duodenum the size of 0.6 cm × 0.6 cm and she was admitted with an intention of "polypectomy". There were no remarkable findings from the physician examinations. Biochemical examinations were all in normal range, except that the serum chromogranin (CgA) level was 151 ng/mL (reference: 27-94 ng/mL). The multi-row computed tomography (CT) of the small intestine detected a protrusion of duodenum without lymph node involvement. The protrusion was removed by EMR (endoscopic mucosal resection) successfully (Figure 1). The pathological report revealed a diagnosis of

highly differentiated adenocarcinoma with a clean margin, but the immunohistochemical staining for synaptophysin (Syn), CgA and CD56 was positive (Figure 2). The tumor did not meet the diagnostic criteria for neuroendocrine tumors or carcinoids. Thus, we diagnosed duodenal adenocarcinoma with NEF. The patient refused further surgical treatment and was followed-up without any further treatment. There were no signs of recurrence or metastasis on esophagogastroduodenoscopy and color Doppler ultrasound of the epigastrium at the 2nd and 18th mo after EMR.

DISCUSSION

Previously, Kato *et al*^[4] reported one case of duodenal adenocarcinoma with NEF at the descending part in a 67-year-old woman. However, it was accompanied by acromegaly and other types of tumors, namely GH-producing pituitary adenoma and thyroid papillary adenocarcinoma with familial clustering. As per the published literature, our report is the second instance of duodenal adenocarcinoma with NEF, especially without any concomitant endocrine diseases or other tumors. Moreover, it was located at the duodenal bulb, which is supposed to be physiologically, immunologically or otherwise uniquely privileged to virtually escape oncogenic transformation based on the extreme rarity of duodenal bulb adenocarcinoma^[3]. Only five cases of adenocarcinoma have been clearly defined arising from the mucosa of the duodenal bulb on a diligent review of 724 reported cases in the published literature^[3].

Generally, the prognosis of duodenal adenocarcinoma is poor; lymph node metastasis is the only independent predictor of decreased survival. Early surgery is required, although risk of surgical complications and death due to surgery are not ruled out^[5-8]. Surgical resection can be variable and depends mainly on the location of the tumor. Tumors arising in the first, second or third portion of the duodenum typically require pancreaticoduodenectomy, whereas tumors occurring in the fourth portion may be more amenable to segmental duodenal resection^[8]. However, the neuroendocrine component may have a considerable impact on duodenal adenocarcinoma^[9]. One case of duodenal adenocarcinoma with NEF combined with liver metastasis survived for more than 24 mo without progression of the primary and metastatic lesions. It is plausible that duodenal adenocarcinoma with NEF is predominantly a neuroendocrine tumor of low malignancy potential, rather than a common adenocarcinoma^[4]. Malignant NET, such as type 3 gastric NETs which are considered to be aggressive and often with metastasis, can also be treated successfully by endoscopic treatment, including EMR^[10]. Endoscopic resection appears to be a safe and effective treatment for duodenal carcinoid tumors measuring ≤ 10 mm in diameter and confined to the submucosal layer^[11].

Figure 1 Computed tomography, endoscopic image and endoscopic mucosal resection of duodenal tumor with neuroendocrine features. A: Computed tomography (CT) image of a duodenal tumor (arrow); B: Three dimensional CT image reconstruction of the duodenal tumor (arrow); C: Endoscopic image of the duodenal tumor; D: Positive lifting sign after injection of physiological saline with 1:10000 norepinephrine; E: Endoscopic mucosal resection after snare; F: Clean border of the wound after endoscopic mucosal resection (EMR); G: Scar formation on EMR position at gastroscopic review after two months.

Figure 2 Histopathology of duodenal adenocarcinoma with neuroendocrine features. A: Hematoxylin and eosin (HE) staining (original magnification $\times 40$); B: HE staining (original magnification $\times 100$); C: HE staining (original magnification $\times 200$); D: Immunohistochemical staining for synaptophysin (original magnification $\times 100$); E: Immunohistochemical staining for chromogranin (original magnification $\times 100$); F: Immunohistochemical staining for CD56 (original magnification $\times 100$).

Here, we removed the tumor with the lift and cut EMR method, in compliance with neuroendocrine tumors. This might be an optimal method for the treatment of duodenal adenocarcinomas with neuroendocrine differentiation (NED). The curative therapy in our case

indicates that the treatment for the NEF component in duodenal adenocarcinoma is adequate.

Incidentally, the presence of NED remains a poor prognostic factor in other adenocarcinomas. In gastric adenocarcinomas, even a minor proportion of 10% of

NED is associated with significantly poorer survival^[12]. In a meta-analysis of 1587 cases of colorectal adenocarcinoma, patients with NED had a poor 5 year survival rate compared to those without NED^[13]. In prostate cancer, NED is also a poorer prognostic factor. The levels of serum CgA are increased in patients with prostate cancer compared with benign conditions. The early detection of high serum levels of CgA could be an indicator for a change to a more aggressive therapy^[14]. Although the exact function of the NEF component is still unknown, it is evident that the NEF component in duodenal and other adenocarcinomas apparently has different functions.

Here, we present a rare case of duodenal bulb adenocarcinoma with NED, which was successfully removed by EMR and has remained curative during the follow-up time. Duodenal adenocarcinoma with NEF might be treated as a neuroendocrine tumor and the role of NED in duodenal adenocarcinoma still needs further study.

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COMMENTS

Case characteristics

A 63-year-old Han Chinese woman complained of anorexia and epigastric fullness for over two years.

Clinical diagnosis

One lesion of duodenal bulb was detected by endoscopy.

Differential diagnosis

The differential diagnosis of lesion of the duodenal bulb should be polyps and other benign diseases.

Laboratory diagnosis

Biochemical examinations were all in the normal range, except that the serum chromogranin (CgA) level was 151 ng/mL (reference: 27-94 ng/mL).

Imaging diagnosis

Esophagogastroduodenoscopy from the local hospital revealed a protrusion in the duodenum the size of 0.6 cm × 0.6 cm, which was also detected by the multi-row computed tomography of the small intestine, without lymph node involvement.

Pathological diagnosis

The pathological report revealed a diagnosis of highly differentiated adenocarcinoma with positive immunohistochemical staining for synaptophysin (Syn), CgA and CD56, which did not meet the diagnostic criteria for neuroendocrine tumors or carcinoids and duodenal adenocarcinoma with neuroendocrine features (NEF) was diagnosed.

Treatment

The duodenal protrusion was successfully removed by endoscopic mucosal resection (EMR).

Related reports

One case of duodenal adenocarcinoma with NEF with liver metastasis reported in a 67-year-old woman was accompanied by acromegaly and other types of tumors, namely GH-producing pituitary adenoma and thyroid papillary adenocarcinoma with familial clustering.

Term explanation

EMR is an endoscopic technique used to remove cancerous or other abnormal lesions found in the gastrointestinal tract.

Experiences and lessons

Lesions at the duodenal bulb should not be underestimated due to the risk of malignant diseases.

Peer-review

Duodenal adenocarcinoma with NEF might be a low malignant neuroendocrine tumor rather than a conventional adenocarcinoma. Endoscopic treatment, including EMR, might be an ideal option for adenocarcinomas with NEF. However, only one case cannot provide strong support for the idea.

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