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*Case Report*

# Duodenal gangliocytic paraganglioma with lymph node metastases: a case report and review of literature

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**Abstract:** Duodenal gangliocytic paraganglioma is an extremely rare tumor. The part of the rare place is esophagus, jejunum, pylorus, pancreas and upper mediastinum. Though considered benign the disease can spread to regional lymphatics. Gangliocytic paraganglioma is a rare tumor that is located in the duodenum in 90% of cases and has been regarded as benign in general with a favorable prognosis. Local excision is used to treat the disease, and radical surgery and lymph node dissection can be avoided if gangliocytic paraganglioma is confirmed. Now, we report a 65-year-old man with lymph gland metastases of duodenal gangliocytic paraganglioma. Local resection was performed. Gangliocytic paragangliomas (GPs) are rare tumors of the duodenum and demonstrates low malignant potential.

**Keywords:** Duodenal gangliocytic paraganglioma, lymph gland metastases

## Introduction

Gangliocytic paragangliomas (GP) are infrequent tumors of the gastrointestinal tract usually found in the second portion of the duodenum. Gastrointestinal bleeding is the most common clinical presentation, followed by abdominal pain and anemia. These submucosal tumors have been generally considered as benign and non-functional. GP is characterized by its triphasic cellular differentiation, consisting of epithelioid neuroendocrine cells, spindle-shaped cells, and ganglion-like cells [1]. This tumor has been regarded as benign in general, but a few cases with lymph node metastasis have been reported which required extensive surgical removal [**Table 1**]. GP was first described by Dahl in 1957 [2]. The most common clinical presentation includes gastrointestinal bleeding and abdominal pain. Rarely, gangliocytic paragangliomas are manifest by pyloric or duodenal obstruction. Surprisingly, almost all patients with GP, including those having lymph node or distant metastasis, gain a good outcome without recurrence. Herein, we report a case of surgically resected duodenal gangliocytic paraganglioma with lymph gland metastases and review the pertinent literature.

## Case report

A 65-year-old male patient presented with complaints of hemafecia for 3 days. During this period, the patient had dizziness and hemafecia occurred 5 times. He showed no epigastric soreness, abdominal pain, or weight loss and experienced vomiting at the time of hospitalization. These symptoms show that he should receive treatment for selective gastroduodenal artery embolization in invasive technology department. As a result, the patient was transferred to our hospital for examination and treatment. No specific features arose from his family or social history. Physical examination results were normal. His hemoglobin was 88 g/L, and renal and liver function, as determined by blood tests, was also normal. No lesions were found in the esophagus or stomach by esophagogastroduodenoscopy. However, an exophytic tumor with a bleeding surface ulcer was observed in the above of duodenal papilla in the second portion of the duodenum. The mass was demonstrated 2 cm. The lesion did not obstruct the ampullary orifice. The bleeding from the ulcer was controlled by endoscope. Digital subtraction angiography (DSA) revealed a vascular malformation group in the second portion of the

**Table 1.** Gangliocytic paraganglioma showing lymph node metastasis

| Reference | Year | Age | Sex | Chief clinical presentation | Site | Size (cm) | Operation | Outcome (months) |
|---|---|---|---|---|---|---|---|---|
| Inai et al [12] | 1989 | 17 | Male | Hematoemesis | Duodenum | 2.0 | WP | NED32 |
| Hashimoto et al [13] | 1992 | 47 | Male | Incidental findings | Duodenum | 6.5 | WP | NED14 |
| Tomic et al [14] | 1996 | 74 | Female | Abdominal pain, vomiting, weight loss | Pancreas | 4.0 | WP | NED19 |
| Henry et al [15] | 2003 | 50 | Male | Abdominal pain | Pancreas | 2.5 | WP | NA |
| Sundararajan et al [11] | 2003 | 67 | Female | Incidental findings | Second part of duodenum | 5.0 | WP | NED9 |
| Bucher et al [16] | 2004 | 31 | Female | Anemia, subclinical jaundice | Papilla of Vater | 3.0 | WP | NED44 |
| Wong et al [17] | 2005 | 49 | Female | Melena | Duodenum | 1.4 | WP+RT | NED12 |
| Witkiewicz et al [18] | 2007 | 38 | Female | Abdominal pain | Papilla of Vater | 1.5 | LR+WP | NA |
| Mann et al [19] | 2009 | 17 | Female | Abdominal pain, vomiting, weight loss | Duodenum | NA | WP | NED12 |
| Okubo et al [1] | 2010 | 61 | Male | Abdominal pain, Melena | Papilla of Vater | 3.0 | WP | NED16 |
| Uchida et al [20] | 2010 | 67 | Female | Stiff shoulders and anemia | Second part of duodenum | NA | WP | NA |
| Ogata et al [21] | 2011 | 16 | Male | Exertional dyspnea | Ampulla of Vater | 2.5 | WP | NED36 |
| Rowsell et al [22] | 2011 | 52 | Female | NA | Duodenum | 1.0 | LR | NED27 |
| Barret et al [23] | 2012 | 51 | Female | Melena | Duodenum papilla | 3.5 | WP | NED96 |
| Shi et al [24] | 2014 | 47 | Male | Abdominal pain, weight loss | Papilla of Vater | 4.0 | WP | NED24 |
| Present case | 2015 | 65 | Male | Melena | Second part of duodenum | 3.0 | LR | NED2 |

LR: Local resection; WP: Whipple procedure; NA: Not available; NED: No evidence of disease. RT: Radiotherapy.

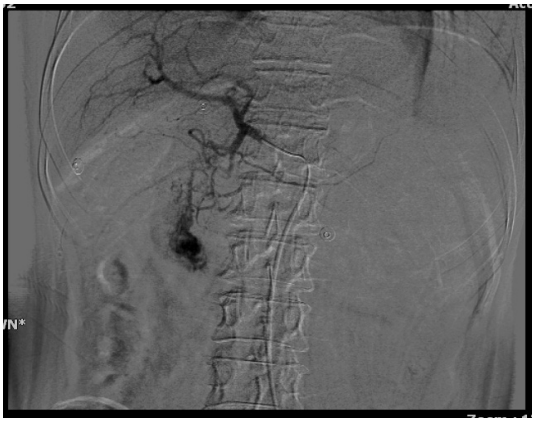

**Figure 1.** DSA reveal a vascular malformation group in the second portion of the duodenum.

duodenum (**Figure 1**). The macroscopic aspect showed a 3×2×2 cm mass (**Figure 2**). Histopathology revealed ganglion-like cells (**Figure 3**), spindle cells (**Figure 4**), and epithelioid cells (**Figure 5**) in the submucosal layer of the duodenum. Immunohistochemical staining for the CgA, CD56, Syn revealed strong positive reactions.

## Discussion

Gangliocytic paraganglioma is a rare tumor that is located in the Second part of duodenum. The part of the rare place is esophagus, jejunum, pylorus, pancreas and upper mediastinum. Several theories about the origin of gangliocytic paragangliomas have been proposed: origin from an embryonic celiac ganglion; the tumor deriving from pluripotent stem cells located at the base of intestinal glands; the lesion deriving from hamartomatous proliferation of ectodermally derived epithelial cells originating from the ventral primordium of the pancreas, and neuroectodermally derived ganglion and schwannian spindle cell; and the lesion arising from ectopic pancreatic tissue [3, 4]. But the histogenesis of these tumors is uncertain.

Histologically, the tumor shows triphasic cellular differentiation with the presence of epithelioid neuroendocrine cells, spindle-shaped cells with Schwannian features and ganglion cells. The proportion of these three cell types is variable. The epithelioid and ganglion-like cells expressed synaptophysin, chromogranin, somatostatin, pancreatic peptide, and serotonin. Spindle cells showed strong expression of S-100 protein.

The mean age at presentation is 52 years (range 15-84). The incidence of GPs is slightly higher in males than in females (1.8:1); the usual clinical manifestation is gastrointestinal bleeding owing to overlying mucosa ulceration, followed by abdominal pain and anemia, and the biliary obstruction or duodenal obstruction with low frequency. Exceptional cases of gangliocytic paragangliomas originating in the third or fourth portion of the duodenum have been reported [5-9].

The diagnostic by endoscopic biopsy is usually negative because the tumors are submucosal. Ultrasound shows a solid isoechogenic mass.

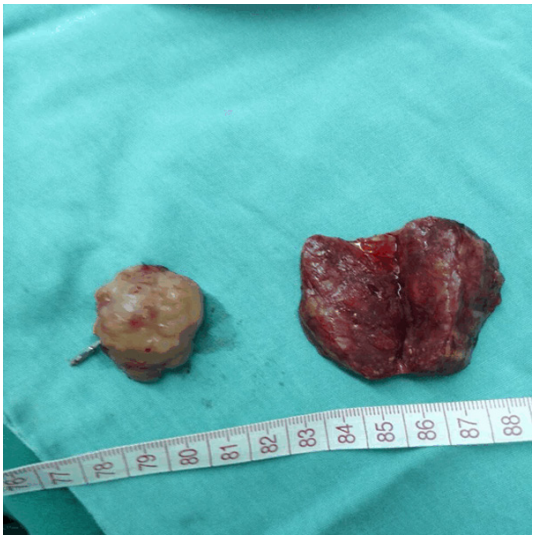

**Figure 2.** The macroscopic aspect showed a 3×2×2 cm mass.

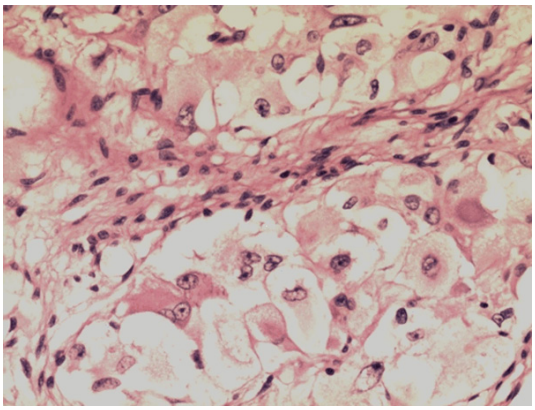

**Figure 3.** Photomicrograph showing the ganglionic cells. (Hematoxylin and Eosin original magnification ×400).

Contrast-enhanced CT scan will usually show a mass with soft tissue attenuation and homogeneous enhancement. On magnetic resonance imaging, the tumor is solid and homogenous in appearance. Endoscopic ultrasound features of gangliocytic paragangliomas are isoechoic or hypoechoic. Although PG is difficult to be diagnosed precisely, sufficiently using endoscopic ultrasound is an effective preoperative diagnosis for the accurate diagnosis of PG. The lesion is submucosal, non-encapsulated and well-circumscribed [10]. Differential diagnosis includes pancreatic head or duodenal cancer, choledochal cyst, duodenal tumors such as lipoma, hamartoma, hemangioma, lymphoma, leiomyosarcoma and inflammatory fibroid polyps.

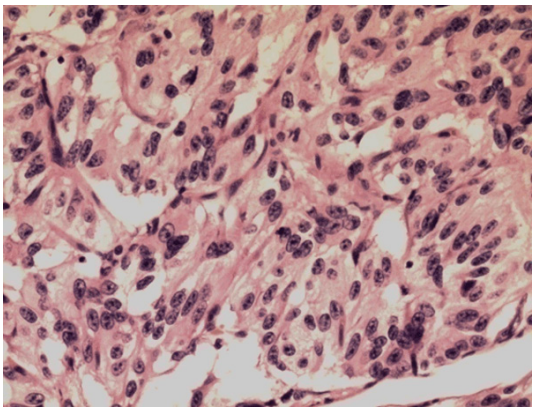

**Figure 4.** Photomicrograph showing the epithelioid cells are arranged in nests of ribbon-like structures. (Hematoxylin and Eosin original magnification ×400).

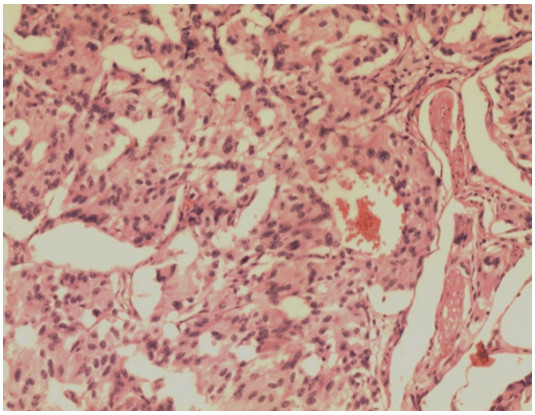

**Figure 5.** Photomicrograph showing the spindle cells. (Hematoxylin and Eosin original magnification ×400).

The majority of the reported duodenal gangliocytic paragangliomas is of benign and nonfunctional nature. Local surgical excision of the lesion is preferred. Metastasis and the recurrence of gangliocytic paraganglioma are rare. At present, no chemotherapy is advised. Endoscopic resection of duodenal gangliocytic paraganglioma appears to be safe and effective [11]. Tumor location, depth of invasion, risks of procedure-related complications and possible lymph node affection should be considered. Complete resection of the tumor combined with adjuvant chemo- or radiotherapy is suggested as a treatment for GP patients with lymph node and/or distant metastasis to avoid the potentially rapid progression of the disease.

In conclusion, we report a rare case of duodenal GP with regional lymph node occurring in an adult patient.

## Disclosure of conflict of interest

None.

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