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## Duodenal gangliocytic paraganglioma: A case report and literature review

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## ABSTRACT

**INTRODUCTION:** Gangliocytic paraganglioma is a rare tumor that is most commonly located in the duodenum. At presentation, it may be confused with a gastrointestinal stromal tumor (GIST), but distinguishing between these tumors is critical because the natural history and treatment of these two tumors differs markedly. Duodenal gangliocytic paraganglioma typically exhibits benign behavior with occasional regional lymph node metastasis and no reports of tumor associated deaths. Recurrence after resection is rare.

**PRESENTATION OF CASE:** A 50 year-old male presented with melena and hemoglobin concentration of 4.6 g/dl. Esophagogastroduodenoscopy demonstrated a submucosal mass in the third portion of the duodenum with no active bleeding. CT scan identified no regional lymphadenopathy or distant metastasis. The tumor was resected through a longitudinal duodenotomy with negative margins.

**DISCUSSION:** Endoscopic resection of duodenal gangliocytic paraganglioma appears to be safe and effective when tumor may be removed in its entirety by this method. If the tumor is not suspended by a stalk or there is suspicion for regional lymph node disease then surgical management is preferred. Radiation oncologists at high volume centers have endorsed utilization of adjuvant radiotherapy to the postsurgical bed in cases involving lymph node metastasis. Utilization of chemotherapy for management of this disease has not been reported.

**CONCLUSION:** Localized duodenal gangliocytic paragangliomas are best managed by resection with negative margins. In cases in which the tumor is resected with negative margins, it appears to be safe to embark on a course of surveillance and forego adjuvant therapy.

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## 1. Introduction

Gangliocytic paraganglioma is a rare tumor that is located in the duodenum in 90% of cases with a particular predilection to the region of the ampulla of Vater [1]. At presentation, it may be confused with a gastrointestinal stromal tumor (GIST), but distinguishing these tumors is critical because the natural history, and therefore, treatment of these two tumors differs markedly. About 200 cases have been reported since the first description of this tumor in 1957 [2]. The most common clinical presentation includes melena and abdominal pain, each of which occurs in about half of all patients [1]. This tumor typically exhibits benign behavior with regional lymph node metastasis occurring in only 5–7% of

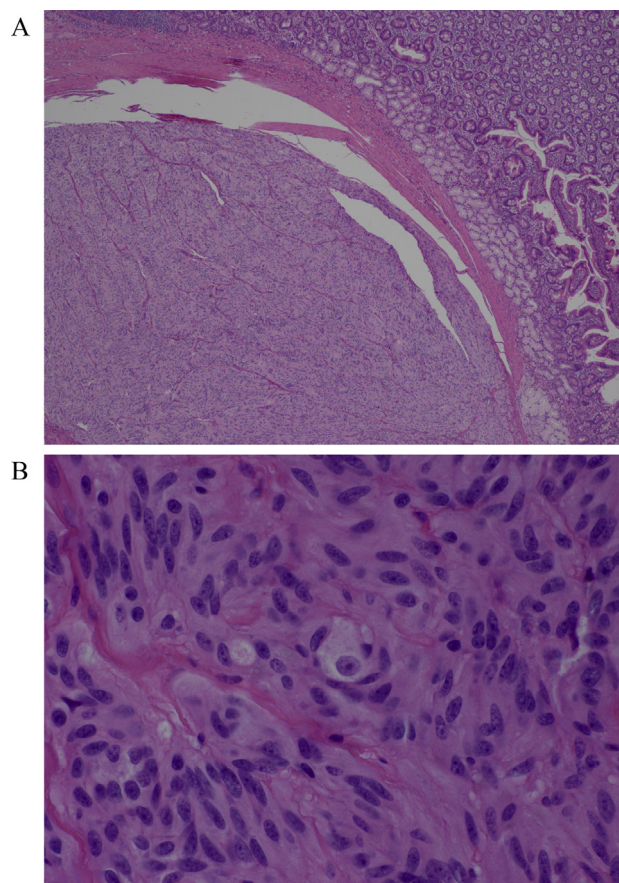
cases with no tumor associated deaths reported [1,3–8]. Recurrence after resection is rare, with only a single reported recurrence many years after initial resection [9]. Herein, we report a case of surgically resected duodenal gangliocytic paraganglioma and review the pertinent literature.

## 2. Exemplary patient

A 50 year-old male presented to an outside hospital with melena and hemoglobin concentration of 4.6 g/dl. He received a total of four units of packed red blood cells and underwent esophagogastroduodenoscopy, which demonstrated a submucosal mass in the third portion of the duodenum with no active bleeding. Computed tomography visualized a 2.5 cm duodenal mass with no evidence of regional lymphadenopathy or distant metastasis. He was transferred to our facility for consideration of a pancreatoduodenectomy due to the close proximity of the mass to the superior mesenteric artery and vein.

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**Fig. 1.** (A) Hematoxylin and eosin-stained section shows a circumscribed submucosal mass with spindle cells arranged in nests and trabeculae. (B) Rare ganglion cells were identified, scattered within the spindle cell compartment.

Exploratory laparotomy confirmed the absence of distant disease and regional lymphadenopathy. The mass was mobile within the third portion of the duodenum, and it appeared amenable to local resection. Through a longitudinal duodenotomy, a 2.5 cm mobile tumor was delivered and noted to be suspended on a wide mucosal stalk. A linear cutting stapler was used to transect the base of the stalk, and the tumor was removed with a clear margin. Initial frozen section pathological assessment of the mass was consistent with a GIST. The duodenotomy was closed transversely in two layers.

Postoperatively, the patient did well with no complications and was discharged on postoperative day five. He returned to clinic tolerating his diet without evidence of ongoing bleeding.

### 3. Pathology review

Histologic evaluation revealed a well-circumscribed, non-encapsulated tumor based in the submucosa and muscularis propria of the duodenum (Fig. 1A). The tumor was predominantly composed of spindled cells arranged in a nested/trabecular pattern. Only rare scattered ganglion-like cells were identified (Fig. 1B). Areas of the tumor show palisaded arrangements of spindled cells resembling Verocay bodies, classically found in schwannomas, yet also described in some GISTs. Epithelioid cells arranged in nests (so called “endocrine” pattern) were not identified. A definitive epithelioid-endocrine component was not apparent. The tumor was immunoreactive for neuron-specific enolase, chromogranin and S-100 highlighted sustentacular cells. The tumor displayed a relatively low mitotic rate of 2/10HPF, confirmed by immunostain

for phosphohistone-H3 (PHH3). The overlying small bowel mucosa demonstrated ulceration and reactive epithelial changes including foveolar metaplasia. The morphologic findings, along with the immunohistochemical profile, support a diagnosis of gangliocytic paraganglioma.

Due to its spindle-cell predominance, our main differential diagnosis included nerve sheath tumors (schwannoma) and GIST. The lack of diffuse positivity for S-100 and the presence of scattered ganglion cells excluded the possibility of a schwannoma, a very rare tumor in this location. Negative immunohistochemical stains for DOG-1, CD117 and CD34 as well as the sustentacular pattern of S-100 (Table 1) protein argued against a GIST.

### 4. Discussion

Duodenal gangliocytic paraganglioma consists of three distinct cellular elements: spindle cells, epithelial cells originating from the ventral primordium of the pancreas, and ganglion cells [3,4,10]. The proportion of these three cell types is variable. In spindle-cell predominant tumors, the differential diagnosis includes schwannomas and GIST. If there is a predominance of ganglion cells or epithelioid cells, the differential diagnosis includes ganglioneuromas for the former and well-differentiated neuroendocrine tumors or carcinomas for the latter. These tumors range in size from 0.5 cm to 10 cm and average 2.5 cm in largest diameter [1,4,5]. The neoplasms most often affect individuals ranging from the second to the ninth decade of life and exhibit a slight male predominance with a ratio of 1.5:1 [1]. Gangliocytic paraganglioma appears on ultrasound as an isoechoic mass and on computed tomography as a homogeneously iso-attenuated mass [6]. Due to the submucosal location of this tumor, preoperative pathologic diagnosis is difficult based on endoscopic biopsy alone [5]. Tumor behavior is usually benign with regional lymph node metastasis occurring in about 5–7% of cases [1,3–8,11,12] with rare invasion of nearby structures [13]. Younger patients appear to be at increased risk for lymph node metastasis [1]. Recurrence is rare, and has occurred as late as 11 years after initial resection [9]. Although gangliocytic paraganglioma from primary sites other than the duodenum have demonstrated the potential for distant metastasis [14], there have been no reported cases of distant metastasis from gangliocytic paraganglioma of duodenal origin [1–5,7–11]. Although there are isolated cases of gangliocytic paragangliomas in association with neurofibromatosis, the vast majority of these cases are sporadic [15].

Endoscopic resection of duodenal gangliocytic paraganglioma appears to be safe and effective in cases in which the tumor may be removed in its entirety by endoscopic methods [1,4,5]. One case has been reported in which a positive margin following endoscopic resection was managed with pylorus preserving pancreaticoduodenectomy [11]. Pathologic review of the pancreaticoduodenectomy specimen identified metastasis in 2 of 7 lymph nodes, indicating the aggressive nature of this particular tumor. Surprisingly, this tumor was only 1.5 cm in its greatest dimension. Eight cases of surgically resected duodenal gangliocytic paraganglioma are listed in Table 2. Of note, the patients with lymph node metastases appeared to have epithelial components in the tumor. Whether this is coincidence or indicative of a causal relationship is unknown. In our case, surgical resection through a duodenotomy was performed without complication and achieved a negative margin.

Radiation oncologists at high volume centers have endorsed utilization of adjuvant radiotherapy to the postsurgical bed in cases involving lymph node metastasis [3]. There has not been a documented recurrence of duodenal gangliocytic paraganglioma in patients who harbor no residual disease after initial resection, and

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