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## Duodenal gangliocytic paraganglioma, successfully treated by local surgical excision—a case report



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### ABSTRACT

**INTRODUCTION:** Duodenal gangliocytic paragangliomas are rare neoplasms often arising in proximity to the major duodenal papilla of Vater. These neoplasms are considered to have a benign behavior with lymph node metastases being a rare phenomenon and distant metastatic disease even more so. Resection of the tumor is the only definitive therapy.

**PRESENTATION OF CASE:** A 67 year old male presented to a referring hospital with symptoms of fatigue and malaise. Evaluation with CT imaging revealed a 3.1 cm intraluminal mass situated grossly at the junction of the third with the fourth portion of the duodenum. The tumor was found to be situated near the ampulla of Vater and was excised through a longitudinal duodenotomy followed by myotomy of the sphincter of Oddi.

**DISCUSSION:** Complete resection of duodenal gangliocytic paragangliomas by surgical or endoscopic means is the only potential cure. Endoscopic removal is the first option and is both safe and adequate. However, localized excision may be utilized instead in those cases in which endoscopic removal is not possible or cannot achieve negative margins. Recurrent disease after complete resection is unlikely.

**CONCLUSION:** Cases of duodenal gangliocytic paragangliomas are best managed with endoscopic resection. However, local surgical excision remains as a second-choice procedure. Adjuvant chemotherapy and radiotherapy are unnecessary after complete excision.

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## 1. Introduction

Duodenal gangliocytic paragangliomas (DGPs) are rare neoplasms often arising in proximity to the major duodenal papilla of Vater [1]. These tumors are often mistaken for other gastrointestinal tract neoplasms, such as gastrointestinal stromal tumors (GISTs), and accurate diagnosis is unlikely without a histologic examination demonstrating the three key characteristic components of the tumor: epithelioid, spindle-shaped, and ganglion-like cells [1]. These neoplasms are considered to have a benign behavior with lymph node metastases being a rare phenomenon and distant metastatic disease even more so [2–4]. Resection of the tumor is the only definitive therapy. This may be achieved through either endoscopic or surgical resection. The aim of this case report is to present

a challenging case of a DGP, situated near the ampulla, treated with local surgical excision instead of a radical procedure. Our report is in accordance with the SCARE criteria [5].

## 2. Case report

A 67 year old male presented to a referring hospital with symptoms of fatigue and malaise, which were attributed to anemia with hemoglobin value of 6.1 g/dL, and hematocrit of 19.4%. Further hematological investigation including white blood cells, liver function tests, INR, blood urea, blood creatinine, and electrolytes were within normal ranges. Tumor markers (CEA, CA 19.9, CA 125, CA 15.3, and  $\alpha$ -fetoprotein) were normal as well. Initial work-up consisted of a computed tomography (CT)-scan imaging which revealed an intraluminal mass measuring 3 cm in diameter, and situated grossly at the junction of the third to the fourth portion of the duodenum (Fig. 1). No regional lymphadenopathy or evidence of metastatic lesions were noted. A colonoscopy was normal and an upper GI endoscopy did not demonstrate any lesion aside

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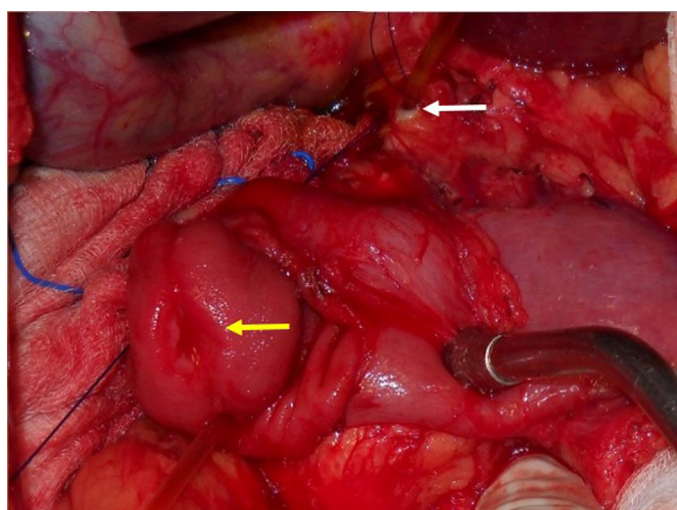
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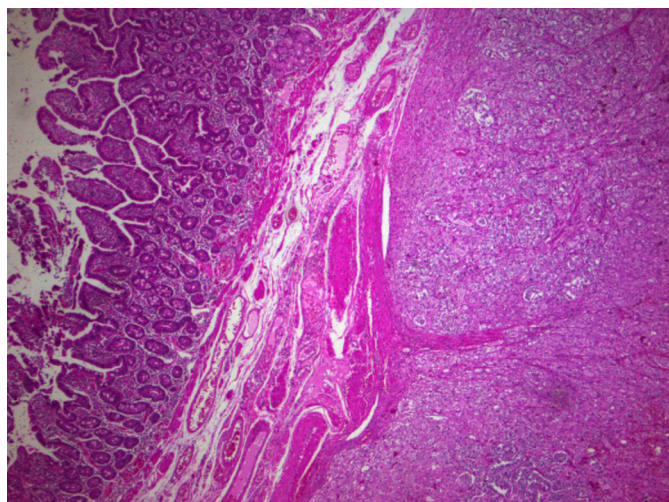
**Fig. 1.** Axial CT-scan of the abdomen after IV contrast medium administration shows an oval shape (white arrow), well circumscribed mass in the third part of the duodenum, with maximum diameter of 3 cm.



**Fig. 3.** Circumscribed tumor with yellow to white cut surface.



**Fig. 2.** After catheterization of the common bile duct (white arrow), a longitudinal duodenal incision was performed to visualize the tumor (yellow arrow).



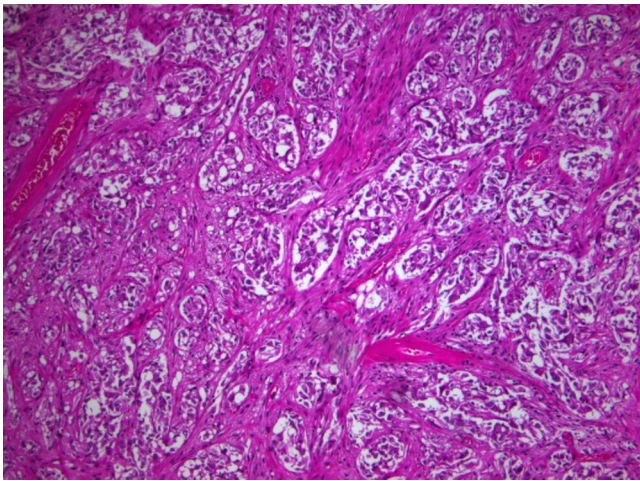
**Fig. 4.** A circumscribed, but not encapsulated, tumor is noticed on scanning magnification (Haematoxylin and Eosin stain: 4×).

from a diffuse gastritis. Capsule endoscopy employed thereafter demonstrated a non-occluding mass lesion in close proximity to the duodenal papilla. The patient was referred to our hospital, a tertiary care center, for further surgical management of this lesion which was initially considered to be a gastrointestinal stromal tumor (GIST). The patient underwent an exploratory laparotomy, revealing no metastatic burden and local excision of the mass was contemplated. After sufficient mobilization of the second and third portions of the duodenum with a Kocher maneuver, the common bile duct was catheterized through a small incision, and the tip of the catheter advanced into the lumen of the duodenum through the ampulla of Vater. Subsequently, a longitudinal duodenal incision was performed to allow direct visualization of the ampulla and the adjacent tumor (Fig. 2). The mass was excised together with the submucosa of the duodenum and the ampulla, with the catheter allowing proper identification and preservation of the major duodenal papilla. Myotomy of the sphincter of Oddi was performed to prevent postoperative obstruction from scar tissue. The third portion of the duodenum was divided with a linear cutter and reconstruction followed, with a duodenojejunal anastomosis and a jejunojejunal anastomosis performed at 40 cm of length. The post-

operative course was uneventful, with improvement of hemoglobin values and the patient was discharged on day 13 postoperatively. A CT-scan (not mentioned here) at 8 months postoperatively was normal and at a 5-year follow-up, the patient is alive and free of disease.

### 2.1. Pathological findings

Macroscopically, after formalin fixation, a fairly circumscribed tumor with a yellow to white cut surface was seen (Fig. 3). Excision appeared, just complete in the sections examined. The lesion was, focally, extending within less than 0.1 mm from the circumferential margin. Pathology review of the specimen revealed a fairly circumscribed tumor located in the duodenal submucosa and muscularis propria (Fig. 4), composed mainly of epithelioid cells with a variable admixture of spindle Schwann like cells and ganglion like cells (Fig. 5). The epithelioid element of endocrine origin was diffusely positive for synaptophysin (Fig. 6a) and immunoreactive, albeit focally, for CKAE1/AE3 (Fig. 6b). In contrast, spindle cells were positive for S-100 (Fig. 6c) whereas both spindle and ganglions like cells were positive for neurofilament staining (Fig. 6d). In addition synaptophysin and chromogranin staining of ganglion like



**Fig. 5.** Three distinct types of cells are evident: epithelioid, spindle Schwann like and ganglion like cells.

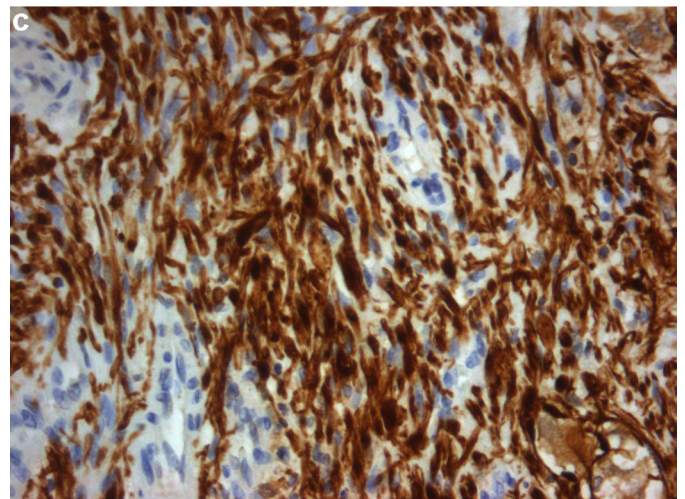
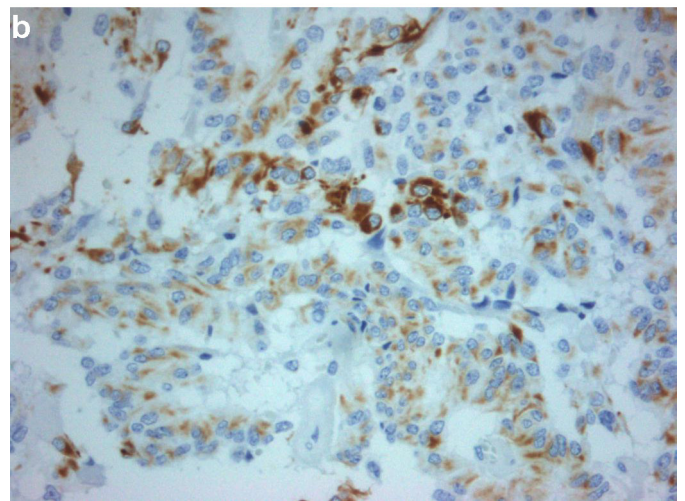
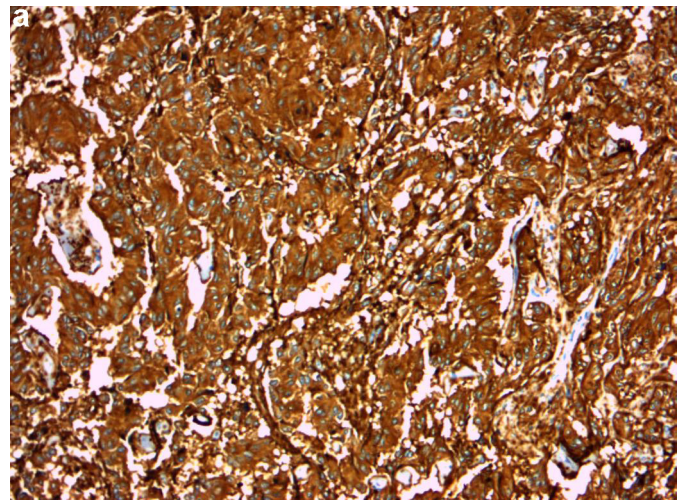
cells was evident. The immunohistochemical analysis together with the morphologic findings of the specimens therefore supported the diagnosis of a duodenal GP.

### 3. Discussion

GPs, in the majority of cases, arise in the duodenum and show a slight male predominance (reported 1.5:1 male-female ratio). They affect individuals of age ranging from 15 to 85 years of age, with GI bleeding and anemia being the most common presenting symptom [2], with abdominal pain and obstructive jaundice having been reported as well [6]. Abdominal CT imaging and endoscopy are unlikely to yield an accurate diagnosis and therefore, mistaking this tumor for a GIST or a neuroendocrine neoplasm is common. The main focus of treatment is removal of the lesion either by endoscopy or surgery. Although GPs are considered to be benign, 23 cases of lymph node metastasis, 3 cases of distant metastases and one death have been reported [2–4,6]. This suggests a metastatic potential for these tumors. Factors associated with lymph node metastases have been reported to be young age and possibly the vertical extent of the tumor in the submucosa [2].

Complete excision of the mass by surgical or endoscopic means is the only cure. Endoscopic removal is the first option in GPs and is both safe and adequate [7–9]. In the case of a periampullary or a large lesion, however, endoscopic resection may be challenging or impossible and therefore surgical resection of the lesion remains as the only therapeutic option. A pancreaticoduodenectomy may be employed for removal of the primary tumor as well as lymph nodes harboring possible metastases. However, taking into account the fact that lymph node metastases are a rare phenomenon in this type of disease, a less radical approach towards the removal of the tumor may be selected, in those cases in which lymphadenopathy is absent in preoperative imaging studies [8,10,11]. The surgical approach we employed consisted of complete removal of the tumor situated in proximity to the major duodenal papilla, followed by a sphincteroplasty and catheterization of the ampulla. This procedure avoids the mortality and morbidity of a Whipple procedure and its postoperative complications such as pancreatic fistula and bile leak, while ensuring adequate resection margins. A sphincteroplasty is mandatory in this scenario to avoid postoperative biliary obstruction from scar tissue.

Recurrent disease after resection is unlikely, with only one such case reported in literature in a patient who underwent a R1 resection [12]. Adjuvant radiotherapy, although having been employed in two cases [4,13], is unnecessary, since disease recurrence in the



**Fig. 6.** a) Staining for synaptophysin in epithelioid (endocrine) cells. b) Focal positivity for CKAE1/AE3 in epithelioid cells. c) S-100 highlights the spindle (Schwann like) component. d) NF positivity in both spindle and ganglion like cells.

setting of a margin negative resection has not been documented in literature. Postoperative chemotherapy for this particular disease has not been employed. Accurate long term outcomes are limited due to the rarity of GPs. Follow-up by a multidisciplinary team is the recommended course of action after a complete excision.

#### 4. Conclusion

DGPs are rare entities that are best managed by a margin free resection. In those cases in which preoperative imaging does not suggest regional disease, endoscopic resection is the treatment of choice, with local surgical excision through a duodenotomy reserved for those in which endoscopic R0 resection is impossible. Adjuvant chemotherapy or radiotherapy are unnecessary after complete excision and routine surveillance appears to be a safe option.

#### Conflict of interest

There is no conflict of interest to be declared.

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#### Consent

Consent has been taken.

#### Author contribution

Papakonstantinou D: He wrote the paper.

Zavras N: He reviewed the paper

Damaskou V: Pathologist performed the histological examination.

Contopoulou Chr: Radiologist performed the evaluation of the CT-scan.

Mahairas A: Surgeon performed the operation and follow-up.

#### References

- [1] Y. Okubo, T. Nemoto, M. Wakayama, N. Tochigi, M. Shinozaki, T. Ishiwatari, et al., Gangliocytic paraganglioma: a multi-institutional retrospective study in Japan, *BMC Cancer* 15 (2015) 269.
- [2] Y. Okubo, M. Wakayama, T. Nemoto, K. Kitahara, H. Nakayama, K. Shibuya, et al., Literature survey on epidemiology and pathology of gangliocytic paraganglioma, *BMC Cancer* 11 (2011) 187.
- [3] C. Henry, H. Ghalel-Méchaoui, N. Bottero, T. Pradier, H. Moindrot, Gangliocytic paraganglioma of the pancreas with bone metastasis, *Ann. Chir.* 128 (2003) 336–338.
- [4] Bin Li, Yang Li, Xiao-Ying Tian, Bo-Ning Luo, Zhi Li, Malignant gangliocytic paraganglioma of the duodenum with distant metastases and a lethal course, *World J. Gastroenterol.* 20 (2014) 15454–15461.
- [5] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, for the SCARE Group, The SCARE Statement: consensus-based surgical case report guidelines, *Int. J. Surg.* (2016) (in press).
- [6] C. Rowsell, N. Coburn, R. Chetty, Gangliocytic paraganglioma: a rare case with metastases of all 3 elements to liver and lymph nodes, *Ann. Diagn. Pathol.* 15 (2011) 467–471.
- [7] A. Sathyamurthy, A. Choudhary, D. Ng, S. Okponobi, A. Diaz-Arias, A. Grewal, Obstructive jaundice due to a rare periampullary tumor, *World J. Gastrointest. Oncol.* 5 (2013) 195–197.
- [8] S.J. Park, H. Kim do, H. Lim, J.H. Lee, K.D. Choi, H.J. Song, et al., Endoscopic resection as a possible radical treatment for duodenal gangliocytic paraganglioma: a report of four cases, *Korean J. Gastroenterol.* 63 (2014) 114–119.
- [9] J.I. Yang, J.S. Choi, G.H. Lee, B.W. Kim, S.J. Moon, M.S. Kang, et al., A case of ampullary gangliocytic paraganglioma, *Korean J. Internal Med.* 29 (2014) 375–378.
- [10] J. Chen, E. Sharon, S. Morgenstern, A. Geller, F. Greif, Local surgical resection of gangliocytic paraganglioma of the duodenal papilla, *Isr. Med. Assoc. J.* 11 (2009) 311–312.
- [11] J. Sucandy, G. Avers, D.J. Bertsch, Surgical management of endoscopically unresectable duodenal gangliocytic paraganglioma in a patient with partial upper gastrointestinal obstruction, *N. Am. J. Med. Sci.* 2 (2010) 547–551.
- [12] D.B. Dookhan, M. Miettinen, G. Finkel, Z. Gibas, Recurrent duodenal gangliocytic paraganglioma with lymph node metastases, *Histopathology* 22 (1993) 399–401.
- [13] A. Wong, A.R. Miller, J. Metter, C.R. Thomas, Locally advanced duodenal gangliocytic paraganglioma treated with adjuvant radiation therapy: case report and review of the literature, *World J. Surg. Oncol.* 3 (2005) 15.

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