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Case report

Gangliocytic paraganglioma treated with ampullectomy, A case report

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HIGHLIGHTS

- Gangliocytic paraganglioma are rare neuroendocrine tumors.
- Gangliocytic paragangliomas are typically found within the duodenum.
- These tumors are commonly benign, but have malignant potential.
- There is no defined standard of treatment for gangliocytic paraganglioma.

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ABSTRACT

Background: Gangliocytic paragangliomas are rare, and typically benign neuroendocrine neoplasms usually found in the second portion of the duodenum. Though recurrence is rare, metastatic cases have been noted. A standardized treatment has not been determined. An endoscopic resection can be carried out, unless metastasis has been noted. We present the case of a patient who underwent an ampullectomy, with successful removal of the tumor, and no recurrence on follow-up.

Case summary: We present a case involving a 58-year-old gentleman, who presented with dysphagia, and ultimately diagnosed with a periampullary mass proven to be a gangliocytic paraganglioma. The tumor was resected in-toto via an ampullectomy. The patient had no recurrence after twenty-one months.

Conclusion: In conclusion, we present the case of an incidental gangliocytic paraganglioma occurring in the periampullary region of the patient. Ampullectomy was carried out and to date the patient has had no recurrence. Rarity of this tumor, along with treatment choice, makes it especially worthy of notability.

1. Introduction

Gangliocytic paraganglioma (GP) is a rare neuroendocrine neoplasm that is predominantly located in the second portion of the duodenum, within the ampulla of Vater [1]. Normally this tumor presents asymptomatically, and is found incidentally through diagnostic studies. Histologically, GP is comprised of three distinct cellular components including epithelioid endocrine cells, ganglion-like cells, and spindle shaped cells [2]. Duodenal GP is considered a benign tumor; however, occasional lymph node metastasis has been reported in literature. Furthermore, the recurrence of this tumor following complete resection is rare [3]. To date, there is no standard of care regarding resection options. We hereby present a case of an asymptomatic periampullary GP, treated with an ampullectomy. The treatment of choice and rarity of the lesion makes it particularly notable in literature.

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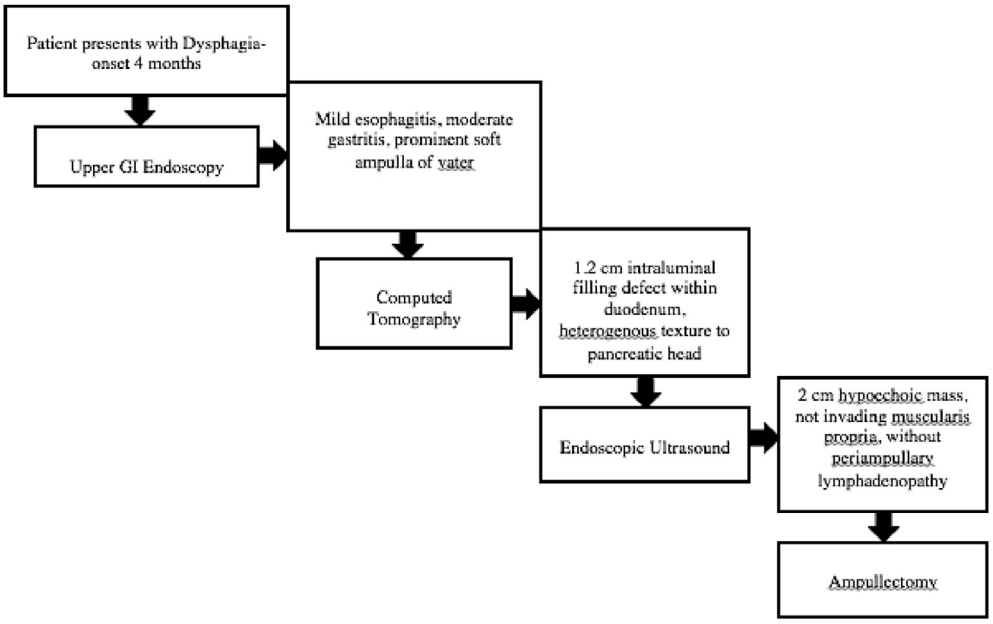
2. Timeline



3. Patient information

A 58 year old man with no significant past medical history presented with a four-month history of intermittent dysphagia to solid foods. The patient is a nonsmoker. He was prescribed omeprazole, which improved his symptoms.

4. Physical exam

General: awake and alert, no signs of distress.
HEENT: No palpable lymphadenopathy.
Cardiac: S1/S2 heard. No murmurs.
Respiratory: Clear to auscultation bilaterally.
Abdomen: Abdomen nondistended, nontender.

5. Diagnostic assessment

The patient was sent for an upper endoscopy, a prominent soft ampulla was noted along with esophagitis was noted (Fig. 1a and b). Gastric biopsy showed mild chronic gastritis. Duodenal biopsy at the ampulla of Vater identified a benign neoplasm composed of neurofibromatous and ganglion cells. Computed tomography scan of the abdomen showed a 1.2 cm intraluminal filling defect within the duodenum at the junction of the second and third portion, below the expected location of the ampulla (Fig. 2a and b). Slight heterogenous texture to the pancreatic head was noted. Laboratory evaluation was negative for anemia or hepatic dysfunction. Endoscopic ultrasound showed a 2 cm hypoechoic mass, not invading the muscularis propria, and without periampullary lymphadenopathy. While the pancreatic duct and pancreatic head appeared normal, the common bile duct seemed to be blending into the area of suspicion.

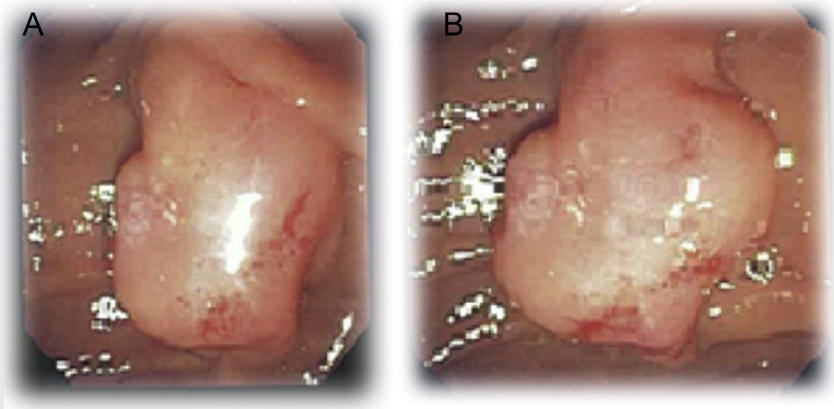


Fig. 1. a: Esophagogastroduodenoscopy showing prominent ampulla of Vater without any mucosal abnormality. b: Close picture of the same.

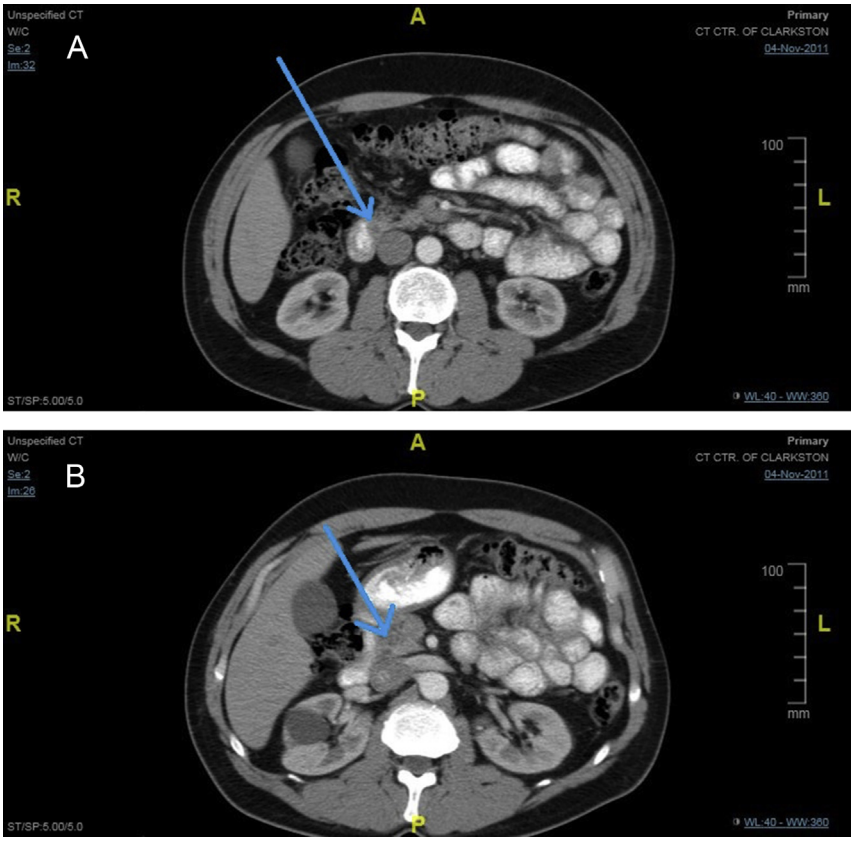


Fig. 2. a: Sagittal sections of computed tomography scan of abdomen showing filling defect within the duodenum lying at the junction of the second and third part. B: Sagittal sections of computed tomography scan of abdomen showing slight heterogeneous texture to the pancreatic head.

6. Interventions

Owing to the questionable involvement of the common bile duct, an endoscopic resection was considered risky. Hence, the patient was scheduled for an ampullectomy with possible conversion to a pancreaticoduodenectomy. Due to lack of signs of local invasion and common bile duct involvement intra-operatively, the ampullectomy was performed with confirmed negative margins. Gross examination of the resected tumor showed a 1.0 × 1.0 × 0.7 cm yellow circumscribed-appearing mass noted in the submucosa. Microscopically, it involved the muscularis mucosa, which was noted to be markedly thickened. Nests and cords of nerve and ganglion cells were observed, which were concordant with GP (Fig. 3). No signs of cellular atypia, significant mitotic activity or malignancy were identified.

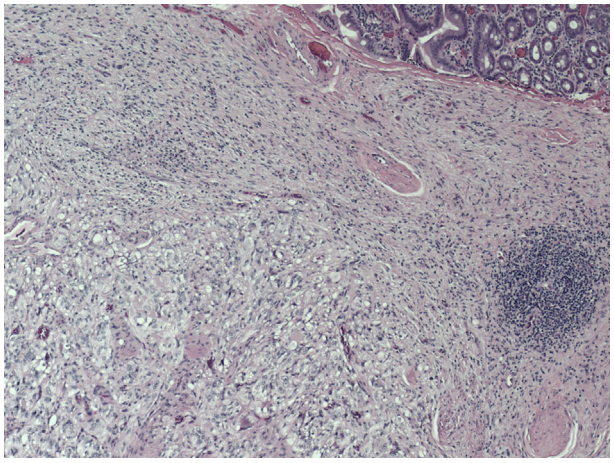


Fig. 3. Microscopy from resected mass showing nests and cords of nerve and ganglion cells.

7. Follow-up and outcomes

Follow up esophagogastroduodenoscopies were done at nine and twenty-one months, which showed no recurrence.

8. Discussion

GP is a rare tumor with only around 200 cases reported since it was first noted in literature in 1957 by Dahl et al. [4,5] It most commonly affects individuals in their 5th decade of life, but can range from 15 to 84 years of age, and has a male to female predominance (1.5:1) [4]. In 90% of cases, this tumor is located in the duodenum, with a particular predisposition to be near or at the ampulla of Vater [4]. Rarely it has also been noted in the lower spinal cord, respiratory tract, jejunum, esophagus, stomach, appendix, retromediastinum, pancreas, and as a mature teratoma [4]. The average size that these tumors reach is 2.5 cm at their widest diameter, but can range in size from 0.5 cm to 10 cm. [4], Histologically, GPs are composed of 3 types of cells: epithelial cells, spindle cells, and ganglion cells.

Typically and most commonly, these tumors are found incidentally on upper endoscopy, computed tomography scan, or ultrasound [6]. When present, the most common symptom is melena due to the ulcerated mucosa, followed by abdominal pain and

anemia [4]. Other signs and symptoms include pyloric/duodenal/biliary obstruction, dyspepsia, obstruction jaundice, and pancreatitis [7–10]. The patient presented with dysphagia, which was unrelated to the tumor itself.

As previously stated, these tumors are most commonly found on endoscopy and can present in either polyp or sessile form. Local and distant invasion needs to be determined to guide treatment options. Endoscopic ultrasound determines the depth of invasion and local lymph node metastasis, while computed tomography shows local lymph node metastasis as well as any distant metastasis. On both computed tomography and ultrasound, gangliocytic paragangliomas present as a homogeneously isoattenuated mass [11]. Tumors invading beyond the submucosal layer typically represent a risk factor for lymph node metastasis [4].

The course of GP is typically benign, with only 5–7% presenting with metastasis [4,7,11–15]. From recent literature, 18 cases of local lymph node metastases have been noted. Along with this, 3 distant metastases to the liver, bone, and concurrently to the liver and pelvic cavity have been observed [2]. There has been only one documented case of death due to this neoplasm. In this case, the tumor did not exhibit any histological atypia, high mitotic figures, nor proliferative index [2]. This case exemplifies the unpredictability of this neoplasm. To date, it is still unknown which histological features lead to a more malignant path. Ki-67 proliferation along with bcl-2 and p53 have been studied and have appeared to offer limited prognostic value [4]. Follow up monitoring by upper endoscopy has been recommended in the literature, and we concur with this. Subsequently, our patient had follow-ups at nine and twenty-one months postoperatively [16].

A true consensus has not been met on a standard of management, which in part is due to the rarity of the lesion. Many authors agree that endoscopic resection is a safe option [4]. Notably, the largest tumor resected by endoscopy was 5 cm [5]. An ampullectomy in this case was chosen because, even though no metastatic lymph nodes were viewed on computed tomography or endoscopic ultrasound, there was still questionable heterogenous texture of the pancreatic head as well as common bile duct involvement. In the case of local lymph node metastasis, a more radical approach is typically carried out: pancreaticoduodenectomy and lymph node dissection. The use of chemotherapy and radiation are questionable, with no response in the case of Li et al. to cyclophosphamide and vincristine [2]. We suggest to first determine the depth of local invasion as well as distant lymph node involvement. If, as in this case, there is questionable local invasion, the determination of the extent of invasion must be carried out.

In conclusion, we present the case of a gangliocytic paraganglioma occurring in the periampullary region of the patient. The presenting symptom of dysphagia was due to the esophagitis. Hence, it can be concluded that the lesion described in this report is incidental. With questionable common bile duct involvement, endoscopic resection was deemed too risky for the patient, but also we did not feel a radical surgery, such as a pancreaticoduodenectomy was reasonable. Regardless of the treatment choice, the rarity of gangliocytic paragangliomas makes it worthy of notability in literature as well as to add more data regarding this tumor for future studies.

9. Patient perspective

None.

10. Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request. IRB approval was received before initiation.

Conflict of interest statement

We have no conflict of interest. We have no financial disclosures.

Care criteria checklist

All work in the manuscript has been reported in line with the CARE criteria. You can find a completed checklist submitted along with the manuscript.[21]

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Ethical approval

We obtained IRB approval from our institution prior to initiation of the case report.

Author contribution

Dr. Mina Guerges, M.D.: Contribution to Case Report concept and design, literature review, writing the paper, editing the paper, final submission of the paper.

Eliza Slama, B.Sc.: Contribution to Case Report concept and design, literature review, writing the paper, editing the paper, final submission of the paper.

Bashar Maskoni, B.Sc., M.B.A.: Contribution to Case Report concept and design, literature review, writing the paper, editing the paper, final submission of the paper.

Dr. Sherwin Imlay, M.D.: Contribution to Case Report concept and design, literature review, writing the paper, editing the paper, final submission of the paper.

Dr. Malik McKany, M.D.: Contribution to Case Report concept and design, literature review, writing the paper, editing the paper, final submission of the paper.

Guarantor

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