

A Common Clinical Presentation and an Uncommon Diagnosis ‘Gangliocytic Paraganglioma’: Case Report

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Abstract

A common clinical presentation, however, the diagnosis was different from the common differentials. Presenting to you a case of malena, headache, dizziness, fatigue. He presented as a diagnostic dilemma. A battery of tests performed to ascertain the diagnosis. After multidisciplinary team approach, it was unanimously decided to subject the patient to surgery. The histopathological examination showed spindle cells, epithelioid cells and ganglion cells, thereby clinching the diagnosis of ‘Duodenal Gangliocytic Paraganglioma’. The first case was reported in 1957, however only a few hundred cases are available in the literature mostly in the form of case reports. Existing case series contain only single digit cases. Because of the rarity of the case, it is important to report thereby adding to the existing literature.

Keywords: Duodenum; Gangliocytic paraganglioma; Case report; Excision

1. Introduction

Gangliocytic Paraganglioma (GP) was first described in 1957 as duodenal ganglioneuroma by Dahl et al. [1]. In 1971, Kepes and Zacharias first described these tumors as GP because of the presence of both ganglion cells and epithelioid cells [2]. Gangliocytic paraganglioma mainly arises from the second part of the duodenum in close proximity to the ampulla of Vater, although the tumor can be seen throughout the gastrointestinal tract. GPs are exceedingly rare tumors, with approximately 280 cases identified in a MEDLINE search through November 2022 [3-6]. Hence it is important to report this case. Thereby adding to the existing literature. This case report has been reported in line with the SCARE Criteria 2023 [7].

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2. Case Report

2.1 Patient information

A 39yr/Male presented to a peripheral hospital with complains of headache, weakness, giddiness, fatigue, malena for a period of 1 week. On subsequent evaluation, Hb- 12.6gm/dl, Stool- occult blood positive. Upper GI endoscopy showed hiatal hernia, antral gastritis and duodenal ulcer. He was given symptomatic treatment and discharged home in 2 days.

2.2 Clinical findings

After 2 weeks he was presented to our hospital with complains of headache, giddiness and malena. He was again subjected to a battery of investigations, the reports were as follows: Hb- 9.4gm/dl, Platelets- 2,87,000.

2.3 Diagnostic assessment

Upper GI scopy showed bleeding from ampulla, with a bulky ampulla (FIG. 1). Side viewing duodenoscopy was done which showed periampullary neoplasm with ulcerations. Biopsy was taken from it. Lower GI Endoscopy was within normal limits. Histopathology report: benign ampullary mucosa, lamina propria shows mild inflammatory infiltrate of lymphocytes and plasma cells. No evidence of granulomas, dysplasia or malignancy. CEA: <0.5ng/ml. CA 19-9: 16.77 U/ml.

Computed Tomography (CT) Scan of abdomen and pelvis with oral+ IV contrast: polypoidal, heterogeneously enhancing lesion of size 2.4*1.5*1.5 cm seen in second part of the duodenum along medial wall just below periampullary region (FIG. 2a). No dilation of pancreatic duct or common bile duct. Periduodenal fat planes appear normal. Possibility of gastrointestinal stromal tumour (GIST) likely. No significant abdominal lymphadenopathy.

During this his Hb dropped to 8gm/dl. He was transfused with 2 pints of packed cell volume (PCV). Whole body PET CT showed small, intraluminal, polypoidal lesion in 2nd part of duodenum, closely abutting ampulla. (SUVmax: 3.6, size: 14*15mm) (FIG. 2b)



FIG 1. Showing upper GI endoscopy appearance.



FIG. 2a. CT Scan (blue arrow shows the lesion).

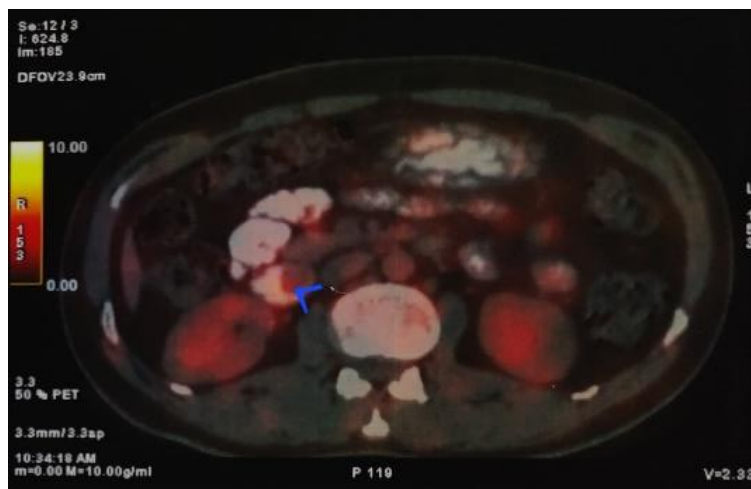


FIG. 2b. PET CT.

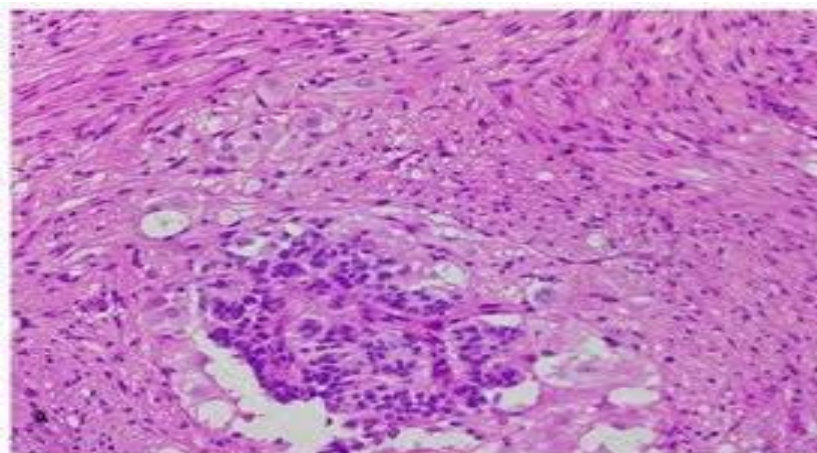


FIG. 3. H & E Staining showing spindle & ganglion cells.

2.4 Therapeutic intervention

Now this patient presented a diagnostic dilemma. With no definite diagnosis to the exact nature of the periampullary lesion, further management was contemplated. Being in close proximity to the ampulla and owing to ambiguity about the nature of the lesion and its size, gastroenterology opinion did not favour endoscopic management. Hence, we decided to go ahead with surgery.

An upper midline vertical incision was taken. The 2nd part of duodenum was opened in the right lateral part vertically. The lesion was seen in very close proximity to ampulla. Wide excision of the lesion was done. Duodenal incision was closed primarily in two layers. The drain kept. Patient did well postoperatively. He was discharged on post-op day 5, tolerating a full diet. Histopathology (FIG. 3) report showed gross size of 1.5 cm × 1.2 cm × 0.8 cm. Submucosal tumor with relatively circumscribed borders composed of spindle cells, small clusters of epithelioid cells and ganglion cells. These features are suggestive of duodenal gangliocytic paraganglioma. Resection margins are free.

2.5 Follow up and outcome

On routine subsequent follow-up of 20 months, patient is doing well. There is no evidence of local or systemic recurrence of the disease.

3. Discussion

GP is a rare diagnosis, usually not even considered in the differentials in the early diagnostic stages. GP shows slight male predominance (1.5:1) and a mean tumor size of approximately 2.57 cm, the most common clinical presentation is gastrointestinal bleeding; other symptoms include abdominal pain, anemia, nausea, weight loss, fatigue, and jaundice [3]. Our case report has a male patient with symptoms suggestive of anaemia and upper GI bleeding. Initially, on endoscopy he was diagnosed as antral gastritis and duodenal ulcer and was discharged with symptomatic treatment.

GP is difficult to diagnose. Radiologically, GP usually presents a diagnostic dilemma [8]. Due to the submucosal location of this tumor, preoperative pathologic diagnosis is difficult based on endoscopic biopsy alone with a diagnostic rate of only 11.4% [9]. As in this case, preoperative biopsy was inconclusive showing mild inflammatory infiltrate. Ct scan report suggested it to be tumor of 2.4 cm, with possibility of it being GIST. Due to diagnostic uncertainty, a PET Scan was performed. Whole body PET CT showed small, intraluminal, polypoidal lesion in 2nd part of duodenum, closely abutting ampulla (SUVmax: 3.6, size: 14 × 15 mm).

A multidisciplinary team decided that surgical resection should be undertaken. Hence open D2 duodenotomy with wide excision of the lesion was done. Postoperative HPE report diagnosed it as GP. Resection of the tumor is the only definitive treatment [10].

Patients with local disease and no malignant features or lymph node metastasis on preoperative workup can be considered for endoscopic mucosal resection [11]. Depending on the size and location of the tumor and the expertise available, laparoscopic

or robotic or open approach can be undertaken. Complete resection of the tumor with free margins constitutes adequate treatment. On the routine follow-up of 20 months the patient is doing well with no evidence of recurrence or metastasis.

4. Conclusion

Duodenal gangliocytic paraganglioma is a rare lesion commonly arising in the 2nd part of duodenum. It is usually benign. Preoperative radiology or histopathology diagnosis is very difficult and requires a high degree of suspicion. Resection constitutes definitive treatment for GP.

5. Author Contributions

Drafting the report, design, and revision done by Dr Atiya Lambe. Dr Atiya Lambe and Dr Rajesh Yadav contributed to the concept of reporting the case and the patient data recording. Final approval of the work to be published was done by Dr Atiya Lambe and Dr Rajesh Yadav.

6. Acknowledgements

The authors have no financial or proprietary interest in the subject matter of this article.

7. Conflicts of Interest

None.

8. Sources of Funding

None.

9. Consent

A written informed consent was taken from the patient for reporting the case and the accompanying images.

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