Case report

Gangliocytic paraganglioma of duodenum

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An elderly person, aged 60 years, presented with pain abdomen localized to the epigastric region, of one year duration. There was a past history of a single episode of passing black-colored tarry stools and hematemesis. Ultrasonography (USG) of abdomen reported a suspected duodenal / periampullary growth. Biopsy performed was inconclusive. Contrast enhanced computerized tomography (CECT) scan abdomen suggested suspected submucosal leiomyoma of duodenum with intrahepatic biliary distension.

Whipple’s procedure was performed which recovered a growth measuring 6x5x5cms arising in periampullary region of second part of duodenum. Pancreatic duct and common bile duct (CBD) were dilated. The histopathological findings were suggestive of neuroendocrine tumor of duodenum. The diagnosis of gangliocytic paraganglioma was confirmed by immunohistochemical stains.

Key words: Gangliocytic paraganglioma, duodenum, Whipple’s procedure

Gangliocytic paraganglioma (GP) is a rare tumor, occurring exclusively in the second portion of the duodenum. Generally, this tumor has a benign clinical course, having rare malignant potential with tendency to metastasize to regional lymph nodes.

Case report

An elderly person, aged 60 years, presented with complaint of pain abdomen of 1 year duration, localized to epigastric region. Past history of a single episode of passing black-colored tarry stools, along with history of hematemesis 5 months back was noted. Patient gave history of weight loss and loss of appetite. He was a chronic alcoholic. Family history was not significant. His physical examination was non-contributory. Ultrasonography (USG) of abdomen reported a duodenal / periampullary growth. An endoscopic biopsy performed on the growth was inconclusive. Subsequently, contrast enhanced computerized tomography (CECT) scan abdomen revealed a mass measuring about 6cms in the second portion of the duodenum suggestive of submucosal leiomyoma of duodenum with intrahepatic biliary distension. Surgery was indicated based on these findings indicating high possibility of malignancy. Whipple’s procedure was performed and a growth measuring 6x5x5cms was found in the ampulla of Vater.

No evidence of any distant metastasis or lymph node enlargement was noted.

Macroscopic and microscopic findings

Gross: Whipple’s specimen consisting of gall bladder along with partial distal gastrectomy,
Gangliocytic paraganglioma

Fig 1. Gross: Proximal resected end with pylorus (upper red arrow), duodenum (blue arrow), distal end of duodenum (green arrow), gall bladder (lower left red arrow)

Fig 2. Gross: Tumor (red arrow), proximal resected end, pylorus (black arrow), distal resected end, duodenum (green arrow), gall bladder (blue arrow)

Fig 3. H & E stain showing normal duodenum, pancreas and tumor.

A: 40X View-normal duodenum with brunner’s glands.
B: 4X View-normal duodenum & pancreas.
C: 40X View-Tumour in nests & trabeculae.
D: 40X View-Tumour in zell-ballen like pattern.

Sections revealed a submucosal tumor, having a triphasic pattern composed of epithelioid-neuroendocrine cells, spindle cells with schwannian differentiation and scattered ganglion cells. The epithelioid cells were arranged in solid nests, ribbons, trabeculae and pseudoglandular patterns. Individual cells had moderate to abundant eosinophilic to basophilic cytoplasm with ovoid nuclei. Spindle cells having elongated nuclei, formed slender fascicles wrapping around and sweeping in between the nests of epithelioid cells.
The ganglion cells were seen either in small clusters or individually scattered. Individual ganglion cells had abundant eosinophilic cytoplasm with round eccentric nucleus and prominent nucleolus. Some cells showed intracytoplasmic brown-colored Nissl’s substance. Mitotic count was one-tenth per HPF. Pleomorphism and necrosis was absent. Pancreas, gall bladder, common bile duct were not invaded by the tumor. No lymph node metastasis was detected (Fig 3 & 4).

A provisional diagnosis of neuroendocrine tumor of periampullary region (second part of duodenum) was considered.

Subsequently, immunohistochemistry (IHC) was done (Table 1), which showed positivity for chromogranin, synaptophysin, NSE and S-100 whereas cytokeratin and CEA were negative (Fig 5 & 6). Based on the above IHC features a diagnosis of gangliocytic paraganglioma of periampullary /second part of duodenum was confirmed.

**Discussion**

Gangliocytic paraganglioma (GP) is a rare tumor, occurring exclusively in the second portion of the duodenum. Dahl et al in 1957, first described the lesion and it was further characterized as a benign non-chromaffin paraganglioma by Taylor and Helwig in 1961. The term “gangliocytic paraganglioma” was coined by Keeps and Zacharias in1971, recognizing the features in common with both paraganglioma and ganglioneuroma. Gangliocytic paraganglioma is usually seen in the periampullary region of duodenum, though rare cases have been reported in jejunum, pylorus, esophagus, pancreas and appendix.

**Table 1: Immunohistochemistry report**

<table>
<thead>
<tr>
<th>IHC marker</th>
<th>Result</th>
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<tbody>
<tr>
<td>Chromogranin A</td>
<td>+</td>
</tr>
<tr>
<td>S-100</td>
<td>+</td>
</tr>
<tr>
<td>NSE</td>
<td>+</td>
</tr>
<tr>
<td>Ki 67</td>
<td>1% positivity</td>
</tr>
<tr>
<td>CEA</td>
<td>-</td>
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<tr>
<td>Cytokeratin</td>
<td>-</td>
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Fig 5. Immunohistochemical stain showing chromogranin positivity

Fig 6. Immunohistochemical stain showing S100 positivity
Recently, 3 cases of pulmonary GP have been reported12,16. The age at presentation ranges from 15-82 years, with slight male predominance. Clinically, GPs arising in the gastrointestinal tract present with bleeding, abdominal pain or obstruction; though some cases were incidental findings at autopsy12. Abdominal pain is the most common presenting symptom.

GP has three characteristic histologic components: epithelioid, ganglion and spindle cell. The proportion of the three cell types varies in each tumor, but each component shows characteristic immunohistochemical staining, similar to those observed in our case11. They may also show positivity for pancreatic polypeptide12.

Theories on the origin of GPs vary widely, and yet have not been able to explain the combination of endocrine, ganglion and spindle cells observed in a single tumor. The tumor components are of different embryologic origins; the first being of endodermal origin and the others originating from neural crest tissue. Initially, it was suggested that these tumors were of ectodermal origin, from pluripotent stem cells derived from the neural crest, which were found in Lieberkühn's glands or the celiac ganglion during fetal development11 but in view of the occurrence of GPs in different sites in the duodenum and its variable histology, it has been proposed that they originated from endodermal pluripotent progenitor stem cell that has the potential for divergent differentiation13. Some authors proposed GPs were hamartomas of endodermal (epithelial cells) and neuroectodermal (ganglion and spindle cells) origin11. Most authors considered them to be variants of gastrointestinal tract paragangliomas14. Paragangliomas may differentiate to other neuroectodermal elements, including neurons and schwann cells15.

Most GPs are benign and are amenable to local resection. However, instances of recurrence, tumor metastasizing to regional lymph node involvement and distant metastases have been reported. This tumor metastasis show all the three cell components16. In a single case of regional lymph node metastasis, tumor consisted of only epithelioid cell component17. Some GP may recur with lymph node metatasis10.

Conclusion

Gangliocytic paraganglioma may be misdiagnosed in view of non-specific clinical symptoms like pain abdomen for peptic ulcer disease and radiologically for periampullary adenocarcinomas or gastrointestinal stromal tumor, as observed in our case. Also, endoscopic biopsy may be inconclusive in view of submucosal location of tumor as noted in our study.

This tumor though rare should be considered as a differential diagnosis in neoplasms of periampullary region and it has a good prognosis as compared with other tumors.

Since GP may recur or metastasize, pancreaticoduodenectomy with lymph node dissection may be indicated for large lesions with infiltrative margin, or lesions with pleomorphism and mitoses.

Acknowledgment: None

Conflict of interest: None

References


