Gangliocytic Paraganglioma of the duodenum : A rare entity

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To the Editor,

Gangliocytic paraganglioma (GP) is a rare condition and the available data consist mainly of case descriptions. The lesion was first described by Dahl *et al*. in 1957 (1). The term GP was introduced by Kepes and Zcharias and is derived from his histological features of neurendocrine tumor, paraganglioma and ganglioneuroma. The tumor is typically located in the second or third part of the duodenum (2).

We describe the case of a 68 year old Caucasian female who had a diagnosis of a GP in the third part of the duodenum that illustrates the difficulties in making a final pretreatment diagnosis and choosing the right treatment modality. The patient complained of dyspepsia and regurgitation since 5 months. She denied abdominal pain, nausea and pyrosis. There was no history of weight loss. The past medical history revealed hysterectomy but no other significant problems and the physical examination was normal. The abdomen was soft and no masses were palpable. Laboratory tests were completely normal. An esophagogastroduodenoscopy (EGD) was unremarkable but a computed tomography disclosed an intraluminal mass in the third portion of the duodenum with a diameter of 3,5 cm. Push enteroscopy revealed a submucosal tumor in the third portion of the duodenum (Fig. 1). The bowel mucosa adjacent was injected with India ink for subsequent surgical identification. The patient underwent a robot assisted duodenotomy and resection of the tumour ; the periampullary area was spared. The lesion measured 3,8 × 3,3 × 2,8 cm, was well circumscribed and limited to the submucosa. The resection margin was negative. The neoplasm had a ganglion cell and in some area's spindle cell morphology with neural fibers (Fig. 2). There was no mitotic activity (p-Histon-H3 staining). Staining for synaptophysin was strongly positive and partially positive for chromogranine and anti-trypsine. Staining was negative for cytokeratine 7 and for cytokeratine 20. A diagnosis op GP was made.

GP is a rare tumor. In a recent survey published in 2011 (3) 173 patients with duodenal GP were retrieved using PubMed and a Japanese database and clinical and histological data were reviewed. The duodenum is the most common site of the disease (90%). The tumor is often located at the papilla of Vater. The presenting clinical symptoms are gastrointestinal bleeding (45%), abdominal pain (42.8%) and anemia (14.5%). Biliary obstruction is rare (4.6%) (3). The pathognomonic features of these tumors are the identification of three distinct cellular elements : spindle cells, epithelial cells and ganglion cells. Immunohistological these tumors stain positive for a variety of markers. GP has generally been regarded as a neurendocrine tumor and the pancreatic islet cells may be the origin (3).

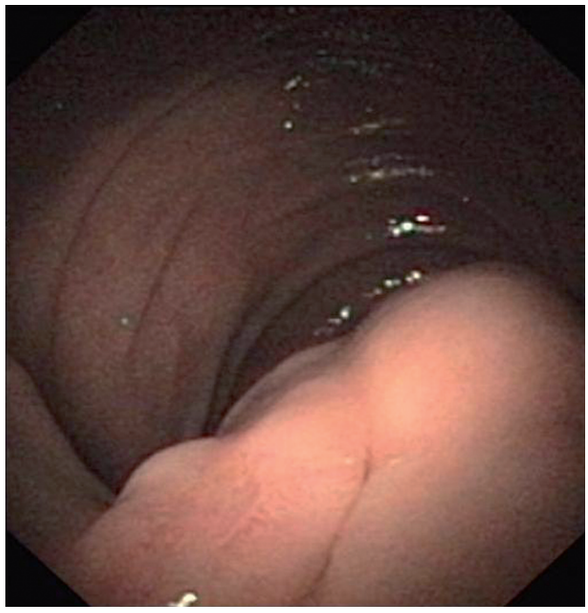

Fig. 1. — Push enteroscopy : Submucosal tumor in the third part of the duodenum.

GP is considered as a benign tumor, however in 61% of cases the tumor is invasive and invades beyond the submucosal layer. In tumors invasive beyond the submucosa, the risk of lymph node metastasis is 10% (3). This confirms that PG has a malignant potential. Immunohistological examination, Ki-67 labeling index and mitotic index are not correlated with malignant behavior and as such not useful as prognostic indicator (4).

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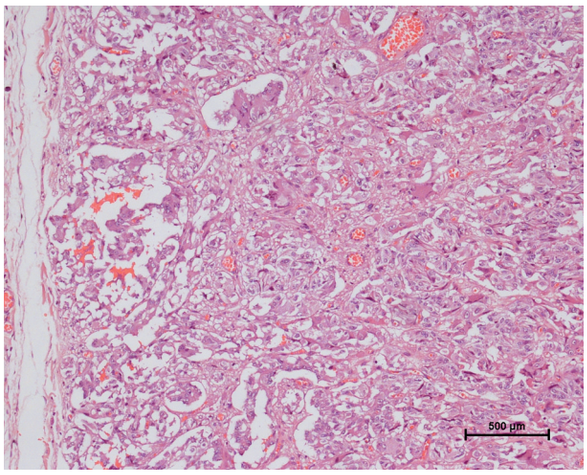

Fig. 2. — Histology : Neoplasm with a ganglion cell and in some area's spindle cell morphology with neural fibers.

Treatment consists of complete resection either surgical or endoscopic based on location and preoperative staging. Usually there is no pretreatment anatomopathological confirmed diagnosis and the tumor is not thought to be a GP. Other tumors such as neuroendocrine tumors or gastrointestinal stromal tumors are suspected.

Although pancreaticoduodenectomy has been proposed as treatment of choice for periampullary lesions (5) several cases treated with local surgical or endoscopic resection have been described. Because the possibility of recurrence or metastasis is not completely excluded, follow up is advised.

References

1. DHAL E.V., WAUGH J.M., DAHLIN D.C. Gastrointestinal ganglioneuromas : brief review with report of a duodenal ganglioneuroma. *Am. J. Pathol.*, 1957, **33** : 953-65.
2. TAYLOR H.B., HELWIG E.B. Benign nonchromaffin paragangliomas of the duodenum. *Virchows Arch. Pathol. Anat. Physiol. Klin. Med.*, 1962, **335** : 356-66.
3. OKUBO Y., MEGUMI M., NEMOTE T., KITAHARA K. *et al.* Literature survey on epidemiology and pathology of gangliocytic paraganglioma. *BMC Cancer*, 2011, **11** :187.
4. OKUBO Y., YOKOSE T., TUCHIYA M., MITUDA A. *et al.* Duodenal gangliocytic paraganglioma showing lymph node metastasis : A rare case report. *Diagnostic Pathology*, 2010, **5** : 27.
5. BUCHER P., MATHE Z., BÜHLER L., CHILCOTT M. *et al.* Paraganglioma of the ampulla of vater : a potentially malignant neoplasm. *Scand. J. Gastroenterol.*, 2004, **39** : 291-295.