

Gangliocytic paraganglioma of the duodenal papilla associated with esophagogastric adenocarcinoma

A 3-cm painless epigastric mass was found on examination in 53-year-old man with abdominal pain and vomiting for the past 6 months. An abdominal computed tomography (CT) scan showed asymmetric thickening of the distal esophagus, and upper endoscopy showed a neoplastic lesion in the distal esophagus (histological examination of the biopsy revealed adenocarcinoma). In the duodenum, we found a subepithelial lesion in the papilla, measuring about 2 cm, which was covered by normal mucosa, and bile drainage was also normal. Endoscopic biopsies did not reveal neoplastic cells (● Fig. 1).

Endoscopic ultrasound showed a hypoechoic heterogeneous lesion in the cardia, infiltrating the serosa, accompanied by perilesional hypoechoic, round, well-defined nodes smaller than 1 cm in size. In the duodenal papilla, we found a hypoechoic homogeneous lesion restricted to the deep mucosa, measuring 25×15 mm, with no invasion of the biliary and pancreatic ducts (● Fig. 2).

The patient was referred for surgical resection of the esophageal tumor, duodenotomy and papillary tumor excision, and biliary drainage. Pathological specimens showed T3N2 esophagogastric adenocarcinoma and gangliocytic paraganglioma of the duodenal papilla (● Fig. 3), with neoplastic cells positive for chromogranin (● Fig. 4a), synaptophysin (● Fig. 4b), and S100 protein, and with a low mitotic index and Ki67 < 1.

Gangliocytic paragangliomas are rare tumors, mainly arising in the duodenal papilla. Only a few cases have been reported in the literature, and we did not find any reports of the tumor in association with other upper gastrointestinal malignancies [1,2]. Endoscopic ultrasound can aid in evaluation of the invasiveness of the lesion and in some cases, in obtaining histological specimens for immunohistochemical analyses to confirm the diagnosis [3,4]. Despite its benign behavior, most patients reported in the literature were treated with surgical resection [1–5]. In the present case endoscopic resection was not considered because of the difficulty in accessing the duodenal papilla



Fig. 1 Endoscopic view of a subepithelial lesion in the duodenal papilla of a 53-year-old man with abdominal pain and vomiting.



Fig. 2 Endoscopic ultrasound view of the papillary lesion.

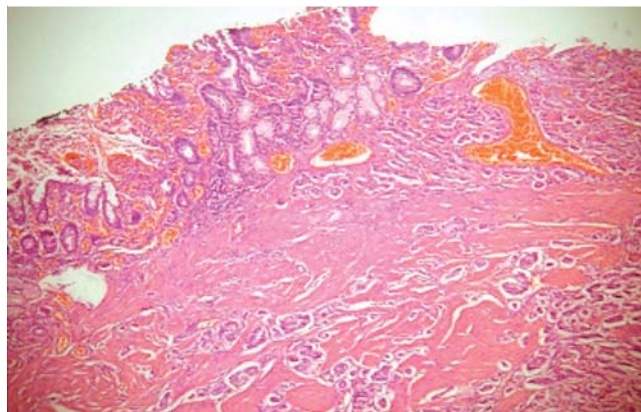


Fig. 3 Histological section (hematoxylin and eosin stain) showing gangliocytic paraganglioma.

la post-gastrectomy if required. Thus, in the present case, we elected to carry out local surgical excision in the form of a duodenotomy during the esophagogastrectomy procedure.

Endoscopy_UCTN_Code_CCL_1AB_2AZ_3AB

Competing interests: None

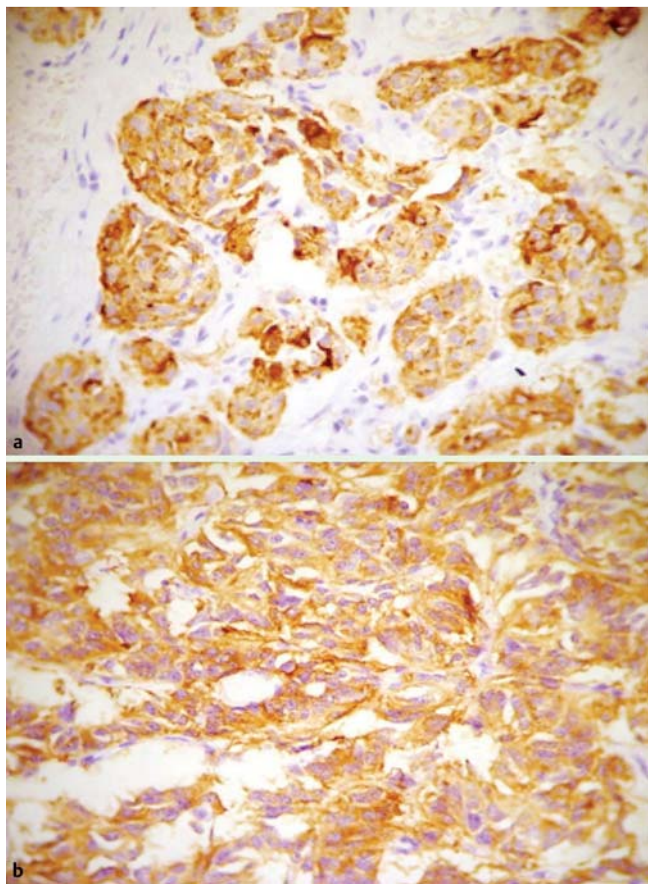


Fig. 4 Histological sections showing positive staining for **a** chromogranin and **b** synaptophysin.

M. S. Assef¹, A. P. C. Carbonari¹, O. Araki¹, F. Nakao¹, I. Marchetti¹, M. T. Medeiros², P. Kassab³, C. A. Malheiros³, L. B. Rossini¹

¹ French-Brazilian Center of Endoscopic Ultrasound (CFBEUS), Department of Endoscopy, Santa Casa Faculty of Medicine São Paulo, Brazil

² Department of Pathology, Santa Casa Faculty of Medicine, São Paulo, Brazil

³ Department of Surgery, Santa Casa Faculty of Medicine, São Paulo, Brazil

References

- 1 Sundarajan V, Robinson-Smith TM, Lowy AM. Duodenal gangliocytic paraganglioma with lymph node metastasis: a case report and a review of the literature. *Arch Pathol Lab Med* 2003; 127: e139–141
- 2 Nakamura T, Ozawa T, Kitagawa M et al. Endoscopic resection of gangliocytic paraganglioma of the minor duodenal papilla: case report and review of the literature. *Gastrointest Endosc* 2002; 55: 270–273
- 3 Hashimoto S, Kawasaki S, Matsuzawa K et al. Gangliocytic paraganglioma of the papilla of Vater with regional lymph node metastasis. *Am J Gastroenterol* 1992; 87: 1216–1218
- 4 Bucher P, Mathe Z, Bühler L et al. Paraganglioma of the ampulla of Vater: a potentially malignant neoplasm. *Scand J Gastroenterol* 2004; 39: 291–295
- 5 Nwakakwa V, Kahaleh M, Bennett A et al. EMR of ampullary gangliocytic paragangliomas. *Gastrointest Endosc* 2005; 62: 318–322

Bibliography

DOI <http://dx.doi.org/10.1055/s-0031-1291759>
Endoscopy 2012; 44: E165–E166
 © Georg Thieme Verlag KG
 Stuttgart · New York
 ISSN 0013-726X

Corresponding author

A. P. C. Carbonari

French-Brazilian Center of Endoscopic Ultrasound (CFBEUS)
 Department of Endoscopy
 Santa Casa Faculty of Medicine
 Martinico Prado 106
 ap 41. São Paulo
 Brazil 01224-010
augustocarbonari@yahoo.com.br