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Case Report

Gangliocytic Duodenal Paraganglioma: A Difficult Preoperative Diagnose

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Abstract

Gangliocytic paraganglioma (GP) is a rare tumor, considered benign, generally located in the second portion of the duodenum. In most cases comma it is an incidental finding. We report a case of GP discovered during a gastroscopy in a woman, which subsequently underwent endoscopic mucosal resection. Fine-needle aspiration biopsy during endoscopic ultrasound examination is not often enough for the diagnose and it cannot replace histological examination. Malignant potential is still unclear.

Keywords: *Paraganglioma; NET; Duodenal; Chromogranin; EUS; FNA*

Background

Gangliocytic paraganglioma (GP) is unusual neoplasia mainly observed in the second portion of the duodenum, near the ampulla of Vater. It is considered a benign tumor but, in literature, cases with lymph nodes metastasis and even few cases with distant metastases have been reported since the first description by Dahl., *et al*. in 1957 [1] Among duodenal neuroendocrine tumors it is considered the third most frequent, preceded by gastrinoma and somatostatinoma [2]. Typical symptoms reported are gastrointestinal bleeding, abdominal pain, anaemia, nausea and weight loss but it is more commonly found incidentally [3].

Case Report

We report a case of a 68-year-old woman with a duodenal GP. She presented to our department for abdominal pain, diarrhoea with weight loss (she referred 5 kilograms in the prior two weeks) and a history of gastroesophageal reflux disease. An esophagogastroduodenoscopy (EGDS) was performed to investigate better and it revealed a polypoid lesion in the first portion of the duodenum. It was described as a sessile polyp of 6 mm, with a mucosal surface appearing smooth, with no ulceration or bleeding (Figure 1). Subsequently, Endoscopic Ultrasound (EUS) characterized the lesion as ovoidal, mildly ipoechoic, involving the first 2 wall layers, mucosal and submucosal (Figure 2), no suspicious lymphadenopathies were identified in the areas explorable near the diaphragm. Fine-needle aspiration biopsy (FNA) was executed using a 22 Gauge needle and it deposed for neuroendocrine tumor well differentiated (NET G1, Ki-67 2%). Due to these findings and considering the lesion had not entered the muscle layer, endoscopic mucosal resection was set as treatment. Histological examination of the resected specimen revealed a lesion arranged in solid and trabecular pattern, with triphasic cellular differentiation, deposing for gangliocytic paraganglioma: epithelioid neuroendocrine cells, spindle-cells with Schwannian cell differentiation and ganglion-like cells (with eccentric nuclei, large nucleoli and abundant clear cytoplasm). There were no necrosis or mitotic figures, Ki67 was 2% (conform to FNA) and as immunohistochemical characteristics, was reported the presence of the typical neuroendocrine markers as Synaptophisine, Chromogranin A and moreover, of cytokeratins AE1/AE3 and S-100 (Figure 3). To improve differential diagnosis

with NET G1, as recently evidence have demonstrated, we tested immunoreactivity for the progesterone receptors (PR) and pancreatic polypeptide (PP) but it resulted negative [4]. The patient showed neuron-specific enolase (NSE) and CgA serum levels increase, but normal level of plasma free and urine metanephrines. Ga68-DOTATOC PET/CT, performed after the endoscopic treatment, resulted negative for the presence of tissue expressing somatostatin receptors. The first follow-up EUS, after 4 months, revealed no residual disease in the duodenal wall nor regional lymph nodes. After two years no evidence of recurrence was found so the patient is still in follow up for the long-term outcome.

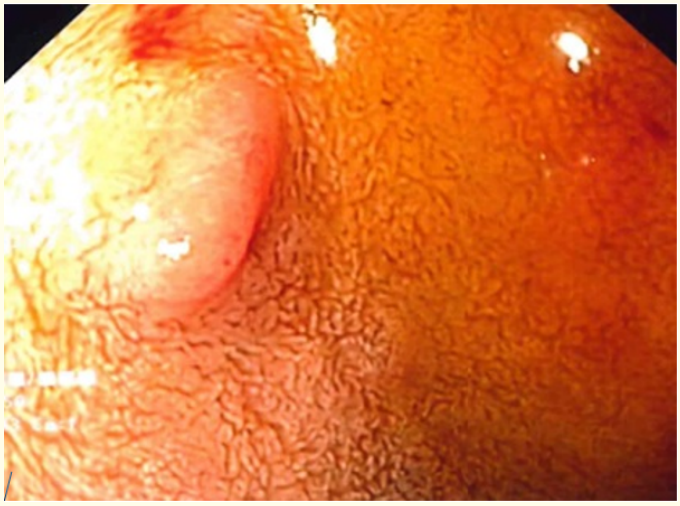

Figure 1: *Polypoid lesion in the duodenal bulb in EGDS.*

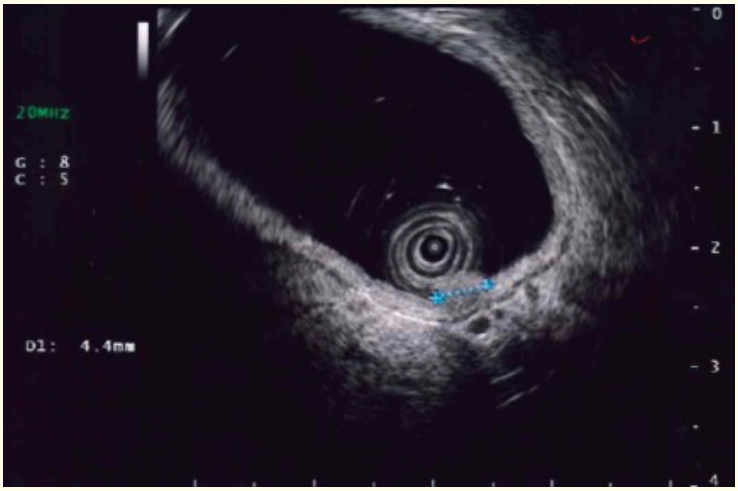

Figure 2: *EUS imaging: ovoidal, mildly ipoechoic lesion, involving mucosal and submucosal layers.*

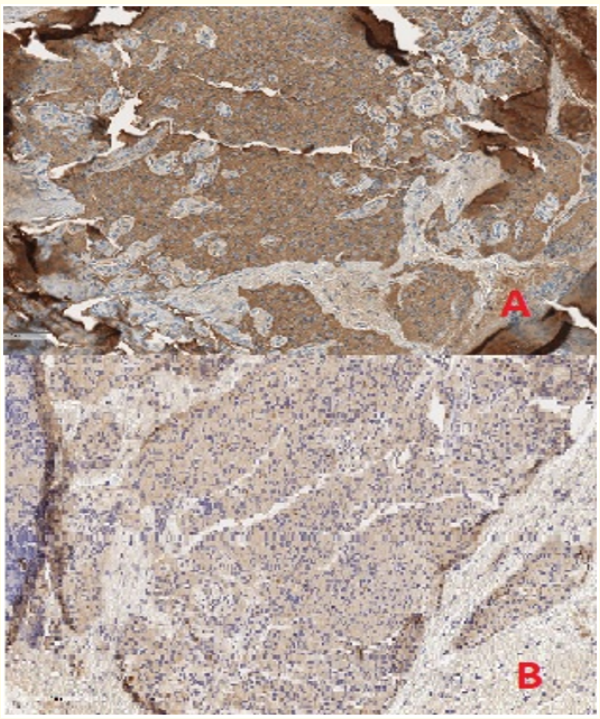

Figure 3: *Positivity for CgA (A) and for cytokeratins AE1/AE3 (B). Courtesy of Dr F. Maletta.*

Discussion

GP is a very rare neoplasia and generally appears in the periampullary region. In this case we describe a GP located in the duodenal bulb. In addition to being easily misdiagnosed as a duodenal neuroendocrine tumor (NET), this tumor goes into differential diagnosis including gastrointestinal stromal tumor (GIST), adenocarcinoma, ganglioneuroma and smooth muscle tumor [5,6]. Basing on one of the most interesting theories about GP origin, the three cellular elements seem to originate from pluripotent stem cells located at the bottom of crypts in the duodenum but further investigations are needed [7]. Diagnosis depends on histopathological and immunohistochemical examinations and it's usually hard to diagnose it only with cytological material from FNA, before a radical treatment of the lesion. Due to its unclear malignant potential, surgical or endoscopic resection is necessary, there is no specific adjuvant treatment strategy in case of regional or distant metastases [8] and it's not defined if a residual tumor after endoscopic procedures can be controlled by radiotherapy or chemotherapy alone without undergoing surgery. Based on this, a close post-treatment surveillance is needed.

Disclosure, Funding and Consent

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