Letters to the Editor

Ampullary carcinoid tumor. An atypical location

Key words: Carcinoid tumor. Ampulloma. Extrahepatic bile ducts. Ampulla of Vater.

Dear Editor,

A 41-year-old male patient came to emergency because of conjunctival jaundice after suffering an infection of upper respiratory tract. His medical history included amputation of right leg after occupational accident and neurofibromatosis type I or Von Recklinghausen disease. On examination, patient had typical "café au lait" spots and subconjunctival jaundice, soft and depressible abdomen with one finger-breadth hepatomegaly. Analytically, he showed moderate macrocytic anemia (hemoglobin 12.7 g/dL) and elevated liver enzymes (total bilirubin 1.75 mg/dL, direct bilirubin 1.12 mg/dL, AST 186 U/l, ALT 331 U/l, GGT 1185 U/l and alkaline phosphatase 563 U/l). After classified as obstructive jaundice, imaging tests were requested (ultrasound and CT scan) to assess the cause, which showed a distal obstruction of bile duct without demonstrable cause. The study was supplemented by oral endoscopy where was appreciated a protrusion of duodenal mucosa in second portion, suggestive of ampuloma or pancreatic head cancer. An endoscopic retrograde cholangiopancreatography (ERCP) demonstrated a friable tumor of 1 cm in diameter. Biopsy was highly suggestive of neoplastic proliferation, which had immunohistochemical characteristics of neuroendocrine carcinoid tumor (CR-, chromogranin A + + +, Vimentin +/-, CEA -, EMA + + +, S100 -, CK7 -, CK20-). With the diagnosis of ampullary carcinoid tumor, it was decided performing elective surgery, realizing a pancreaticoduodenectomy (pylorus-

preserving modification of classic Whipple procedure). Postoperative course was favorable and uneventful; at present, after more than 8 years after surgery, patient is asymptomatic and without signs of local or distant recurrence.

Carcinoid tumors are the most common neuroendocrine tumors. Incidence has increased considerably in recent years, but can not be ruled to be due to an improvement in diagnostic methods (1).

The first ampullary carcinoid tumor was described by Oberndorfer in 1907 (2), being published to date approximately 120 cases in the literature (3). These tumors are very rare (2% of all ampullary tumors and 0.05% of all gastrointestinal carcinoids), with an incidence even lower than duodenal carcinoids (2%) (4). Most common symptoms are, in order of frequency, jaundice (53.1%), abdominal pain (24.6%), pancreatitis (6%) and weight loss (3.6%) (5).

Diagnosis is established by histological and immunohistochemical study of the lesion, being generally postoperative, because biopsy during endoscopy or ERCP has a high failure rate, due to its often submucosal location (6).

It has been demonstrated that tumor size has no correlation with metastatic potential, unlike duodenal carcinoids where size, invasion of muscularis propria and mitotic activity rather correlated, so that duodenal tumors smaller than 2 cm can be resected locally (7). So, given the propensity of ampullary carcinoid tumors to develop lymph node metastases, we recommend performing a Whipple procedure, regardless of tumor size, although there is no concrete evidence that this technique improves survival (8).

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References

1. Varas-Lorenzo MJ, Muñoz-Agell F, Espinós-Pérez JC, Bardají-Bofill M. Tumores carcinoides gastrointestinales. *Rev Esp Enferm Dig* 2010;102:533-7.

2. Oberndorfer S. Karzinoide Tumoren des Dunndarms. *Frankf Zschr Pathol* 1907;1:426-32.
3. Ozsoy M, Ozsoy Y, Canda AE, Nalbant OA, Haskaraca F. The rare malignancy of the hepatobiliary system: ampullary carcinoid tumor. *Case Report Med* 2011;17:30-6.
4. Hartel M, Wente MN, Sido B, Friess H, Buchler MW. Carcinoid of the ampulla of Vater. *J Gastroenterol Hepatol* 2005;20:676-81.
5. Senda E, Fujimoto K, Ohnishi K, Higashida A, Ashida C, Okutani T, et al. Minute ampullary carcinoid tumor with lymph node metastases: a case report and review of literature. *World J Surg Oncol* 2009;22:7-9.
6. Selvakumar E, Rajendran S, Balachandar TG, Kannan DG, Jeswanth S, Ravichandran P, et al. Neuroendocrine carcinoma of the ampulla of Vater: a clinicopathologic evaluation. *Hepatobiliary Pancreat Dis Int* 2008;7:422-5.
7. Rohan VS, Narendra H, Patel JJ, Tankshali RA. Carcinoid of ampulla of Vater. *Indian J Cancer* 2007;44:90-2.
8. Carter JT, Grenert JP, Rubenstein L, Stewart L, Way LW. Neuroendocrine tumors of the ampulla of Vater: biological behavior and surgical management. *Arch Surg* 2009;144(6):527-31.