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Neuroendocrine tumor of the ampulla of Vater. Rreview of the literature

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Medical case

Neuroendocrine tumors (NETs) or carcinoids represent a small percentage of gastrointestinal neoplasms (1). An unusual case is described due to its location in the ampulla of Vater, being an extremely rare cause of biliary obstruction. The therapeutic approach remains controversial today.



A 43-year-old male, without other medical history, referred to the Digestive outpatient consultation for elevation of liver enzymes, with no associated symptoms, and dilatation of the bile and pancreatic duct in ultrasound image. CT findings (Figure 1A) suggests an ampulloma and a duodenoscopy (Figure 1B) and echoendoscope (Figure 1C-D), confirmed a 1.6cm ampullary lesion. Endoscopy biopsies from the lesion were compatible with well-differentiated NET, grade 1 and Ki67 <3%. PET-CT rules out metastasis. Preoperative stage II and TNM classification T2N0M0 (2) was diagnosed. The patient was then, undergone to a Whipple cephalic duodenopancreatectomy with histological confirmation of the piece and absence of affected lymph nodes.

NETs represent 2% of overall gastrointestinal neoplasms. The ileum (41.8%), rectum (27.4%) and stomach (8.7%) are the most common locations (1). Only around 150 cases of NET originating in the ampulla of Vater have been described in the literature currently. The clinical presentation is jaundice (60%), abdominal pain (40%) and weight loss (10%); most of them are non-functioning without carcinoid syndrome (3). Duodenoscopy and abdominal CT/MRI are diagnostic image test. Routine echoendoscopy (EUS) is not indicated, but individualized assessment is recommended in case of small lesions (<1-2cm) for a better characterization of local invasion and UES guided biopsy if necessary (4).

The treatment remains under review. In small intestine of small size NETs, endoscopic resection is considering as a therapeutical option. However, the ampulla of Vater is a highly vascularized area with a risk of dissemination and need for surgical approach. Unlike carcinoid tumors of these ampullary locations, small size does not predict the absence of lymph node involvement, with up to 50% of cases (3,5), the literature shows that they metastasize in about half of the cases (5). Therefore, the curative treatment of choice for NETs is a Whipple surgery with lymphadenectomy, regardless the size and histological grade, although there are no updated official guidelines. Endoscopic or laparoscopic ampullectomy are recommended in cases of elder and high-risk surgical patients (3-5).



Figure



Figure 1. Complementary tests compatible with ampulloma A. Toracoabdominal CT: 15-mm solid nodule (arrow) at the level of the ampulla of Vater. B. Duodenoscopy: prominent and indurated duodenal papilla showing initial exteriorization of the tumor through the papillary orifice. C and D. EUS: homogeneous, well-defined round lesion that does not exceed 16x16 mm margins of the papillary wall, causing dilation in the common bile duct and Wirsung (shotgun barrel image) from the papillary area.





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