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MIXED NEUROENDOCRINE TUMOR OF HEAD OF PANCREAS

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Abstract: This report describes a rare case of a mixed neuroendocrine tumor of the head of the pancreas. The diagnosis was based on imaging studies with anatomopathological confirmation after a wide resection surgical procedure. This type of tumor must be included in the differential diagnoses in jaundiced and young patients, especially when other more common diseases have been ruled out.
Keywords: Neuroendocrine tumors. Chromogranin A.

INTRODUCTION

Neuroendocrine tumors are rare neoplasms responsible for less than 5% of those originating in the pancreas (1). It occurs at an annual incidence of 1 case per 100,000 people (2), but the incidence is higher in autopsy studies, ranging from 0.8 to 10%, suggesting underdiagnosis as they are asymptomatic in most affected patients (3). These tumors are classified according to clinical manifestations into functional and non-functional and according to histology in carcinoid tumors (well, little and moderately differentiated), mixed adenoneuroendocrine carcinomas and hyperplastic and pre-neoplastic lesions (4). About 50-60% are non-functional and 48% are located in the pancreatic tail (5). Several clinical syndromes are associated with these tumors, depending on the hormone produced in excess, in the case of functional ones (6). In non-functional patients, symptoms are associated with their dimensions and metastases (7). The diagnosis is histopathological and Chromogranin A is increased in up to 80% of the cases, regardless of whether they are functional or not (8), however imaging tests such as computed axial tomography and magnetic resonance imaging may be necessary (9). Surgery is the only treatment with potential for a cure (5).

CASE REPORT

Male patient, 34 years old, brown, was admitted to Acre Clinic Hospital from a city in the countryside with a history of epigastric abdominal pain with 3 months of evolution associated with progressive jaundice, choluria and weight loss of 15 kg in the period. He denied traumatic, alcoholism and smoking. On physical examination, he was emaciated, icteric (3+/4+) and without peripheral lymphadenopathy. The abdomen was flat, flaccid but painful on deep palpation in the epigastrium. Examinations on admission showed hyperbilirubinemia at the expense of direct, increased alkaline phosphatase and gamma-GT, negative serology for viral hepatitis, CA19-9 4448, Chromogranin A 51.7. She also had an abdominal ultrasound showing a slight increase in liver volume, hydropic gallbladder with biliary sludge and microlithiasis, normal intra and extrahepatic bile ducts and pancreas with increased volume in the head. Computed tomography of the abdomen showed an enlarged head of the pancreas with a nodular area measuring 4.9 cm with heterogeneous enhancement and another in the tail without enhancement after contrast with 2.5 cm, in addition to moderate intra- and extra-hepatic bile duct dilatation.

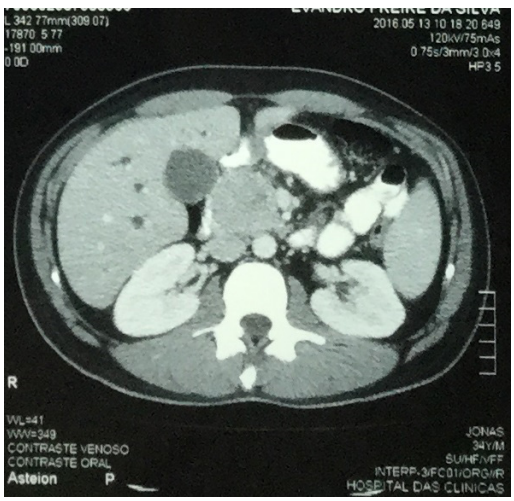

Figure 1. Computed tomography showing tumor lesion in the head of the pancreas

He was then submitted to exploratory laparotomy, which showed a tumor mass in the head of the pancreas measuring approximately 6 cm, without involvement of surrounding structures, and gastroduodenopancreatectomy was performed. The histopathological report reported acinar cell neoplasm with focal areas of neuroendocrine differentiation and the immunohistochemical mixed acinar-endocrine carcinoma. After successful postoperative recovery, adjuvant therapy with gemcitabine 1560mg was initiated. Patient is currently being followed up by the Asymptomatic Cancer Hospital.

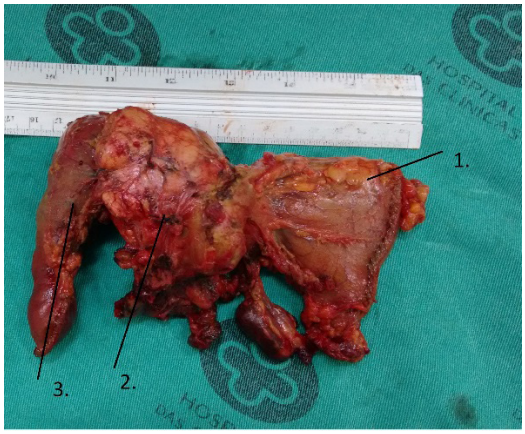

Figure 2. 1- gastric antrum; 2- tumor lesion in the head of the pancreas; duodenum.

DISCUSSION

Neuroendocrine tumors originate from neuroectodermal or pluripotent cells, have neurosecretory granules identified by electron microscopy or by immunohistochemical study (10) and, therefore, if they are functional tumors, they produce a specific clinical syndrome. The patient reported was admitted with symptoms not associated with functional tumors because the clinical picture was more associated with tumor growth and compression of neighboring structures. Regardless of this classification, studies show that 80% of patients have increased plasma chromogranin A, but in the report in question the levels were below the positivity value. The decision for extensive tumor resection by means of gastroduodenopancreatectomy was decided because the Pathology service of the Hospital das Clínicas do Acre does not have a frozen section biopsy or echo-endoscopy, which, respectively, would offer support for precise tissue resection or guided biopsy for better diagnostic elucidation and therapeutic planning before definitive surgery. Lymph node dissection is also recommended for all pancreatic neuroendocrine tumors (11), being performed in the reported patient D2 lymphadenectomy. Immunohistochemistry results showed a mixed tumor with an adenocarcinoma and neuroendocrine component, with the best option being the therapeutic complementation with chemotherapy (12). Ductal adenocarcinom are responsible for most pancreas tumors, being twenty-five times more common than neuroendocrine tumors, and therefore, it is necessary to distinguish between these two types, since neuroendocrine tumors have a better prognosis even in cases of metastatic disease (1,9). However, it is difficult to predict the prognosis of these patients because these tumors have a very variable biological behavior and clinical course (9).

CONCLUSION

The case report in question is important not only because of its rarity but because it illustrates an exceptional diagnosis in jaundiced patients, especially in this case in which the patient was young and all the most common differential diagnoses were ruled out. In addition, we observed that the propaedeutic limitation of the Clinic Hospital service precludes more conservative approaches due to the lack of more complex imaging and laboratory tests and also due to the absence of frozen section biopsy.

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