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Case Report

Neuroendocrine Stomach Cancer: A Rare Entity

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Abstract

Neuroendocrine tumors (NETs) are a group of neoplasms characterized by a variable biological behavior, with an average age at the time of diagnosis of 60 - 64 years. We report a very rare case of neuroendocrine tumor of the stomach, in a young and asymptomatic patient.

Keywords: *NET; MRI; Stomach; Somatostatin Receptor*

Abbreviation

NET(S): Neuroendocrine Tumor(S); US: Ultrasound Examination; MRI: Magnetic Resonance Imaging; TSE: Turbo Spin Echo; T2w: T2-Weighted Sequence; T1w: T1-Wighted Sequence; WHO: World Health Organization; CT: Computed Tomography

Introduction

Neuroendocrine tumors (NETs) are a heterogeneous group of neoplasms characterized by a variable biological behavior. In fact, the NETs can cause the typical symptoms of carcinoid syndrome (recurrent peptic disease, skin disorders, alteration of glucose metabolism, chronic diarrhea and flushing), but they can also be asymptomatic and discovered as an incidental finding [1]. They are less than 0.5% of all malignancies. The clinical incidence of NETs is estimated to be around 2/100,000 people/year for men and 2.4 for women [2]. In the last thirty years, there has been an increase in the number of new cases of neuroendocrine tumors and this figure is not due solely to improved diagnostic techniques and the spread of knowledge about this class of tumors, but also to a real increase of incidence of this pathology [2,3]. The average age at the time of diagnosis of NET, in the United States and Europe, is 60-64 years, while slightly higher in Taiwan [4,6,7]. The present paper reports a very rare case of neuroendocrine tumor of the stomach, in a young patient.

Case Report

A 21-year-old man came under our observation in December 2014 because he reported a suspected pancreatic mass detected by ultrasound (US). The patient was asymptomatic, it used to do regular US as follow-up for surgical correction of a right ureter malformation due to renal rotation. Until December 2015 there were no radiological signs indicating clinical remission for his surgical problem. In October 2014, an US was performed, evidencing not other abnormal findings except for a possible cyst-like over or peri-pancreatic mass.

2 months after the patient proceeded at our Radiology Unit to perform an abdomen Magnetic Resonance Imaging (MRI), with 3Tesla equipment, before and after intravenous administration of paramagnetic contrast medium; in particular, were made Turbo Spin Echo (TSE) T2-weighted (T2w) sequences on axial, sagittal and coronal planes sequences, TSE T2w with fat suppression, DWI and TSE T1w DIXON on axial plane, this last before and after intravenous administration of contrast medium.

MRI confirmed the presence of an intra-peritoneal epigastric mass, about 6 x 4 cm, bilobed, solid and fair CE after contrast medium, associated with some satellite lymph nodes associated with the presence of a gastric lesion vegetating along the greater curvature of stomach (Figure 1 and 2).

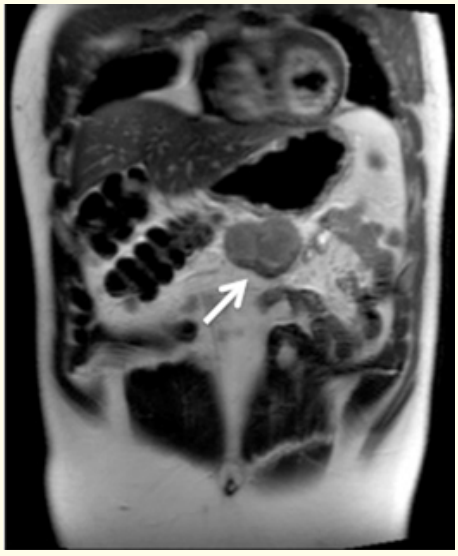

Figure 1: *MRI TSE T2 coronal images, showing an epigastric mass.*

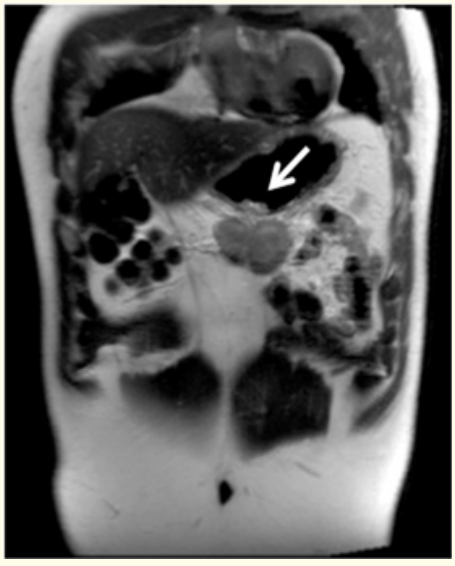

Figure 2: *MRI TSE T2 coronal images, showing a vegetating gastric lesion.*

MRI, through the use of various sequences allowed to evaluate the morpho-structural alterations of that gastric lesions which appeared hypo-intense on T1 and hyper-intense on T2 and showed inhomogeneous enhancement in dynamic phase of study.

In December 2014, the patient underwent a further esophagus-gastro-endoscopy (EGDS) examination that showed "the presence, in the middle third lower level of the great gastric curvature, of a nodular formation with central depression of 2.5 cm with regular margins. Esophagus revealed regular in caliber, course and mucosal surface. Hiatus hernia from slipping was discovered too. Normal gastric cavity. Pylorus patent. No injury in the bulb and in the second duodenal portion". Histological examination concluded for a low-grade neuroendocrine tumor (NET-G1 according WHO 2010) [5] with a follicular chronic active and micro-erosive gastritis.

A total body computed tomography (CT) examination was also performed, showing along the distal part of the greater gastric curvature in antral-pre-pylorica region, a vegetating lesion of about 5.8 x 3.0 cm with significative contrast enhancement and peri-lesional lymph nodes of up to 15 mm (Figure 3 and 4).

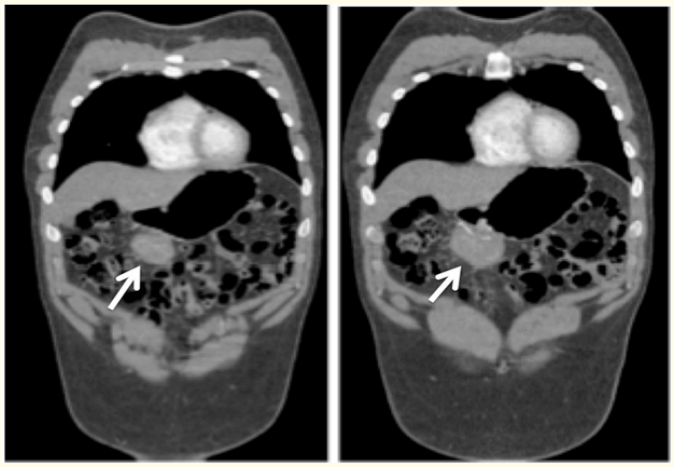

Figure 3: *CT scan, coronal images, showing along the distal end of the great gastric curvature in antral-pre-pylorica region, a vegetating lesion with contrast enhancement.*

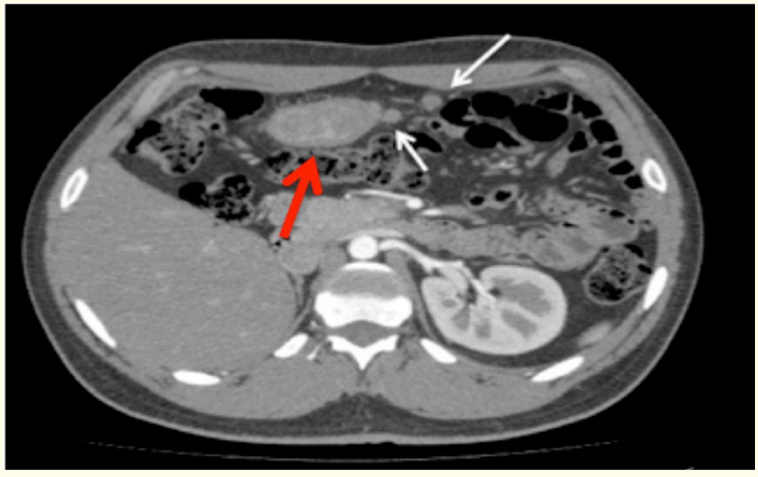

Figure 4: *CT scan: axial images, showing a vegetating lesion (red arrow) with associated local lymph nodes (white arrows).*

Blood tests performed on 30.12.2014 report the following values: HBsAg: negative; HCV: negative; AC-125 = 27; N.S.E Enolase Neur. Spec. = 10.84; Ca 19-9 GICA < 0.8; Blood type: A, RH Factor = positive; Fibrinogen 356; Dibucaine Number = 88; Prothrombin activity = 11.70, INR = 1.09; Partial thromboplastin time = 30.8; Sodium= 141 and Potassium = 4; Chromogranin A = 33.9.

The histopathological examination suggested the need to perform a SPECT/octreoscan to confirm the metabolic activity of the lesion and if there were other different localization of the disease. The SPECT highlights a single focal area of hyper-metabolic of the radiopharmaceutical accumulation, projected at the level of the gastric antrum, in correspondence of the note anamnestic pathology, evidencing the expression of somatostatin receptors presence.

The January 24, 2015 the patient undergoes subtotal gastrectomy for neuroendocrine cancer, with D2 lymphadenectomy and in addition to perigastric lymph nodes, an excision of hepatic hilum lymph nodes, of the celiac and the origin of the splenic artery was performed.

The histological report of the surgical specimen dated 02.04.2015 showed: tumor neuroendocrine (CK-pan, synaptophysin: positive) intermediate grade (NET-G2 sec. WHO 2010; proliferation index Ki-67: focally 5%; Mitosis 1/10HPF) infiltrating the tunica gastric submucosa with images of vascular embolization. Margins of gastric and duodenal resection free from neoplastic infiltration. Two lymph nodes of the big curve (the largest 4 cm) were metastatic among 16 examined (10 of the big curve and 6 of the small curve); fibro-histiocitary perinodal reaction. Omentum free from neoplastic infiltration. pT2 N1 sec. TNM VII and G2 R0.

Discussion

The NETs of the stomach account for about 5% of all neuroendocrine tumors of the gastro-entero-pancreatic district [8]. According to the clinical picture where they develop, NETs can be classified into three groups [9,10]:

1. Type 1 associated with a chronic atrophic gastritis;
2. Type 2 associated with type 1 Multiple Endocrine Neoplasia (MEN-1) syndrome;
3. Type 3 sporadic.

Beyond the genetic and pathological differences, the three categories mainly differ from the prognostic point of view. Type 1 constitutes 70 - 80% of gastric carcinoid and affects mainly women in the sixth and eighth decade of life. It can be considered almost a sort of benign tumor, associated, in very limited circumstances, with spread of metastases and for which a conservative approach is suggested [11,12].

Type 2 is developed in association with MEN-1. This disease affects equally men and women, typically during the fifth decade. Type 3, in contrast to the previous two forms, is not associated with chronic atrophic gastritis and has a predilection for male. It is associated with the presence of metastases in about 70% of cases. This form requires an aggressive treatment and a tight follow-up similar to those applied for the not endocrine forms. However, the poorly differentiated stomach NETs are very rare entity characterized by a high biological aggressiveness and poor prognosis. For NET, it is indicated regional staging and distance with Ultrasound, MSCT thorax, abdomen, pelvis or MRI [13].

We reported a case of stomach NETs, in a patient of 21-year-old, highlighted with US, MRI, CT and SPECT/octreoscan examination. In agreement with previous results, the patient undergoes to subtotal gastrectomy.

MRI and CT are two basilar methods of imaging in the staging, in order to choice of the best therapeutic strategy, providing important information on the response to the treatment, disease recurrence and prognosis of long-term survival. In our case, the concern has focused on the small gastric curvature and antrum. To date, our patient performs a follow-up with total body CT and full abdomen MRI with and without contrast medium, at intervals of six months, with no evident visceral recurrence of disease.

Conclusion

Gastric cancer accounts for the differential diagnosis with other benign and malignant formations of the stomach (polyps, soft tissue tumors, lymphoma, neuroendocrine tumors, sarcomas, GIST).

The accurate diagnosis and appropriate treatment of NETs currently gives in most patients a good prognosis, as long as it is discovered early. For this reason, patients with NETs should be managed in specialized centers, by an interdisciplinary team fundamental for the optimal patient care.

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