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Pancreaticoduodenectomy for Neuroendocrine Tumor of Duodenum- A Case Study

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Abstract

Neuroendocrine Neoplasm are a group of cancers that start in neuroendocrine cells. These cancers may also be referred to neuroendorin tumor, or even Carcinoids. Neuroendocrine Neoplasm occur when neuroendocrine cells stop working normally and start to grow or behave abnormally. Surgery has a large part in treatment because it is the only potentially curative therapeutic modality if resection can be complete. We have 2 types of Neuroendocrine Neoplasm; NETs (neuroendocrine tumours) are called 'well-differentiated' and tend to have a slow to moderate growth pattern and NECs (neuroendocrine carcinomas) - are called 'poorly differentiated' and tend to grow rapidly with significant metastatic potential. In this case we will study a duodenal neuroendocrine tumor simulating a neuroendocrine carcinoma at the beginning, an MRI was carried out revealing a duodenal neuroendocrine mass judged to be extirpable. Pancreaticoduodenectomy (PD) according to Whipple was made confirming the neuroendocrine nature.

1. Introduction

Cephalic duodeno-pancreatectomy (CPD) is a heavy and mutilating procedure, with postoperative mortality for trained teams in the range of 1% to 5% and morbidity of 30% to 40% related to post-operative pancreatic fistula in half cases. The two major indications for this intervention are tumors of the head of the pancreas, the ampullary region or the biliary duct however it finds its place in certain duodenopancreatic neuroendocrine tumors [1].

The endocrine cells present in the duodenal wall are at the origin of neuroendocrine neoplasms (NNE). NNEEs are a heterogeneous group of lesions with common anatomopathological characteristics.

Neuroendocrine duodenal tumours (TNE-D) represent between 2% and 8% of digestive TNE [2,3]. They correspond to 1%-3% of duodenal tumors and 11% of small intestinal tumours [4,5]. There is a slight male predominance [2]. Their discovery is mostly accidental (70%). Other modes of discovery are related to the tumor syndrome (20%) or genetic predisposition (10%) [6]. The vast majority of duodenal neuroendocrie tumors is non-metastatic at diagnosis [7,4]. They are localized in 75%-82% of cases, with lymph node invasion in 15%-20% of cases and with distant metastasis in 3%-5% of cases [8,9]. In 75% of cases, it is a single duodenal tumour and less than 2 cm in size. Their preferred location is the bulb (60%) followed by the Vater's bulb (20%). We present the case of a large neuroendocrine tumor occupying all the duodenal frame coming into intimate contact with all the organs of the neighborhood for which a Cephalic duodeno-pancreatectomy was performed.

2. Observation

A 50-year-old women, without any particular medical history, consulting for abdominal pain in her right hypochondrium. For 4 years the patient has complained of pain in the right hypochondrium that radiates towards the right shoulder with an intermittent upper gastrointestinal bleeding type melaena without other associated signs. Vital signs on presentation were within normal limits with an extreme mucocutaneous pallor.

Abdominal examination revealed a painful mass of the right hypochondrium. laboratory tests showed, hypochromic microcytic anemia with a hemoglobin at 6g/L. A contrast-enhanced computed tomography (CT) scan of the upper abdomen and abdominal magnetic resonance imaging (MRI) revealed : A sizable well-limited oval subhepatic heterogeneous tissue mass measuring 92 mm × 87 mm extended over 107 mm evoking a GIST: presenting intimate contact with the 2nd duodenum, the hepatic parenchyma and the head of the pancreas without a net fatty stripe of separation and with the portal vein and the upper pole of the right kidney with the presence of a fine greasy stripe. An esophagogastroduodenoscopy (EGD) revealed at the level of the 2nd duodenum ulcerobudding tumor formation of about 04 cm large axis reducing the duodenal lumen without obstructing it, the rest of the duodenal mucosa is healthy. Biopsies were made, the anatomopathological study of which returned to a poorly differentiated carcinoma Immunohistochemical examination revealed that the neoplastic cells strongly expressed chromogranin A. The Ki-67 index was approximately: 4% in the most proliferative tumor regions, the tumor cells did not express CD 117 Based on all the results, we made a diagnosis of primary neuroendocrine carcinoma of the second part of duodenum, and the histological grading is Glade 2. Disease staging was negative as well as the tumor markers CA19.9 ACE). The tumor is classified cT4N0M0, considered to be of limited «borderline» resectability and the therapeutic decision was to put the patient on induction chemotherapy (08 sessions based on Gemcitabine) followed by 30 cures of Somatostatin analogues (Somatulin tablets 120 mg 01 cures every 28 days) .Given the stability of mass measurements on the different CT monitors over the 4 years and the worsening of symptoms; the patient is discussed again in a Meeting of multidisciplinary hepatobiliary consultation in the presence of an expert radiologist who confirms the resectibility of the mass from which the decision for surgical treatment. The patient underwent a cephalic dudeno-pancreatectomy (Whipple) after laborious dissection and liberation of the mass (which had intimate contact with: the lower side of the liver and the hepato-duodenal ligament compressing backwards the inferior vena cava that was flattened down, with the right colic angle and the transverse colon, with the pancreas and with the portal vein and upper mesenteric axis). Pathological anatomy of the CPT-T piece came of neuroendocrine tumor (FIG. 1-3).

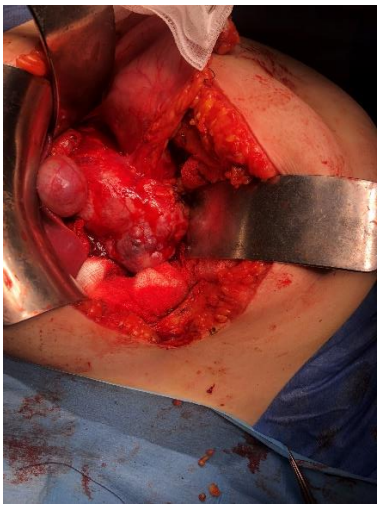

FIG. 1.

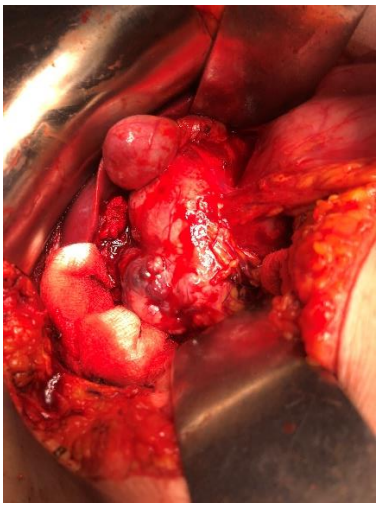

FIG. 2.

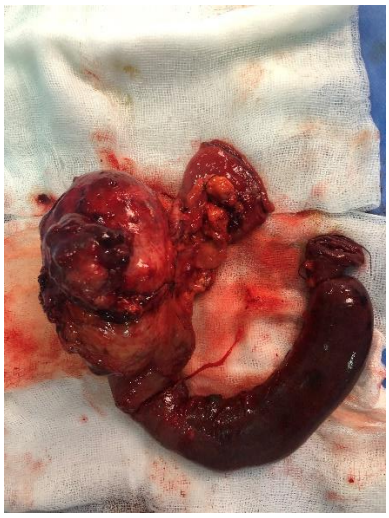

FIG. 3.

3. Discussion

Symptoms that may or may not include Neuroendocrine Cancer associated syndromes. (Syndrome is where 2 or more related symptoms occur). In Duodenal Neuroendocrine Cancer - you may hear the terms “Functioning” (meaning “with hormone related symptoms”) or “Non-functioning” (meaning “without hormone related symptoms”). Nb. Functioning or non-functioning may also be terms used to describe whether these cancers show up on nuclear medicine imaging* (see diagnostic tests). Many of Neuroendocrine Cancers of the Duodenum are non-functioning, meaning that they do not over produce hormones. Symptoms, if/when they occur, tend to be related to the size and / or position of the cancer and can include back pain, jaundice, stomach pain, nausea, vomiting and / or weight loss.

Functioning Duodenal Neuroendocrine Cancers can produce syndromes - due to producing too much of specific hormone. These include:
Gastrinoma (Gastrin): Zollinger-Ellison syndrome - acid reflux, heartburn, burping, stomach/chest pain, diarrhoea, anaemia
Somatostatinoma (Somatostatin): can cause symptoms of diabetes - like feeling tired, going to pee a lot, dry mouth, nausea, weight loss and anaemia. They may also cause diarrhoea and / or steatorrhoea (fatty pale stools).

A very small percentage may also produce symptoms of Carcinoid Syndrome: dry flushing of face and torso but can occur over whole body, palpitations, headache and blood pressure alterations, diarrhoea, asthmatic-like wheezing, weight loss or gain, malnutrition and / or fatigue.

Other, rarer symptoms, including Paraneoplastic syndrome and oncological emergencies, (a specific set of health concerns that can occur in any cancer), such as raised calcium levels (Hypercalcaemia), may occur.

Further information about Neuroendocrine Cancer associated and Paraneoplastic Syndromes - including Oncological emergencies - can be found duodenal neuroendocrine tumors are well encapsulated tumors that pushing neighboring structures without invading them and this is the advantage of these tumors compared to neuroendocrine carcinoma which infiltrates neighboring organs, which have a very poor prognosis and inextirpable in the majority of cases.

Surgical treatment has a central place because it is the only potentially curative treatment if the resection is complete [10]. Pancreatoduodenectomy (PD) is associated with the highest post-operative mortality among all pancreatic operations. The expected risk/benefit ratio has to be measured with extreme caution, particularly if the lesion is benign or of borderline malignancy [11-13].

4. Conclusion

We do not know exactly what causes NET; however, research is ongoing to truly understand both pathology and biology with the hope that this will lead to a much better understanding of the causes [14]. Pancreaticoduodenectomy (PD) according to Whipple is a major surgical procedure which finds its place in the treatment of the neuroendocrine tumor exceeding 2 cm.

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