

## Non-Functional Neuroendocrine Carcinoma of the Pancreas: about a Case at the Health Centre of Sikasso (Mali)

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### Summary

Neuroendocrine tumors of the pancreas are rare with an estimated incidence of 1 to 2 per 100000 inhabitants. We report the case of a non-functional neuroendocrine carcinoma of the pancreas in a 23-year-old patient, with no medical and surgical history, admitted to the Sikasso referral health center for a painful epigastric mass. Programmed for antropyloric tumor of the stomach or mesenteric, the intraoperative diagnosis was a tumor of the body of the pancreas of incidental discovery. Treatment consisted of tumor excision by enucleation of the tumor. Histopathological examination of the tumour revealed non-functional neuroendocrine carcinoma. The discovery of such a rare pathology in our health center motivated this work.

Our objective was to report this clinical case and to make a clarification within our health structure.

**Keywords:** Neuroendocrine carcinoma; non-functional; Pancreas; Sikasso Ref

### Introduction

Neuroendocrine tumors are defined by their structural proteins and hormonal secretion products common to neurons and all endocrine cells [1-4]. They have a common phenotype in immunohistochemistry [1].

A small number of these tumors (10 to 20%) secrete polypeptides responsible for a clinical syndrome of hormonal hypersecretion that allows them to be detected at an early stage.

According to [3], nearly 80% of these tumors are non-functional, because they are not accompanied by any clinical manifestation of hormonal hypersecretion and are revealed late by a locoregional tumor syndrome or by metastases.

We report the observation of a 23-year-old patient, with no medical and surgical history, operated on in our facility for an epigastric tumor syndrome with intraoperative discovery of a tumor of the pancreatic body whose anatomical and pathological examination of the room revealed a neuroendocrine carcinoma of the pancreas.

### *Patient et observation*

This is a 23-year-old patient, with no known medical-surgical history, multi-procedure who consulted in our structure for a painful epigastric mass.

The history of the disease dates back to about 1 year, marked by the onset of intermittent epigastric pain with the progressive appearance of an epigastric mass, accompanied by vomiting of food.

On general examination: WHO score = 1, BMI = 21.3 kg/m<sup>2</sup>, well-coloured conjunctivae and integuments, temperature = 37.2 C, Blood pressure = 110/80 mm Hg, Pulse = 106 beats/min, SPO<sub>2</sub> = 98%.

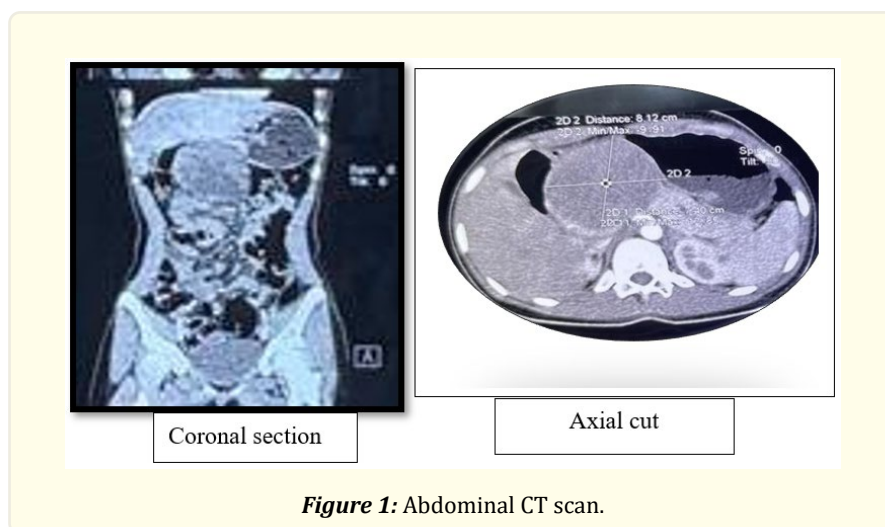
The physical examination found, on inspection, an epigastric arch, without scratching lesions or mucocutaneous jaundice.

Palpation revealed a painful, non-pulsatile epigastric mass of firm consistency, mobile in relation to the two planes and measuring 10 cm/6 cm of long axis. The lymph node areas were free, no Troisier's lymph node. The pre-hepatic dullness was preserved. Cardio-pulmonary auscultation was normal. The pelvic touches were normal, in particular. The rest of the examination was unremarkable.

We had put forward the following diagnostic hypotheses: mesenteric tumor and antropyloric tumor.

### *The requested FODG returned to normal*

The Thoraco-abdominopelvic computed tomography had objectified a voluminous epigastric mass 81x74 mm, of tissue content with heterogeneous enhancement after injection of the contrast medium and exerting a mass effect on the isthmus and the head of the pancreas of probably mesenteric origin. (See Figure 1).

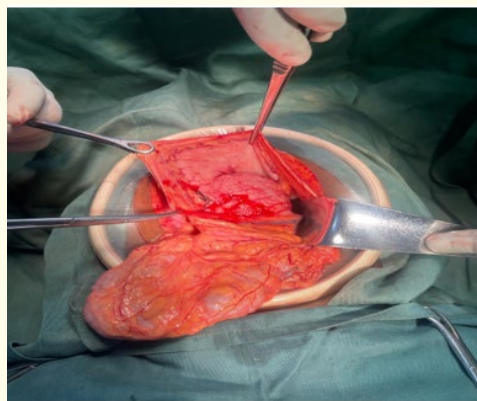


The patient was clustered O, rhesus positive, blood glucose at 0.90 g/l, serum creatinine at 0.82 mg/dl, PT at 95%, white blood cells at  $5.5 \times 10^3/\text{mm}^3$ , hemoglobin at 11 g/dl, platelets at  $336 \times 10^3/\text{ul}$ ,  $\alpha$ -fetoprotein level  $< 5 \text{ IU/ml}$ .

Anesthesia was of the general type with orotracheal intubation with provision of two units of blood iso group iso rhesus for mesenteric tumor.

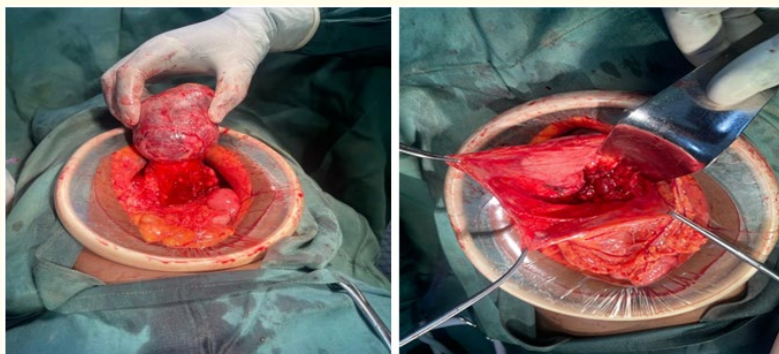
We performed a median laparotomy above and below umbilical.

On exploration, there was no ascites, the liver surface was smooth, no deep lymphadenopathy, no peritoneal carcinomatosis. The opening of the rear cavity of the omentums revealed a large tumor at the expense of the pancreatic body, without locoregional invasion (See Figure 2).



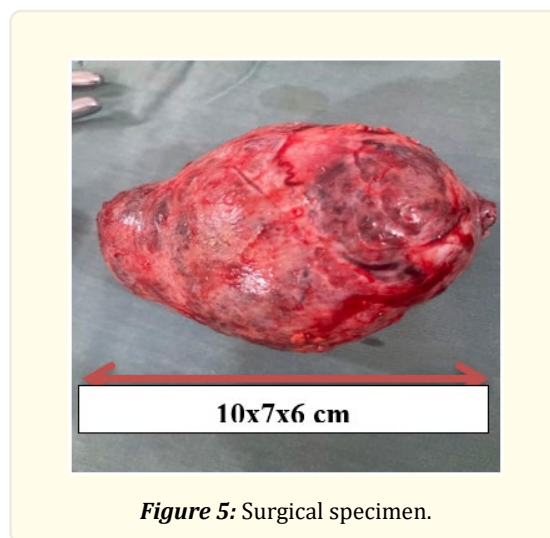
**Figure 2:** Opening of the rear cavity of the omentums.

The surgical procedure performed was tumor excision by enucleation and hemostasis control. (See Figure), peritoneal toilet with saline serum, a drain in the left parieto-colonic gutter and a drain in Douglass' posterior cul de sac and parietal closure. The immediate post-operative effects were simple.



**Figure 3, 4:** Tumor excision by enucleation and hemostasis control.

Immunohistochemistry of the operative specimen had objectified 'a synaptophysin-positive neuroendocrine carcinoma classified (T3; G3: WHO 2017) [6].



The case was discussed at the multidisciplinary postoperative symposium and a hormonal assessment was carried out, which was unremarkable. We had decided on clinical and biological monitoring.

The follow-up examinations at one month, three months, six months and then one year postoperatively had been unremarkable.

## Discussion

Neuroendocrine tumors of the pancreas are rare tumors with an estimated incidence of 1/100000 inhabitants [4, 5, 7]. The relative frequency of non-functional tumors of the endocrine pancreas has decreased markedly in recent years, due to the systematic search for hormonal polypeptides (normally secreted by the pancreas or ectopic) [3, 7, 12]. Their proportion fell from 41% to 15% between 1950 and 1981 in world literature [8]. As in our case, these tumors do not present a clinical translation related to hormonal secretion and are usually fortuitous discoveries at an often very advanced stage at the origin of a tumor syndrome associating cholestatic jaundice, abdominal pain, weight loss and vomiting or, more rarely, by palpation of an abdominal mass on clinical examination [3, 4]. Due to the increase in non-invasive radiological investigations of the abdominal area, non-functional endocrine tumors of the pancreas can be discovered incidentally during an ultrasound or a CT scan.

The computed tomography (CT) examination had been of very important contribution in our case by objectifying the tumor without specifying the pancreatic location. CT scans can be used to determine the size of the tumor and its location in the most common cases [3-5, 9] Magnetic resonance imaging (MRI) provides morphological information comparable to CT scans and makes it possible to evoke the diagnosis of TPE thanks to the hypo-intense T1 signal and the hyper-intense T2 signal of the tumor [5, 9].

Ultrasound-guided needle aspiration or CT scans confirm the endocrine nature of the pancreatic tumor. This precision is important in the presence of large tumors, with per-pancreatic (vascular) or metastatic extension which would constitute a contraindication to surgical excision in the case of exocrine tumor.

The treatment of endocrine tumors is multidisciplinary. Poorly differentiated endocrine tumors have a very poor prognosis and are treated with systemic chemotherapy. Excision surgery is the only curative treatment for well-differentiated endocrine tumors. Its objective is: to prolong survival by resecting the primary tumor and its possible lymph node and/or liver metastases; to control a possible

hormonal syndrome and to prevent or treat local complications [3, 4, 10].

The treatment of non-functional pancreatic endocrine tumors is surgical, allowing the cure of benign forms and prolonged remissions in the case of malignant and metastatic forms [3, 4]. In our case, we had proceeded to the enucleation of the tumor as described by some authors [3, 4, 9]. Histopathological examination of the operative specimen was in favor of neuroendocrine carcinoma of the pancreas.

The extent of resection of the primary tumor can range from a simple enucleation to a regulated pancreatectomy. This may be a cephalic duodenopancreatectomy "CPD", a caudal pancreatectomy sometimes associated with splenectomy (caudal pancreatectomy "SPC") or a segmental pancreatectomy (median or isthmic) [3, 11].

When the tumor is small in volume (diameter less than 5 cm), and limited to the pancreas, the doubt about its benignity requires a regulated resection: cephalic duodeno-pancreatectomy with pyloric, caudal or corporeo-caudal preservation according to the site of the tumor. In the case of tumors occupying the entire pancreas, total spleno-pancreatectomy associated with a regional lymph node dissection (coeliac, hepatic, superior mesenteric) is a rule [5, 3, 4, 9, 11]. In the case of a very large tumor with a corporeo-caudal site, a subtotal spleno-pancreatectomy is preferred, which will preserve the integrity of the duodenal framework and the main biliary voice with a non-tumoral pancreatic residue [4, 7, 10].

Medical treatment of these tumors has been proposed, either as an adjunct to surgical removal of malignant tumors, or as an isolated treatment in the presence of tumors considered unresectable [11]. Chemotherapy is mainly represented by streptozotocin alone or in combination with doxorubicin or 5-fluorouracil. Interferon has been proposed after chemotherapy has failed, or in combination with chemotherapy. The use of somatostatin has been more recently proposed. Despite response rates ranging from 10 to 50%, the effectiveness of these medical treatments has not been statistically demonstrated, but cases of paradoxical survival have been reported [12]. The immediate post-operative effects and at one month, six months and one year had been simple in our case. The follow-up after treatment is adapted to the histological type of the endocrine tumor. While poorly differentiated carcinomas require close monitoring every 2 to 3 months to assess the response to chemotherapy, well-differentiated endocrine tumors require spaced follow-up (tumors with a long doubling time, several years often being necessary to demonstrate slow progression).

## Conclusion

Non-functional neuroendocrine tumors of the pancreas are rare. Their diagnosis is most often intraoperatively in countries with a low medical technical platform. This should be considered in the presence of any epigastric mass and/or hypochondria with normal gastric fibroscopy. Imaging plays a key role in the localization of the primary tumor, making it possible to make a local and remote extension assessment, to ensure follow-up after treatment, to look for a syndrome of predisposition to tumors and to look for a second associated cancer.

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