A Rare Case of Adenocarcinoma of Heterotopic Pancreas

KALAJI Manhal*, DELAUNOIT Thierry and MANSVELT Baudouin

* Digestive surgery unit, Jolimont Hospital
1 Gastroenterology unit unit, Jolimont Hospital
2 Digestive surgery unit, Jolimont Hospital

ABSTRACT
Heterotopic pancreas is defined as the presence of pancreatic tissue in an unusual anatomical location that has neither anatomic nor vascular continuity with the pancreas. Ectopic pancreas adenocarcinoma is a rare form of ectopic pancreas.

This case study is about a 69-year-old man who was admitted to the emergency department (ED) with fecal vomiting lasting for the last two days. The investigations revealed an antro-pyloric mass. Biopsies performed at gastroscopy argued in favor of duodenal cancer, while those performed at endoscopic ultrasound argued in favor of a gastric tumor.

A surgery was therefore decided. During the operation, the decision was made to perform a subtotal gastrectomy because of the results of the extemporaneous analysis and the suspicion of a gastric cancer. The final histopathological report revealed an adenocarcinoma of an ectopic pancreas. Outcome and a literature review of the pathology, prognosis and treatment will be discussed.

Even if adenocarcinomas of heterotopic pancreas are rare, it should not be forgotten in the differential diagnosis of submucosal tumors. The standard treatment for ectopic pancreatic adenocarcinoma is surgery with a higher survival rate at 5 year compared to pancreas cancer. Frozen section analysis is essential.

Introduction
An ectopic pancreas, also called aberrant or heterotopic pancreas is defined as the presence of pancreatic tissue in an unusual anatomical location that has neither anatomic nor vascular continuity with the pancreas. It was first described in 1729 by Jean Schultz and later studied in 1970 by Potet and Ducleir [1].

The majority of patients are asymptomatic. The reported incidence in autopsy studies is 1 to 14% and it is encountered in one out of five hundred abdominal surgeries [2, 3]. This uncommon submucosal lesion is mostly found in upper gastrointestinal tract, such as duodenum (35%), stomach (30%) and jejunum (15%), but it can also be found in extra-abdominal location. Endocrine or exocrine pancreatic tissue or a combination of both cell types may be seen. Malignant transformation of the aberrant pancreas is very rare.

In this article we report a case of an ectopic pancreas adenocarcinoma developed in the stomach. Outcome and a literature review of the pathology, prognosis and treatment will be discussed.

Case
A 69-year-old man was rushed to the emergency department (ED) with fecal vomiting lasting for the past two days. The patient past medical history includes coronary bypass surgery a couple of month before, active smoking, high blood pressure, chronic renal failure, hypercholesterolemia and insulin-requiring diabetes.

The vomiting occurred night and day without abdominal pain. He also suffered for several months from eructation and burping. His appetite was surprisingly preserved.

The abdomen was soft and painless. No Blood analysis showed impaired renal function with creatinine levels at 1.8mg/dL and a C-reactive protein level at 19.1mg/L.

Due to his chronic renal failure, a non-contrast abdominal CT-scan was realized in the first instance and showed a distended stomach without any abnormality of the duodenum neither of the intestines.
However, an infrarenal abdominal aortic aneurysm with a diameter greater than three centimeter was discovered.

The patient was hospitalized for completion of investigations. He was nil by mouth with parenteral nutrition, rehydration, high-dose proton pump inhibitors (PPI) and a nasogastric tube.

An upper gastrointestinal series showed a severe stenosis of the pylorus with absence of gastric emptying.

A gastroscopy with biopsies was performed. The pyloric stenosis was crossed by the passage of the endoscope. No lesion or ulcer was visualized. The biopsies were analyzed in our hospital and duodenal neoplasia was the first hypothesis.

The echo-endoscopy indicated a high suspicion of antro-pyloric neoplasia. At this stage new biopsies were performed and were analyzed in an external laboratory that suspected a gastric tumor.

The tumor markers, Carcinoembryonic antigen (CEA) and Carbohydrate antigen 19-9 (CA 19.9) were both negative.

The PET-CT showed a very suspicious abdominal mass in the pyloric region (figure 1). It could indicate either a primitive cancer or a gastrointestinal stromal tumor (GIST). No abdominal or thoracic lymphadenopathy was seen.

Following this assessment, a cephalic duodenopancreatectomy was planned. However, we finally opted for a subtotal gastrectomy with a D2 lymphadenectomy without splenectomy because of the extemporaneous results (figure 2). A cholecystectomy was performed because of the adhesion.

The final histopathological analysis described an adenocarcinoma arising in an ectopic pancreas within the stomach wall (figure 3). Resection margins were free. Nine lymph nodes out of eighteen were positive at analysis. The cancer was classified as pT2N3Mx. Thereafter, he underwent chemotherapy postoperatively with six month of Gemcitabine.

In June 2018, he recurred in the form of pulmonary metastases. A new cycle of chemotherapy was started, with Gemcitabine and Abraxane (Protein-bound Paclitaxel). Two year later, in March 2020, the lung nodules were stable. The Abraxane was stopped due to stable lesions and the poorer patient tolerance.

Discussion

Ectopic pancreas, also known as aberrant or heterotopic pancreas was described for the first time in 1729 by Jean Schultz [4]. It is defined as anatomical variation or congenital abnormality in which pancreatic tissue has grown outside its normal location and without vascular or anatomical connections to the pancreas. Aberrant pancreas can be at any position in the abdominal cavity, but it is usually found in the upper gastrointestinal tract. His incidence is very low. Pancreatic rest is usually silent but it can be affected by the same complication as the pancreas itself. Malignant transformation is very rare but when it occurs, it is often as an adenocarcinoma. Only few cases have been reported in the literature.

Patients diagnosed with pancreatic rest are mostly asymptomatic, but they can present abdominal pain, bloating, gastrointestinal bleeding, intestinal obstruction or even pancreatitis.

Differential diagnosis is to be made between a gastric adenocarcinoma due to its location and a GIST, the lesion being submucosal [5]. A perioperative extemporaneous exam is necessary to exclude this diagnosis.

While heterotopic pancreas may be detected with an enhanced CT scan, if a submucosal lesion is noted on upper endoscopy, an endoscopic ultrasound with biopsy is strongly recommended for further evaluation, as small lesions can pass unnoticed on the CT scan [6]. The final diagnosis is reached by the histology.

Guillou and al [7] described three criteria that confirm the ectopic pancreatic origin of the adenocarcinoma and thus exclude a gastric origin. First, the tumor should be located in ectopic pancreatic tissue or in contact with it. Secondly it is necessary to see a histological transition zone between pancreatic structures and the carcinoma. Finally, non-neoplastic pancreatic tissue must contain at least fully developed acini and/or islets of Langerhans. These three criteria were respected in our case.

The prognosis of ectopic pancreas adenocarcinoma is generally favorable, as in the rare cases described in literature, it seemed

Figure 3: Microscopic view of the tumor

Well differentiated adenocarcinoma (G1) with a “pancreatic” phenotype in hematoxylin and eosin.

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to show a slightly higher survival rate than genuine pancreatic adenocarcinoma. This could be explained by the fact that adenocarcinoma of aberrant pancreas are more quickly symptomatic, especially when the lesion is greater than 1.5cm [8]. Our pyloric lesion measured 4x2x2 cm which could explain the symptoms.

The management strategy of pancreatic rest should be guided by symptomatology and by suspicion of malignancy. While asymptomatic lesions can be followed, even though some authors recommend resection, symptomatic or malignant lesions should be removed [9,10].

Endoscopic removal of submucosal lesions carries a risk of perforation and bleeding and should be performed carefully only when the mass is accessible [11]. In the other cases or in front of a malignant tumor a surgery is indicated. In our patient, the malignant nature of the lesion and the symptomatology requested a surgical approach. After acquiring the results of the frozen section, it was decided to perform a subtotal gastrectomy given the suspicion of a gastric cancer. The entire stomach could not be removed due to the patient complicated anatomy and the presence of many adhesions, but the resection margins make us feel totally comfortable.

On ectopic pancreas, it would seem that a limited resection with healthy margins is sufficient, but no recommendations could be established due to the rarity of such cases.

The histopathology results showed an adenocarcinoma arising in an ectopic pancreas. Resection margins were free, but nine lymph nodes out of eighteen were positive and the tumour was classified as pT2N3Mx. It was therefore decided to start adjuvant chemotherapy with Gemcitabine for six month, which is recommended in pancreatic cancer.

Thereafter, the patient was followed closely which allowed early detection of the lung metastasis. A new chemotherapy was performed.

Actually, four years after surgery, the patient still in a very good general state and the lung nodules are stable.

Conclusion
The incidence of ectopic pancreas is low and its diagnosis is difficult but it should be considered in the differential diagnosis of upper gastrointestinal submucosal lesions.

This clinical case illustrates the difficulty of establishing the diagnosis of such lesions.

The standard treatment for ectopic pancreatic adenocarcinoma is surgery with a higher survival rate at 5 year compared to pancreas cancer. Frozen section analysis is essential.

Conflicts of Interest
Authors declared no conflict of interest.

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References

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