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Pancreatic Neuroendocrine Tumor with Intraductal Growth

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Abstract

Pancreatic neuroendocrine tumors (PanNETs) rarely involve the main pancreatic duct. We report a case of 79-year-old-man, diagnosed with an incidental pancreatic cystic tumor. Abdominal CT scan and MRI were compatible with Main Duct Intraductal Papillary Mucinous Neoplasms (MD-IPMN) with "high-risk stigmata" of malignancy. Pancreaticoduodenectomy was performed. Intra-operative histological analysis showed a PanNET instead of an MD-IPMN. We review the literature and discuss this unusual presentation of PanNETs.

A rare case of PanNET with intra-ductal growth simulating a MD-IPMN was presented and discussed.

Keywords: Pancreatic neuroendocrine tumor; Intra-ductal growth; Intra-ductal papillary mucinous neoplasm; pancreatectomy

Introduction

Pancreatic neuroendocrine tumors (PanNETs) are uncommon pancreatic neoplasms, representing only 1-2% of those diagnosed [1]. PanNETs were originally thought to arise from the pancreatic islets of Langerhans however, recent evidence indicates that their origin is more likely from pluripotent stem cells present in the ductal epithelium [1,2]. PanNETs are classified as functioning or non-functioning, depending on their associated symptoms [1,3]. Diagnosis includes: evaluation of symptoms associated with classic endocrine syndromes, serum markers of neuroendocrine tumors (Chromogranin A), Computed Tomography (CT) or Magnetic Resonance Imaging (MRI) as well as Octreotide scanning (somatostatin receptor scintigraphy). Surgery is the first line therapy for patients with resectable disease [4].

Intra-ductal Papillary Mucinous Neoplasms (IPMN) are cystic tumors of the pancreas. IPMNs are classified into 3 groups: main duct IPMN (MD-IPMN), branch duct IPMN (BD-IPMN), and mixed type. IPMN detection is increasingly common owing to the expanding use of CT and MRI. These represent about 20% of all cystic pancreatic neoplasms and may progress to pancreatic cancer [5,6].

IPMN are more commonly seen in males at the 6th and 7th decade, while PanNETs have no gender predilection and are more frequent between the 3rd-6th decade.

Several guidelines have been published for the management of IPMN [6,7,8]. The International Association of Pancreatology (IAP) published The Fukuoka Guidelines in 2012 [6] and reviewed in 2017 [5]. Currently, these are the standard for diagnosis and management of IPMN.

The Fukuoka Consensus Guidelines define "high-risk stigmata" IPMN as follows: obstructive jaundice in a patient with a cystic lesion of the pancreatic head, enhancing solid component >5 mm within the cyst and MD size of >10 mm. In patients fit for surgery, with any of these criteria, resection should be performed without further testing [6]. The analysis of intra-operative frozen-section margin of the pancreas is mandatory, as this plays a role in the intra-operative management of IPMN [9,10].

The typical radiological features of IPMNs are the presence of single or multiple pancreatic cysts

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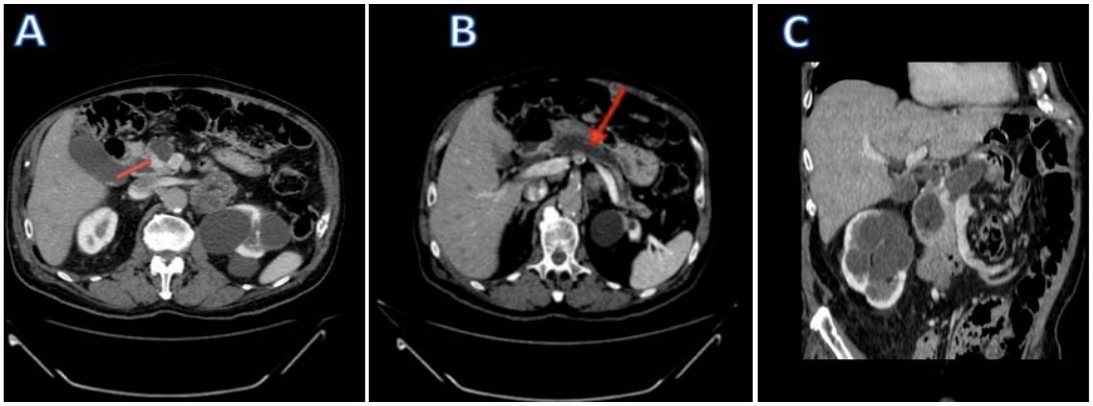

Figure 1: Abdominal CT scan revealed diffuse dilatation of the main pancreatic duct (20 mm) (Figure 1B); and a small hypercaptant lesion (15 mm) at the head of the pancreas (Figure 1A and C).

represented by hypodense lesions, dilatation of the main pancreatic duct and communication with the main pancreatic duct or it's branches. However, PanNETs usually appear on CT as circumscribed solid masses that tend to displace surrounding structures and are homogeneous with a high intake of contrast in the arterial phase (hyper-intense). Pancreatic lesions with an intra-ductal growth pattern other than IPMNs are rare. Intra-ductal growth of PanNETs has been published sporadically [11].

This report details an unusual case of a PanNET with intra-ductal growth that preoperatively mimicked a main duct IPMN.

Case Report

The patient was a 79-year-old-man, diagnosed with an incidental cystic pancreatic tumor. The lesion was found incidentally on ultrasound performed for symptoms of benign prostate disease.

Abdominal CT and Magnetic Resonance Imaging (MRI) demonstrated a diffuse dilatation of the MD (20 mm), with amputation of the MD at the level of the pancreatic head in line with a small hyper-captant lesion (15 mm) at the head of the pancreas suspicious of an enhancing mural nodule (Figure 1). The images were in keeping with MD-IPMN with high risk stigmata of malignancy. Tumour markers were normal: Ca 19.9 13 (<37 U/ml), CEA 0'8 (<4 ng/ml).

Pancreaticoduodenectomy with standard lymphadenectomy was performed [2].

Intra-operative histological analysis showed a PanNET rather than an IPMN (Figure 2). The final pathology report showed a well-differentiated non-functioning PanNET Grade 1 of 2.1 cm, with intra-ductal growth (pT2N0).

The patient was discharged from the hospital four weeks post-operatively. No recurrence was observed up to 2-years follow-up.

Discussion

The vast majority of PanNETs are sporadic, with a minority associated with familial syndromes [1,2]. Based on imaging, it is occasionally difficult to differentiate between pancreatic lesions with prominent intra-ductal growth patterns [11].

The World Health Organization (WHO) classification of PanNETs from 2017 distinguishes between well differentiated and poorly differentiated carcinomas. Grading well-differentiated PanNETs,

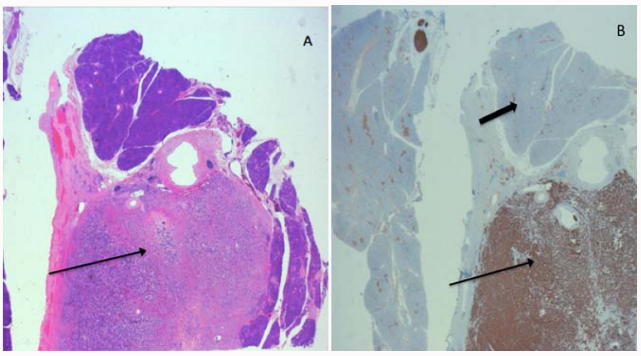

Figure 2: A: Hematoxylin-eosin (10X). Section of pancreatic parenchyma. An infiltrating tumour lesion is observed limited to pancreas with well defined contour (thin arrow) and B: In the immunohystochemical analysis (10X)tumor cell positivity is observed for CD56 (neuroendocrine marker) (thin arrow); in the normal pancreatic parenchyma positivity is observed in the islets of Langerhans (endocrine pancreas) and nerves (thick arrow).

into three grades (G1, G2, G3), based on proliferation assessed by mitotic count and Ki-67 index. Well-differentiated PanNETs(G3) are more aggressive than G1 or G2. Surgery is the treatment of choice for the majority of PanNETs, including this case. In selected cases, conservative treatment with active surveillance might be an option, although this is controversial [3].

Our case was discussed at the local multidisciplinary Hepato-Pancreato-Biliary meeting. Due to MPD dilatation of>10mm and the presence of a enhancing mural nodule, the preoperative diagnosis suspicion was of MD-IPMN with "high-risk stigmata" of malignancy. Resection was agreed following the revised IAP 2017 Fukuoka Consensus Guidelines, resection is usually carried out without further diagnostic tests. In this case it was felt that a negative biopsy on Endoscopic Ultrasound (EUS), would not have changed the surgical indication.

PanNETs with intra-ductal growth are unusual. Most of these cases are located in the distal pancreas and in the vast majority of cases the intra-ductal component, although prominent, is associated with a parenchymal component [11]. In this case it was a neuroendocrine tumor that resembled an IPMN preoperatively.

Conclusion

PanNETs should be kept in mind in the differential diagnosis of intra-ductal lesions owing to therapeutic implications, especially in

high risk patients. The main pancreatic duct can be dilated for a variety of reasons other than MD-IPMN. It is difficult to differentiate on imaging between different pancreatic lesions with a prominent intra-ductal growth pattern. Preoperative imaging although very helpful is not always reliable to fully elucidate the tumor and its extension along the gland. Intra-operative analysis of surgical specimen apart from frozen-section margin of the pancreas can avoid an unnecessary extension of the resection.

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