Solid Pseudopapillary Neoplasm of the Pancreas: A Rare and Unexpected Finding

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Abstract

Solid Pseudopapilary Neoplasm (SPN) are rare tumors usually appearing in young females. Presentations beyond 40 are rare. We describe the case of a 52 years old patient with persistent abdominal pain diagnosed with an SPN of the uncinated pancreatic process. The diagnostic between SPN and neuroendocrine tumor (NET) was challenging.

INTRODUCTION

SPNs are rare tumors comprising only 1 to 3% of all pancreatic tumors [1]. There is a clear female preponderance (95.2% in a 29 case series) of young age (from 24 to 39 years) [2]. SPNs were first described by Frantz in 1959 as pancreatic papillary cystic tumors [3] but the name SPN was finally defined in 2010 by WHO [4]. Usually it is a slowly growing tumor [2] incidentally found in imaging exams, not associated with major symptoms [5], although it can be detected as result of persistent unexplained upper abdominal pain [6]. Despite its gradual growth, SPN has the potential to expand significantly and generate compressive symptoms that manifest as jaundice or a palpable abdominal mass [6]. SPN do not present preferably in any specific part of the pancreas and size do not correlate with malignancy index [2]. Tumors located in the distal pancreas are more commonly larger [7]. SPNs are solitary [8] in most cases and encapsulated [2, 9].

Gold standard treatment is surgical resection by enucleation, distal pancreatectomy or pancreaticoduodenectomy [9] and definite diagnosis is obtained by histological evaluation of the specimen [10]. Solid sheets of tumor cells along with areas showing cells oriented around fibrovascular cores and periodic acid Schiff positive hyaline globules are typical histological findings [2, 11].

The majority of SPNs runs a benign course with an excellent prognosis [12] but large series documented cases of malignancy based on recurrence or metastasis in up to 22.8% cases [13, 14]. Due to its rarity, its diagnosis can be difficult and should be considered in the differential diagnosis of any solid or partly cystic pancreatic neoplasm, especially in women under 35 years of age [2].

The authors present a case of a 52 years old patient with complaints of recurrent upper abdominal pain that was diagnosed with SPN of the uncinated pancreatic process.

CASE PRESENTATION

A 52-year-old woman with no prior relevant medical history or chronic medication was referred to outpatient Gastroenterology (GE) consultation after going to the emergency department (ED) for recurrent abdominal

pain. In the previous five months, she noticed postprandial epigastric discomfort radiating to her back, nausea and bloating; she also linked the pain's escalation to stress and certain foods such as vegetables and fruit. In recent weeks, the pain has become more frequent and acute. The system review was otherwise unremarkable.

Observation reveals a soft, non-tender, and modestly swollen abdomen on physical examination, with no palpable lumps or audible bowel sounds. With the exception of gamma-glutamyltransferase (GGT) in the upper range of normal – 40U/L (34), laboratory tests were unremarkable. The results of the other liver tests, amylase and lipase, were normal. An emergency room CT scan revealed a heterogeneous 3cm mass of uncinate pancreatic process with calcifications; the pancreatic duct was not dilated (figure 1). She was prescribed simethicone and put on a FODMAPs diet before being referred to a GE consultation for further evaluation.

She experienced some symptom relief, with decreased stomach bloating and distention but the abdominal pain did not improve significantly. An endoscopic investigation, abdominal MRI and EUS were requested. Endoscopy of the upper gastrointestinal tract and colonoscopy were unremarkable. EUS revealed a 2.7 cm mixed well-delimited pancreatic uncinate mass with no e-flow (figure 2). The biliary and pancreatic ducts were not dilated. No swollen lymph nodes were found. Fine needle aspiration (FNA) was performed (22Gx1, from the second duodenal region, xanthochromic fluid). Endoscopic findings pointed to a neuroendocrine tumor with cystic degeneration. The smears were sparsely cellular with isolated cells positive for CKAE1/AE3 (dot-like) and CD56, and negative for chromogranin. Although it was insufficient for a conclusive diagnosis, the features suggested a neuroendocrine neoplasia/hyperplasia.

A 2.5cm well-circumscribed ovalated cystic mass with some septs was discovered in the uncinated process of the pancreas on abdominal MRI, with an unclear etiology (figure 3). Serum chromogranin was within normal limits. Ga-68 DOTANOC PET was ordinary. The case was discussed in Hepatobiliary and Pancreatic multidisciplinary meeting and surgical resection (pancreatoduodenectomy) was proposed.

The patient had a pancreatoduodenectomy and recovered well afterward. Histologicaly, it was a neoplasm with a solid component and pseudopapillary structures (figure 4). The cells expressed CD10, CD56, PR and B-catenin but were negative for CAM5.2, chromogranine and trypsin. These findings allowed a diagnosis of SPN. The patient was placed on a surveillance regimen and no additional treatment was given. She is currently asymptomatic six months after surgery.

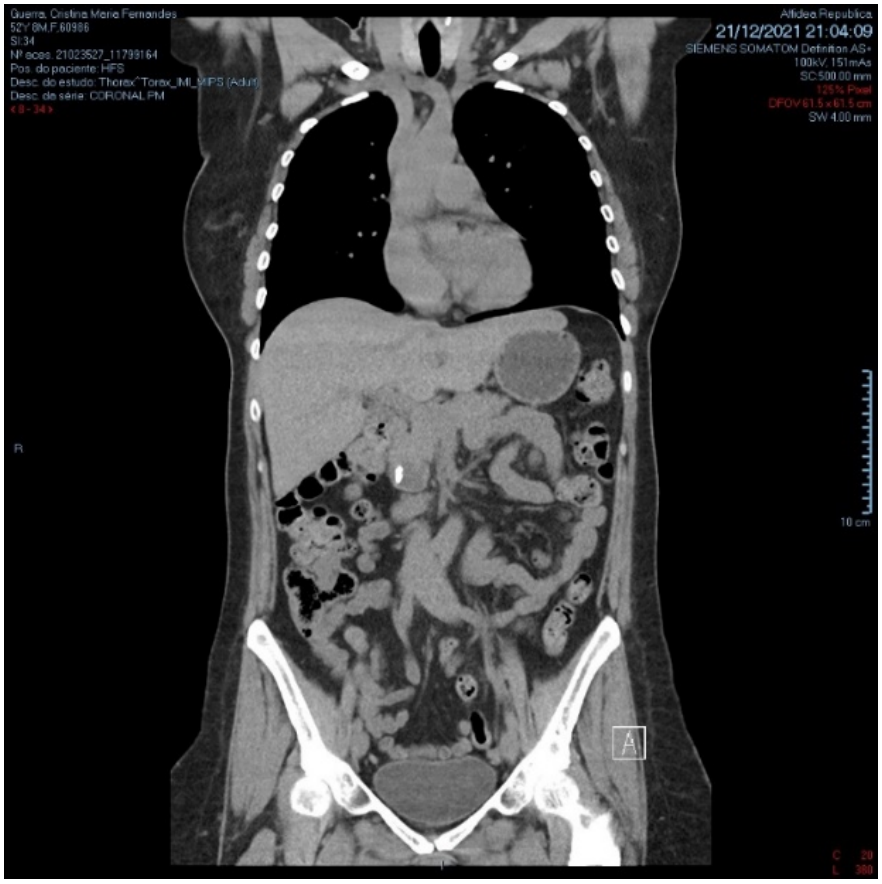

Fig. 1. Abdominal CT showing a heterogeneous 3cm mass of uncinate pancreatic process with calcifications; the pancreatic duct was not dilated.

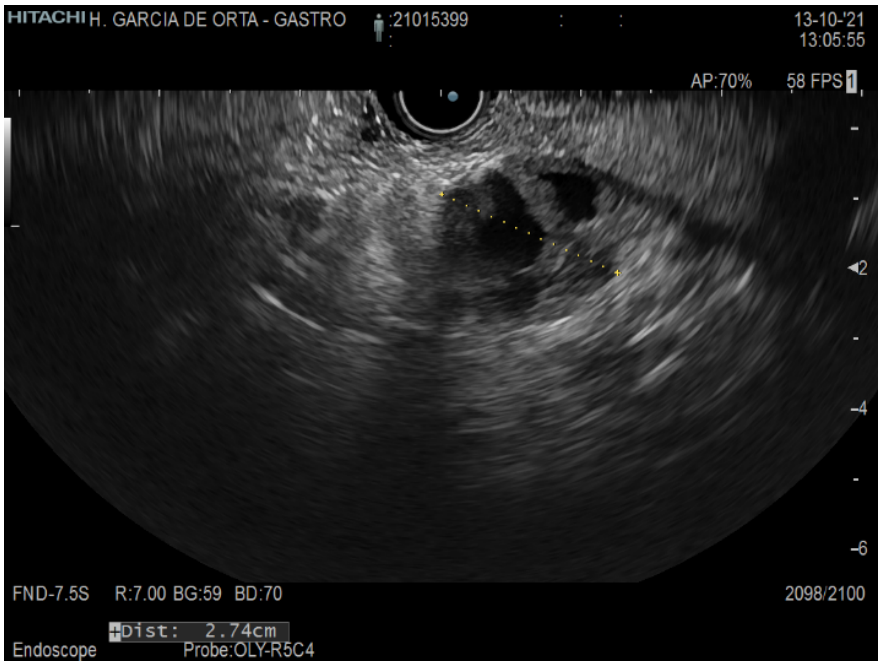

Fig. 2. EUS revealing a 2.7 cm mixed well-delimited pancreatic uncinate mass.

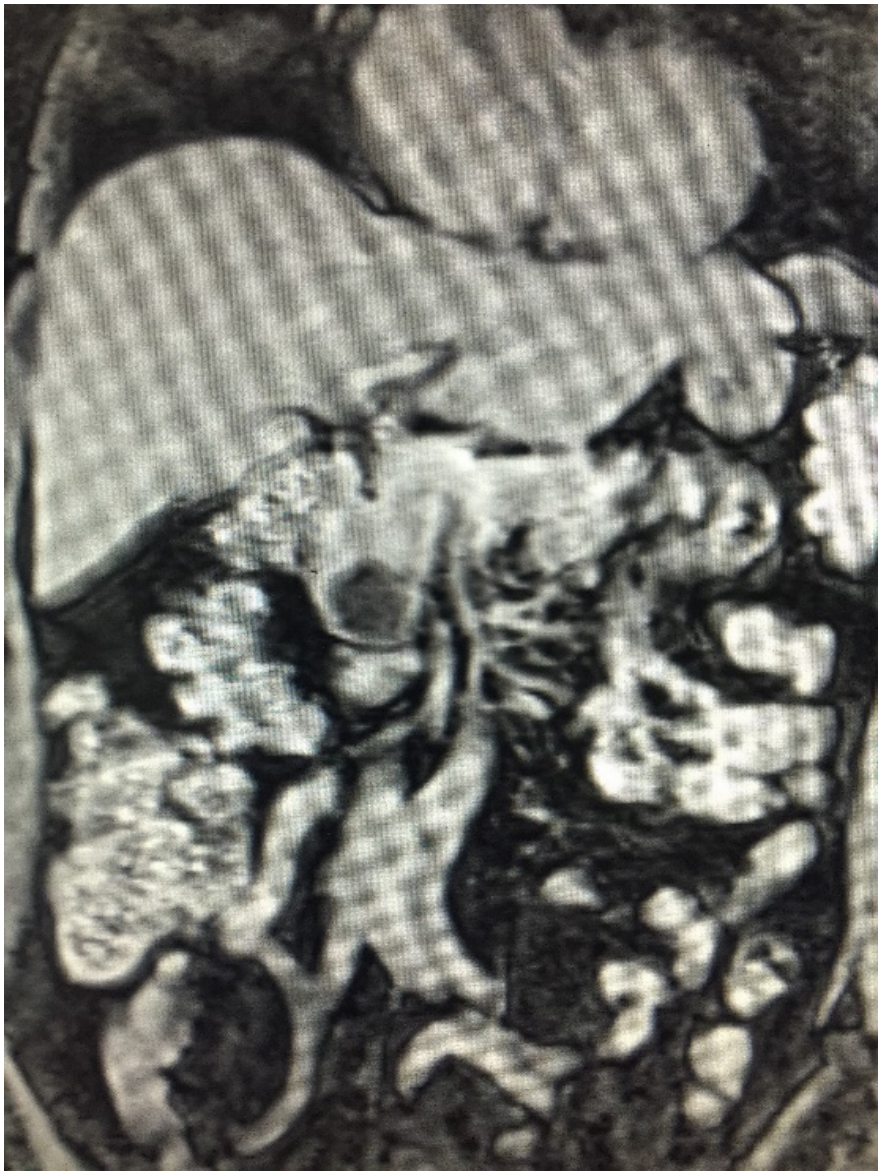

Fig. 3. Abdominal MRI showing a 2.5cm well-circumscribed ovalated cystic mass with some septs was discovered in the uncinated process of the pancreas.

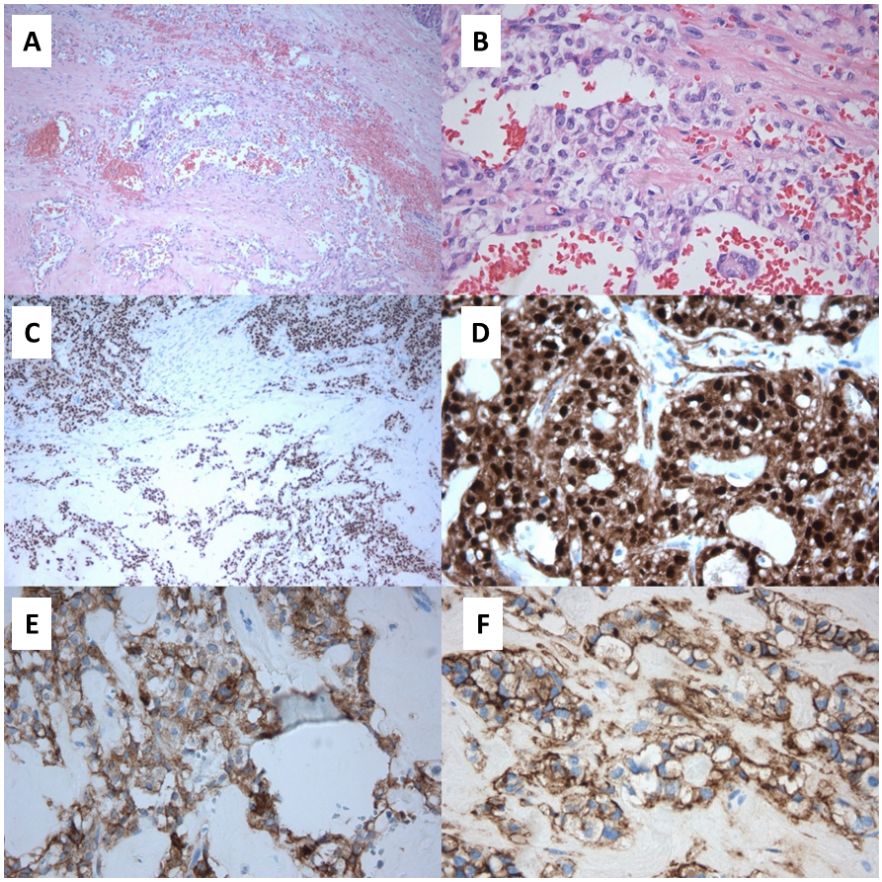

Fig.4. A mixed neoplasm with solid component and pseudopapillary core was discovered on histological examination. CD10, CD56, PR, and B-catenin were expressed, however CAM5.2, chromogranin and trypsin were not. A: HE 100x; B: HE 400x; C: PR 100x; D: B-catenin 400x; E: CD10 400x; F: CD56 400x.

DISCUSSION

Diagnosis of SPN of the pancreas is crucial because its prognosis is completely different from most pancreatic tumors with favorable long term outcomes [15]. In this case, a surgical specimen was needed before a definitive diagnosis of SPN could be made. This might be explained by the lack of pathognomonic clinical presentation with vague and mild symptoms in the beginning, without alarm features, mostly interpreted as functional dyspepsia, which might have delayed the diagnosis. Moreover, SPN are mostly found in female patients under 35 years, in a 29 case series under 20 years of age [2], and this patient was older. In fact, less than 10% of SPN cases have been reported in patients older than 40 years of age [16].

Also, in this case, EUS finding were not specific enough to define the diagnosis of SPN and NET with cystic degeneration was suspected. Pancreatic NET and SPN are frequently a diagnostic challenge [17] and definitive preoperative diagnoses are made in only a minority of cases of SPN [18]. In a large study that included 718 patients, only 52 patients (7%) received a confirmed preoperative diagnosis of SPN based on the findings of FNA [19]. Contrast enhanced EUS could be used to visualize the blood flow inside the tumor, however, in this case, the SPNs was hypovascular compared with the surrounding pancreatic parenchyma. Also, sparsely cellular smears with limited amount of cytological material obtained from fine-needle biopsies pose significant diagnostic challenges since the typical pseudopapillary structures are absent. Due to the partial overlap in immunohistochemistry in SPN and neuroendocrine neoplasm, a diagnosis of a neuroendocrine tumor is a potential pitfall in cytology smears with only isolated cells.

In this case, due to the large cystic component of the lesion, a FNA needle was chosen over a FNB needle. Probably, if a FNB was performed, a larger sample could have allowed a definite diagnosis. Song *et al*proposes some FNA cytomorphologic features that can help define the diagnosis, namely the presence of marked cellularity with pseudopapillary fragments composed of fibrovascular cores lined with one to several layers of tumor cells intermingled with discohesive neoplastic cells and inter- or intra-cellular pink hyaline globules, mucus-like globules surrounded by the stromal cells and cellular debris [20].

Surgical resection is often not difficult because is an encapsulated tumor and prognosis is excellent [2]. However, there are risk of metastasis (most commonly to the liver), or tumor recurrence, which is associated to large tumor size (more than 5 cm), lymphovascular invasion, metastasis to the regional lymph nodes, synchronous metastatic disease and positive resection margins [21]. Furthermore, close follow up of the patients after surgical resection is advised for early diagnosis of local recurrence and metastatic disease [18]. In our case, the patient is asymptomatic with no signs of metastatic disease or recurrence in the 6 months' post-operative follow up.

STATEMENT OF ETHICS

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

CONFLICT OF INTEREST STATEMENT

The authors have no conflicts of interest to declare.

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AUTHOR CONTRIBUTIONS

International Committee of Medical Journal Editors (ICMJE) criteria were used to define authorship. Each author was considered to fulfil these criteria.

Joana Rita Carvalho and Emanuel Vigia: substantial contributions to the conception of

the manuscript and interpretation of the case report; drafting the work and revising it critically for important intellectual content.

Marta Patita, Pedro Pinto Marques, António Figueiredo, and Catarina Lima Vieira: substantial contributions to the conception of the manuscript and acquisition of data for the work; critically revising the work for important intellectual content. All authors gave final approval of the version to be published and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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