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Solid pancreatic pseudopapillary tumor managed laparoscopically: A case report and review of the literature

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ABSTRACT

BACKGROUND: Solid pancreatic pseudopapillary tumors are a rare neoplasms, about 1–3% of all pancreatic neoplasms. This cancer mainly affects women between the third and fourth decade of life.

They are not well known; the molecular origins represent a low degree of malignancy, in which the complete resection is curative. We report our experience with a case report of SPT in a young man.

PRESENTATION OF CASE: Thirty-six years old male patient with a mass about 10 cm in the pancreatic tail and splenic ilum. After following CT and MR, the patient was subjected to surgery. Histophatological result was solid tumor pseudopapillary of pancreas with no pathological lymph nodes.

DISCUSSION AND CONCLUSION: Solid pseudopapillary neoplasm shows histological characteristic solid and pseudopapillary proliferation. Immunohistochemistry detects, among the causes of tumor development, a correlation between the Beta-catenin mutations, alteration of the E-cadherin. In the most cases, therapy is surgical treatment with laparoscopic.

1. Introduction

Solid pseudopapillary neoplasm of pancreas is a rare tumor, making up only 1%–3% of neoplasm of pancreas and his cells are characterized by poorly cohesive uniform cells solid and pseudopapillary growth pattern [1–3].

This tumor was first described in 1959 by Virginia Frantz as "papillary cystic tumor of the pancreas". The patient was a 2 years old boy undergone to pacreatic-duodenectomy [4]. Only in 1970 Hamoudi et al. described the ultrastructural features of the tumor as separate clinicopathological entity [5]. In 1996 its inclusion in the World Health Organization (WHO) classification as "solid pseudopapillary tumor" of the pancreas. The tumor has been given by different names but all to same histogenesis: solid and cystic tumor of the pancreas, papillary epithelial neoplasmos pancreas, papillary-cystic tumor adenocarcinoma of pancreas of childhood", and solid and papillary epithelial neoplasm [6].

The origin of this neoplasm is not well known but it has been postulates that this tumor maya rise from pluripotent embryonic cells of the pancreas or from the ridge ovarian analog-related cells, which were attached to the pancreatic tissue during early embryogenesis [7].

Untill today about 700 case has been described, more the two-thirds of them in the last years [9]. Probably this increase is due to better awareness clinicopathologic and radiographic features of solid pesudopapillary neoplasm associated to uniformity of the nomenclature. The source of the cells of SPT and tumorigenesis is still unknown. They usually affect young women with a female:male ratio of 10:1, between the third and the fourth decade of life. However, sporadic rare cases in men and in the elderly have also been reported [8]. This case report was conducted, and is reported in accordance with the SCARE criteria [15].

2. Case description

A 36-years-old man reported history of upper abdominal pain for about 4 months, associated to decrease appetite and weight loss. He had no comorbidy. All blood test and tumor markers were normal (CEA, Ca19.9, NSE, Cromogranina A). He performed an ultrasound examination showed bright liver; the gallbladder thick-walled appeared normal with evidence an endoluminal stone formation about 9 mm. Pancreatic morphology and echostructure was preserved. Wirsung was not dilated. Furthermore, the exam showed, in perisplenic a voluminous mass of 7.7 × 6.6 cm, heterogeneously hypoechoic with microcalcifications and irregular contours (Fig. 1a and b) Computerized tomography showed massive solid neoformation, next to splenic hilum, which incorporated the pancreatic tail and the splenic hilum, likely attributable swelling confluent lymph nodes. (Fig. 2a and b). The pancreatic tail was poorly defined, while morphology and density of the remaining parenchyma was normal. Preoperative MRI viewed between

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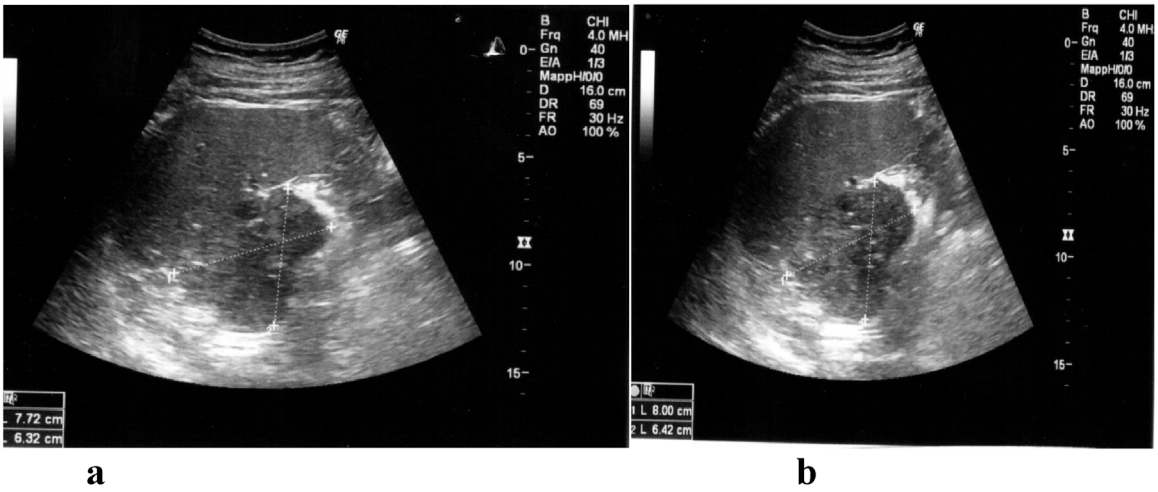

Fig. 1. a and b: US show an irregular and inhomogeneously hypoechoic mass with some calcifications inside.

pancreatic tail and spleen, an inhomogeneous formation of about 6.6 × 5.7 × 6.5 cm. This mass showed hyperintense signal on T2 W sequence (Fig. 3a and b), hypointense signal on T1W and progressive contrast enhancement (Fig. 4a and b), with some calcifications and colliquative areas. The tumor seemed poorly separable from medial profile of the spleen, the pancreatic tail, and from the splenic artery and vein and also compressed the ipsilateral kidney and adrenal gland. MRI concluded for an uncertain nature of the mass between lymph nodal mass or pancreatic tail tumor.

So we decide to perform laparoscopic approach as the first diagnostic and therapeutic act. It showed, on the retrocavity of omentum, a neoformation next to the splenic hilum, to about 10 cm in diameter that displaced the splenic vessels and infiltrated the tail of the pancreas. Then we run multiple biopsies and extemporaneous histological examination revealed epithelial neoplasm. Therefore we decided to proceed with a distal splenopancreasectomy.

2.1. Surgical technique

We performed a pneumoperitoneum open Veres-assisted. A 12-mm trocar was used 1 cm above the umbilicus to accommodate the standard 10-mm 30° laparoscope (T1). Two trocars, a 10-mm trocar was used at the left side in the left midclavicular line (T2), while a 5-mm trocar at right side (T3). An additional 5-mm trocars were used below xiphoid (T4) (Fig. 5). Gastro-colic ligament section and retrocavity of omentum access. Mobilised the splenic flexure of the colon and dissected the gastro-splenic ligament leads to the adrenal slot where the tumor appears no infiltrate the ipsilateral adrenal gland. Then we isolated the splenic artery to the upper margin of the pancreas and we dissected it between titanium clips. Isolation and section of the splenic vein between titanium clips. The next step was to pass under the pancreas and to dissect in disease-free area with Echelon Flex gold clad Seamguard. It then proceeded to dissect short gastric vessels and to mobilize the spleen from adhesions with the diaphragm on top, with the adrenal gland and the lower floor muscle infero- laterally, thus completing spleno distal pancreasectomy.

2.2. Histological examination

Macroscopic histological examination showed a mass of 8 × 6, 5 × 6 cm localized at the splenic hilum and in continuity with the pancreas, with immobilized congested vessels. To cut the neoplasm seemed very hard with some brittle portions and greyish color. Also the mass seemed to invade the splenic parenchyma. None of the 7 lymph nodes removed were malignant.

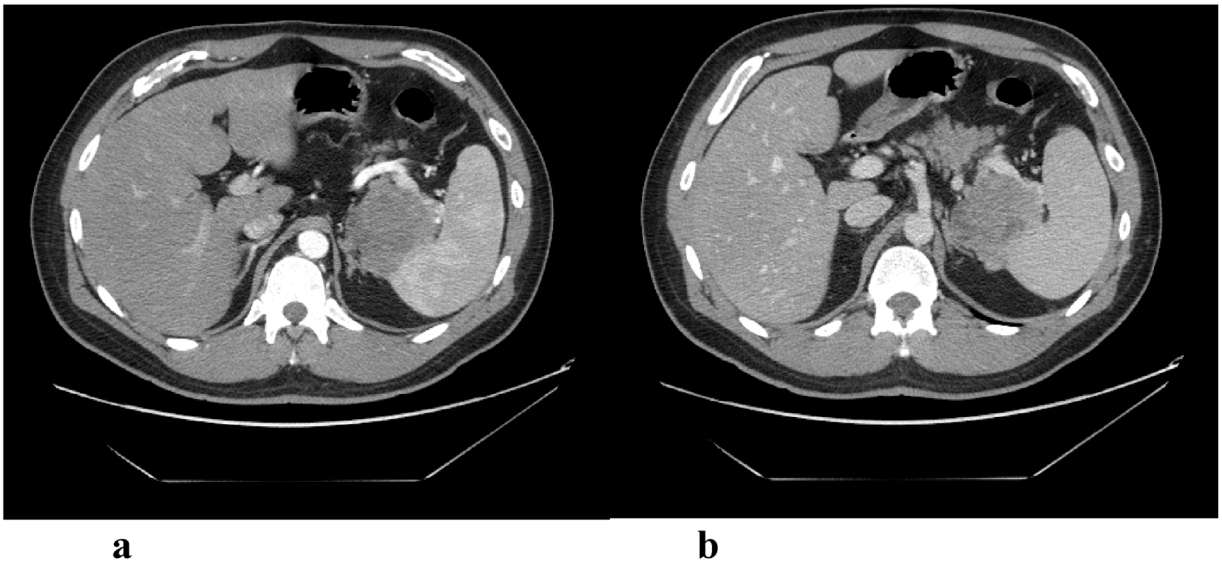

Fig. 2. a and b: CT with contrast enhancement during portal phase show a voluminous mass next to splenic hilum with splenic vein infiltration.

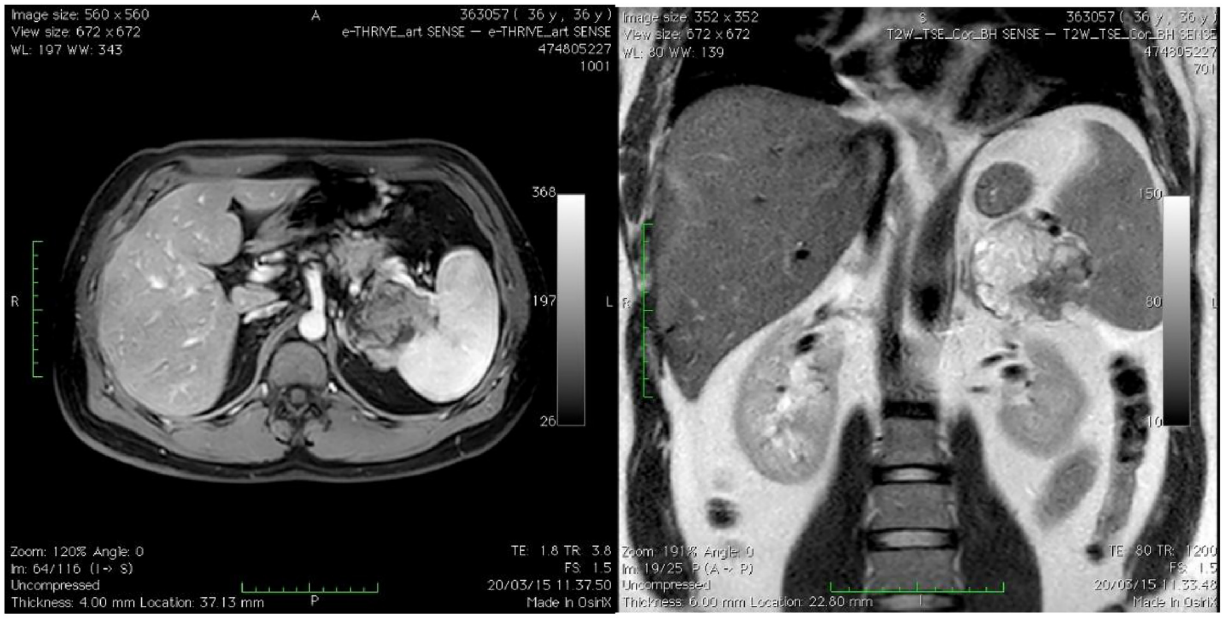

Fig. 3. T2 W TSE-BH sequence axial (a) and coronal (b) views show the inhomogenous neoformation, hyperintense in T2 W sequence with irregular internal septa and partially encapsulated.

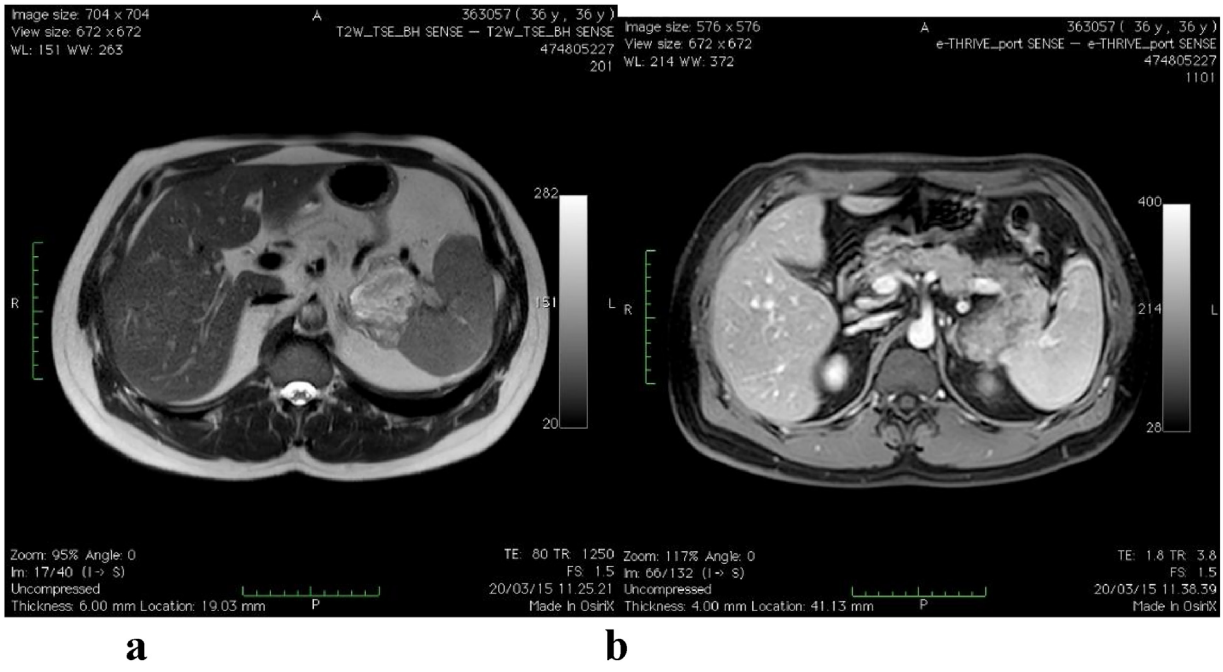

Fig. 4. a and b: THRIVE sequences: the images detect the presence of the mass in perisplenic site infiltrating splenic parenchyma and splenic hilum vessels.

Microscopic histological examination showed neoformation circumscribed by fibrous capsule and consisting of rather monomorphic proliferation of cells with eosinophilic cytoplasm weakly, clear nucleus with nucleolus and occasional small incisions. In the more points you observed cholesterol needles and aggregates istociti foamy. There are focally necrotic areas. The immunohistochemical staining show: CD56+, ß – Catenina+, CD10+, PROGESTERONE+, VIMENTINA+, CK–, Ca19.9–, CD99–, CD117–, CEA–, CK7–, CK20–, CROMOGRANINA–, DOG1–, FATTORE VIII–, INSULINA– Ki67 < 1%, GASTRINA–, ACTH–, CK8/18, CK19–, EMA–, GALECTINA 3–. Histophatological result was solid tumor pseudopapillary of pancreas with no pathological lymph nodes (Fig. 6).

The patient was discharged seven days after surgery. Histologic examination gave unexpected diagnosis of solid tumor pseudopapillary pancreatic node-negative and not infiltrated spleen.

3. Discussion

Solid pseudopapillary neoplasm of pancreas make up 1%–3% of pancreatic tumor [1]. It is more frequent in young women:

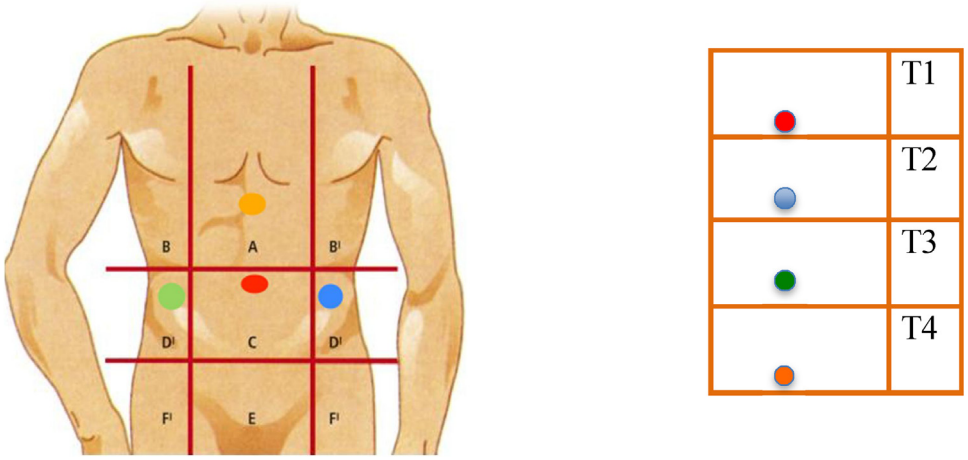

Fig. 5. T1 red; T2 blue; T3 green; T4 orange.

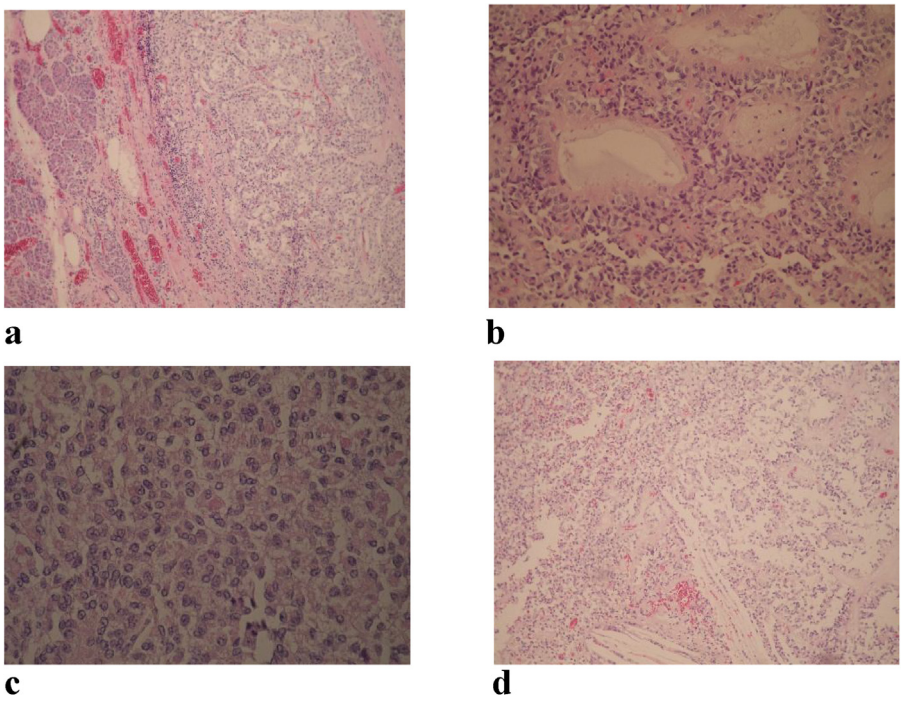

Fig. 6. (a–d): well-developed capsule circumscribes neoplasms from normal pancreas (a); very cellular neoplasm with monotunous lightly eosinofilic cells (b); cells with ovoid nuclei and little nucleoli and occasional hyaline globules (c); pseudopapillae and cholesterol clefts (d).

it affect 80% women and 85% the patients under 30 years old. Rarely it occurs in males or in pediatric people [9]. This neoplasm have a proceeding slow growing and low malignant potential. The common localization of solid pseudopapillary tumor is the tail of the pancreas, following by head and body with metastasis in only 10%–15% of cases. More rarely it can stand as multicentric tumor of the pancreas or extrapancreatic synchronous sites such as the retroperitoneum, duodenum, mesocolon, omentum and liver [10]. Its clinical presentation is nonspecific. The patients often are asymptomatic for a long time, until the tumor reaches a considerble size, because symptoms associated with tumor compression of adjacent organs. Some times the patient may have mild abdominal pain, nausea, poor appetite, loss of weight or palpable abdominal mass [11]. Due to its minimal symptoms it must resort to imaging techniques in order to make a diagnosis. Ultrasound and CT may help us diagnosis showing heterogeneous mass, with solid and cystic component, well- encapsulated. The MRI-scans show cystic degeneration, presence of a capsule and hemorrhagic or necrotic areas that can orient towards a diagnosis.

Histologically solid pseudopapillary neoplasm presents characteristic solid and pseudopapillary proliferation of homomorphous cell without increased mitoses or cytological atypia [12]. This neoplasm seems to originate from primitive pancreatic cells or from cell line of the female genital bud [13].

The presence of well- encapsulated with a mixture of solid, cystic and pseudopapillary patterns are pathognomonic of this disease. Often are visible irregular cystic and hemorrhagic.

Immunohistochemistry shows a possible correlation between the Beta-catenin mutations or alteration of the E-cadherin and tumor development. The expression of progesterone receptor and how often develops in women seem to indicate a hormone-dependent tumor.

The treatment of choice for solid tumors pseudo papillary pancreas, their view of low malignant potential, is surgical resection.

Pancreatic resection should be preferred to enucleation to prevent the risk of dissemination or pancreatic fistulas.

In 1990 it performed the first laparoscopic distal splenopancreasectomia. Today this technique is the gold standard, as countless advantages were found over the open technique: In fact, it was shown that the laparoscopic approach allows to see magnified images with a better view of the splenic vessels and preservation of the spleen with an equal morbidity than the open technique. The laparoscopic approach is seen to be less debilitating allowing a shorter period of hospitalization and better cosmetic results [14].

In our experience, patients with uncertain diagnosis of mass located between the tail of the pancreas and the splenic hilum underwent distal splenopancreasectomy. During the postoperative period the patient underwent clinical observation, blood tests, with dosage of serum amylase and lipase and drainage in the first, third and sixth day. The amylase wes always between normal range, while after an initial rise of amylases in the drainage of 563 U/L there was a normalization of values in silk postoperative day. There were no post-operative complications.

Conflicts of interest

None.

Sources of funding

None.

Ethical approval

Ethical approval was not required from my Institution for this case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Cuccurullo: Diego Study design.
Carbone Gabriele: Writing the paper.
Iovino Giuseppe Michele: data collection.
Fabozzi Massimiliano: data analysis.
De Rosa Ilaria: Histological examination interpretation.
Corcione Francesco: Study concept.

Registration of research studies

N/a.

Guarantor

Fabozzi Massimiliano.

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