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Case Report

Solid Pseudopapillary Tumor of the Pancreas: A Case Report

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Abstract

Introduction: Solid pseudo-papillary tumors of the pancreas (SPT) described in 1959 by Dr. Frantz, are tumors with low malignancy potential, representing 1 - 2% of exocrine tumors of the pancreas. They mainly effect young women [1,2]. They are characterized by atypical symptomatology, ranging from a simple gene to signs of compression or invasion of the pancreas or pancreatic duct, sometimes even revealing metastases or fortuitous discovery during imaging [3-6]. Surgical resection remains the only treatment for SPT. The type and extent of this resection depends on the location of the pancreas, size, and extent of the tumor. The prognosis is generally favorable with a long life without recurrence [2]. In this article, we report a case of a patient with a large SPT of the head of the pancreas that was surgically treated with a cephalic duodeno-pancreatectomy (CDP).

Observation: We report the case of a young woman operated for a pseudopapillary tumor of the head of the pancreas discovered incidentally during an ultrasound. The echoendoscopy (EE) in addition to the characteristics of the tumor allowed the realization of a puncture-biopsy; the histopathological study was in favor of a SPT of the pancreas. The patient underwent cephalic pancreaticoduodenectomy according to Whipple. The postoperative follow-up was simple except for a grade B pancreatic fistula.

Conclusion: Solid pseudo-papillary tumors of the pancreas are rare tumors. Treatment is surgical. Recurrences are rare and late, hence the need for long-term follow-up.

Keywords: *SPT of the Pancreas; Surgery; Recurrence; Pancreatic Fistula*

Introduction

Dr. Frantz [1] initially described solid tumors of the pancreas in 1959. In 1996 WHO defines them as "Solid Pseudopapillary Tumor" or SPT.

They are considered as tumors with low malignancy potential, representing 1 - 2% of exocrine tumors of the pancreas, and affecting especially young women, with a sex ratio 10:1 [2].

They are characterized by clinical latency and atypical symptomatology, ranging from a simple gene to signs of compression or invasion of the pancreas and pancreatic duct or even telltale metastases. They can sometimes be discovered incidentally during imaging.

If the principle of preservation of the pancreatic parenchyma is recommended, in some cases, pancreatic resection remains the only alternative to the treatment of SPT, the type and extent of this resection depends on the location on the pancreas, the size, the extent of the invasion of the pancreas and/or the pancreatic duct.

Resection may be simple enucleation, cephalic pancreatectomy with or without pyloric preservation, central pancreatectomy, or left pancreatectomy with or without splenic preservation.

Prognosis is generally favorable with a long life without recurrence; however, in 10 - 15% of cases, the presence of synchronous liver metastases occurs.

We report a case of a patient with a pseudopapillary tumor of the head of the pancreas whose surgical treatment consisted of CPD and triple montage according to Child.

Observation

A patient was a female aged 28 years, with a BMI of 31, and a history of hypothyroidism, with a pancreatic mass discovered incidentally during an abdominal ultrasound. She did not suffer from any clinical symptoms. CT scan objectified a tumor process of the largely necrotic (55 x 45 x 86 mm) oval pancreatic incus without signs of vascular infiltration (Figure 1a).

MRI: encapsulated cephalic pancreatic mass with solid, cystic and hemorrhagic component, measuring 78x52 mm heterogeneous in T1 and T2, hyper intense in diffusion, moderately raising heterogeneously, without signs of invasion or lymphadenopathy (Figure 1b).

Echoendoscopy (EES) in favor of a voluminous formation of the pancreatic incus not infiltrating the surrounding vascular structures, evoking a solid pseudo-papillary tumor coarsely oval well limited (72.3 x 44.3 mm), hypo-echogenic, heterogeneous by the presence within it of some calcifications. The pancreatic duct is not infiltrated. A transduodenal biopsy was performed, in favour of a SPT rather than a neuroendocrine tumour (Figure 1c).

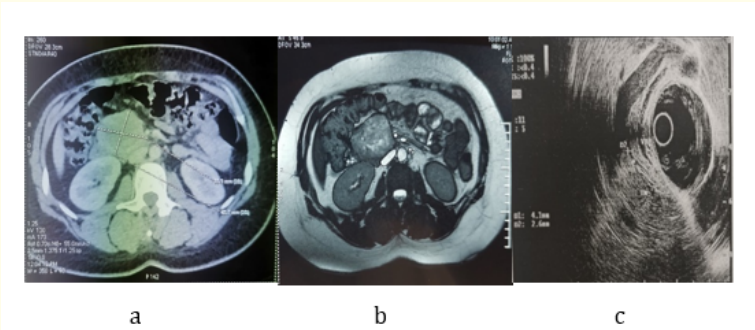

Figure 1: *a. CT scan; b. MRI; c. Echoendoscopy.*

Surgical exploration had found a tumor occupying the head and part of the body of the pancreas, infiltrating the pancreatic parenchyma and in intimate contact with the pylorus. This voluminous mass came into contact with mesenteric vessels without infiltrating them, however the existence of perigastric and mesenteric and even latero-aortic lymph nodes has been noted.

A cephalic pancreatectomy was performed according to Whipple and triple montage according to Child. In addition, we performed lymphadenectomy of the celiac trunk, portal and superior mesenteric vessels (Figure 2).

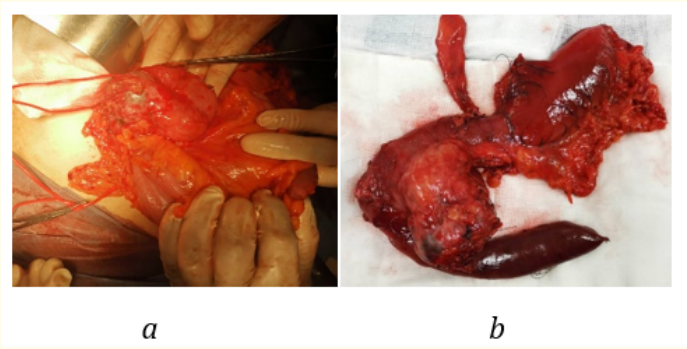

Figure 2: *a. Intraoperative aspect, b. Resection specimen.*

In the immediate postoperative period, the principle of early rehabilitation has been adopted. It consisted of the removal of the naso-gastric tube at the exit of the operating room, a wandering and resumption of early feeding (J1), anticoagulant and sondostatin.

The immediate consequences were simple. On day 5 the patient developed an infectious syndrome (fever at 39.5 c, dyspnea, and tachycardia). The determination of amylase in the drainage fluid (subhepatic drain) was in favor of pancreatic fistula.

The rest of the assessment (biological and radiological) returned without particularities, namely an inflammatory assessment (PCR), and the search for a possible Covid-19 infection (negative PCR). The CT scan found no abdominal collection or signs of release.

This alarming symptomatology was self-limiting and the patient was discharged on the 11th postoperative day, after removal of the drain.

On day 25 the patient came back for flow of a clear liquid through the opening of the drain, the radiological control did not objectify intra-abdominal effusion, it is indeed a pancreatic fistula whose evolution was favorable and completely dried up.

The pathological study of the resection part concludes a well-encapsulated tumor process of (90 x 60 x 50 mm), well limited adherent to the duodenal wall without infiltrating it, and located at 2.3 cm of the surgical boundary of soildo-cystic variegated appearance with necrotic-hemorrhagic rearrangements.

The microscopic study carried out on several levels of sections: pancreatic parenchyma seat of a tumor proliferation well limited and encapsulated epithelial nature of papillary architecture, pseudo-papillary, solid beaches with cystic rearrangements. Tumor cells are monomorphic, rounded with little cohesive eosinophilic or clear cytoplasm and round or oval nucleus with fine chromatin and rare mitosis. Immunohistochemistry in favor of a pseudo-papillary and solid tumor of the pancreas, Vimentin: intense and diffuse cytoplasmic positivate, AE1/AE3: negative. CD56: Moderate chromogranin membrane positivity: negative (Figure 3).

The young patient is seen in consultation regularly. At 26 months after surgery, the patient came back for a consultation and a liver ultrasound with images suspecting a secondary hepatic localization. Hence why we performed morphological examinations (ct scan and MRI) which came back without particularity.

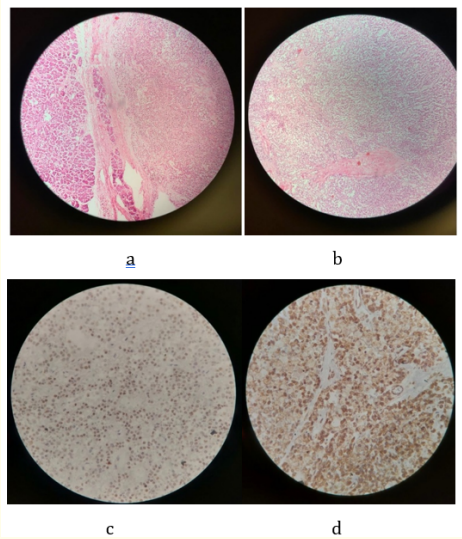

Figure 3: *a. Low magnification: tumor proliferation well limited by a fibrous pseudocapsule. b. Tumor architecture made of acini and pseudopapillary structures composed of monomorphic and slightly atypical cubic tumor cells. c. Progesterone receptors: nuclear labeling of tumor cells. d. Vimentin: cytoplasmic labeling of all tumor cells.*

Discussion

The solid tumors of the pancreas described in 1959 by Dr. Frantz, and defined in 1996 by the OMS as "solid pseudopapillary tumor" SPT. They are considered as tumors with low malignancy potential, representing 1 - 2% of exocrine tumors of the pancreas, and affecting young women, with a sex ratio 10:1 [1,2] and with an average age of 35 years old (extreme 13 to 63 yeas) [7].

Solid pseudo-papillary tumors of the pancreas are characterized by clinical latency, usually of incidental discovery during routine morphological examination (ultrasound) or as part of another digestive or gynecological pathology, vague transient abdominal pain; discomfort and sometimes-palpable mass rarely jaundice [3-6]. The tumors are characterized by slow evolution and low-grade malignancy. The prognosis after surgical resection is very favorable [5,6].

Pancreatic PSDs are usually located in the body of the pancreas, but they can be localized on all segments of the pancreas (head, body, tail) and can come in contact with the pancreatic duct. It is often the case of a large mixed tumor of cystic lesions and solid containing foci of necrosis and hemorrhage, often well encapsulated these tumors can nevertheless infiltrate the pancreatic parenchyma, the pancreatic duct and in some cases the neighboring organs.

Forms of contact with the pancreatic duct to differentiate from TIPMP [7].

Typically, on ultrasound SPT, the tumor appears well-circumscribed, mixed component solid, cystic and hemorrhagic hypoechoic and heterogeneous without dilation of the pancreatic duct. The CT scan specifies the location, size, and extent of the tumor process, necrosis, signs of infiltration of the pancreas and/or adjacent organs and the possible presence of synchronous metastases. On MRI, a heterogene-

ous low intensity signal in T1 and T2, hyperintense in diffusion, moderately heterogeneously and early enhancement of solid variegated appearance and pseudo-papillary cystic with necrotic and hemorrhagic rearrangements of variable proportion [4-6].

Resection may be simple enucleation, cephalic pancreatectomy with or without pyloric preservation, central pancreatectomy, or left pancreatectomy with or without splenic preservation.

If the principle of preservation of pancreatic parenchyma is recommended, in some cases, pancreatic resection remains the only alternative.

Small, well-encapsulated tumors can be treated with simple enucleation. The optimal surgical approach for tumors (large), is to perform a partial resection while respecting the principle of conservation of the pancreatic parenchyma (parenchymal sparing), the extent of this resection depends of course on the localization on the one hand and the possibility of infiltration of the pancreatic parenchyma, the pancreatic duct and/or other organs on the other hand.

When CPD is indicated in the cephalic localization, most authors prefer the preservation of the pylorus, especially when the tumor does not infiltrate the stomach and in the absence of perigastric lymphatic invasion [8].

When there is a doubt (morphological examination and surgical findings) on the possibility of infiltration of the pylorus and the suspicion of lymphatic infiltration, as is the case of our patient, to have a complete resection (R0) it is recommended to perform a cephalic pancreatectomy of Whipple without preservation of the pylorus with triple anastomoses, this resection can be enlarged en bloc to the others organs and/or vessels (portal vein, mesenteric vein) [8].

Routine lymphadenectomy is not recommended, but synchronous liver metastasis (SHM) resection is highly recommended [9].

In some cases the tumor is partially devoid of capsule and directly infiltrates the pancreatic parenchyma, it is not uncommon for the main duct to be incorporated into the capsule [10].

The possibility of infiltration of the capsule and/or pancreatic duct must be taken into account and can determine which type of resection to use, however this infiltration is not easy to highlight by preoperative morphological explorations.

Enucleation may be associated with a risk of fistulas. In forms with suspicion of infiltration of the capsule and/or pancreatic duct, it is recommended to perform a pancreatectomy [8]. As is the case of our young patient, operated for a SPT of the pancreas in whom it was performed a cephalic pancreaticoduodenectomy without preservation of the pylorus.

Lymph metastases are rare in PTS. In a study with more than 1700 patients with PTS, lymph metastases were found in only 5 cases so routine lymphadenectomy is not recommended [8].

However, lymphadenectomy is recommended when intraoperative suspicion of lymph metastases. Complete block resection with removal of suspected lymph nodes and nodal dissections is recommended [11].

Vascular extension and infiltration (portal vein, mesenteric vein), peri-nervous sheathing and infiltration of neighboring organs are in favor of malignancy and therefore monobloc resection remains the only chance to have an R0 resection [12].

Resection of synchronous liver metastases (SLM) is highly recommended, for some authors the prognosis is favorable with aggressive surgical resection, especially for related metastases [9,10].

Recurrences are rare and late. They may be local or give liver metastases and therefore require long-term follow-up. Several clinical factors could predict recurrence, the presence of metastases at the time of the first operation, tumor rupture during manipulation or invasion of adjacent organs.

The presence of larger tumors (diameter > 5 cm), renal invasion, lymph node metastases, positive margins and multiple organ resection were significant factors in locoregional recurrence and cancer-related survival [13,14]. In a review and analysis of 1897 patients operated for PTS, 2% developed a recurrence in 72% of cases; it was a distant recurrence [14].

Conclusion

Solid pseudo-papillary tumors of the pancreas are rare tumors. Treatment is surgical, recurrences are rare and late, hence the need for long-term follow-up.

Contribution

Dr. Meriem Rayen Lamara (Ph.D.), Peterborough, UK.

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