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Pseudopapillary and Solid Tumours of the Pancreas: Series of Five Cases

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Abstract: Case Report

Pseudopapillary and solid tumors of the pancreas (PPSTP), also called FRANTZ tumor, are rare tumors, occur especially in young women, and etiopathogenesis still uncertain. Their local and distant extension is rare, and their prognosis remains good especially after complete excision. Their risk of degeneration is 10 to 20%. We report the cases of a 5 pseudo-papillary and solid tumor of the pancreas in five women with no notable history.

Keywords: Pseudo-papillary and solid tumor, pancreas.

INTRODUCTION

Pseudo-papillary and solid tumors of the pancreas are a rare pathological entity and account for 1 to 2% of pancreatic tumor lesions. First described by Frantz [1] in 1959. They develop preferentially in young women and adolescents, in the form of a voluminous tumor with few symptoms for a long time, without alteration of the general state. The diagnosis is usually incidental or following abdominal pain. There is no typical imaging or specific tumor markers. The risk of degeneration is 10 to 20%.

In this paper we report the case of five patient with a pseudo-papillary and solid tumor of the pancreas; revealed by different clinical signs.

Patient and Observation

Case 1:

A 25-year-old female patient, who presented with abdominal pain in the epigastric region and in the right hypochondrium with a type of heaviness, evolving for 1 year and a half, without notion of epigastric pain with dorsal irradiations, without notion of antalgic position, without notion of a syndrome of cholestasis.

In whom the clinical examination objective a young healthy woman, with a WHO score 1 and a BMI of 24.6kg/m2. The abdominal palpation finds a mass at the level of the epigastrium, slightly sensitive, of hard consistency, fixed in relation to the deep plane, without modification of the skin in front.

An ultrasound scan was performed, showing a caudal pancreatic mass of about 10X12 cm. This was completed by a CT scan, which showed: a large pancreatic process at corporal-caudal level, with lobulated contours, heterogeneously enhanced with cystic areas, with small peripheral nodular calcifications, measuring 113x85X120mm.

This process was in intimate contact with the great gastric curvature and the anterior face of the left kidney, the tumor is distant from the celiac and superior mesenteric vessels, it displaces the splenic hilum with loss of the separating fatty interface in some places (Figure 1).

The parenchyma of the pancreas head is without anomaly. Biological workup revealing no abnormalities, including tumor markers.

The patient underwent a midline operation with a huge mass of about 20 cm long at the corporal-caudal level; solid-cystic in nature with a purplish wall.

The tumor invades the transverse mesocolon and the left 1/3 of the transverse colon and the right colonic angle without surgical cleavage plane, it also invades the splenic hilum without surgical cleavage plane.

It also invades the splenic hilum without any surgical cleavage plane. It comes into contact with the upper pole of the left kidney and the greater curvature of the stomach without invading, and also comes into contact with the lesser omentum with respect to the vascular structures.

The surgical procedure consisted of a caudal spleno-pancreatectomy with resection of the left 1/3 of the transverse colon, ACG and the lumbar colon, with manual colo-colic anastomosis (Figure 2).

The postoperative follow-up was without abnormalities, feeding was authorized on the third postoperative day, and the patient was declared discharged on the sixth day with a prescription for antistreptococcal antibiotic prophylaxis with penicillin M, and anti-pneumococcal, anti-Haemophilus influenzae, anti-meningococcal, and anti-influenza vaccinations on the fifteenth day of her surgery.

An anatomopathological study of the surgical specimen was made with the result: a morphological aspect in favor of a solid pseudo-papillary neoplasm of the pancreas of 16 cm infiltrating the mesocolon opposite and the splenic meso opposite as well as the pedicle and the splenic hilum, without infiltration of the colonic wall, with a limit of pancreatic resection passing in healthy tissue. With a 0N+/18 N lymph node dissection (Figure 3).

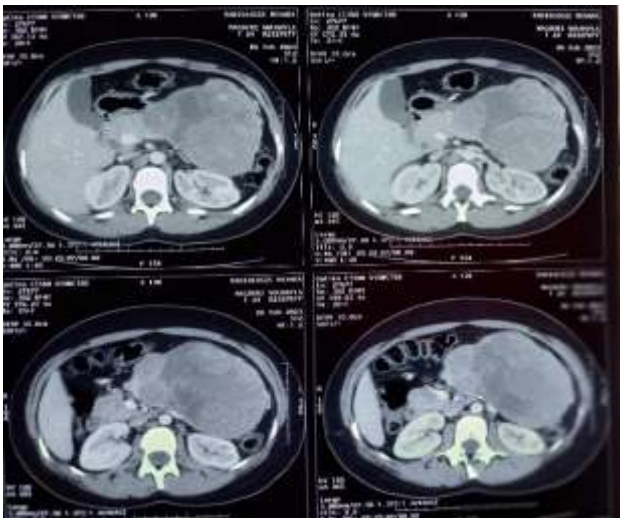

Fig 1: Pseudo-papillary and solid tumor of the pancreas with peripheral calcifications

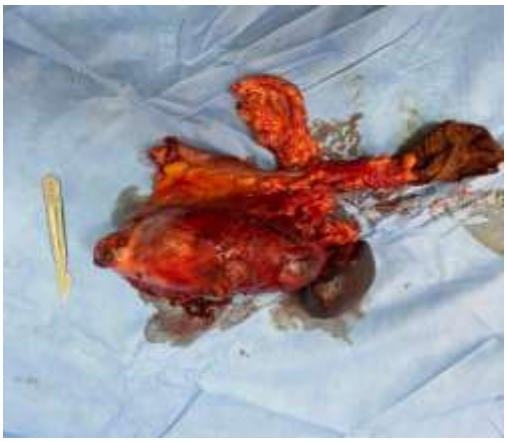

Fig 2: Image of caudal splenopancreatectomy specimen with monobloc transverse colon resection

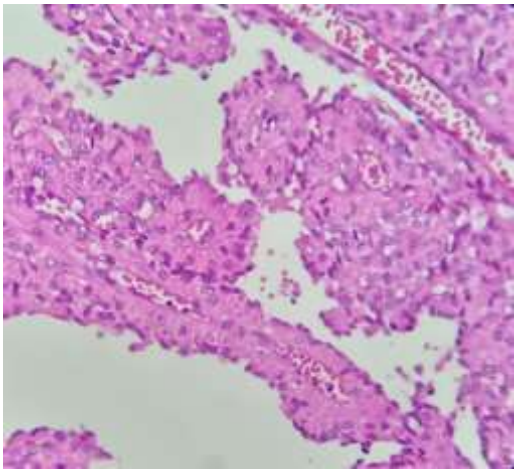

Figure 3: Histological image of the tumour mass

Case 2

A 22-year-old girl who complained of abdominal pain in the left hypochondrium, in a context of asthenia and altered general condition for 4 months. Abdominal examination revealed sensitivity and empathy in the left hypochondria.

Abdominal scan showed swelling of the left flank measuring 66 x 77 mm hypodense; heterogeneously enhanced by the contrast agent. Having a regular wall, coming into contact with the stomach, kidney, splenic pedicle, and pancreatic tail; conculating to gastric GIST pushing back the pancreatic tail (Figure 4). Gastric fibroscopy was without abnormality.

Surgical examination showed a solido-cystic mass of the tail of the pancreas in contact with the splenic vein and transverse mesocolon in accordance with the cleavage plan.

The surgical procedure consisted of a caudal pancreatectomy, carrying the tumor with splenic preservation (Figure 5).

The postoperative follow-up was without anomaly and the patient was discharged on the 5th day the anatomopathological and immunohistochemical study concluded that it was a pseudopapillary and solid tumour of the pancreas thail.

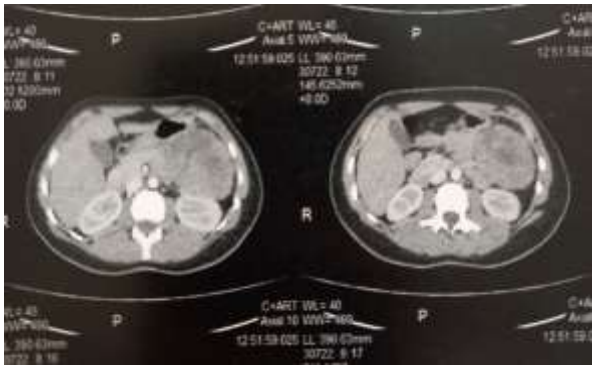

Figure 4: Solidocystic mass of the left flank

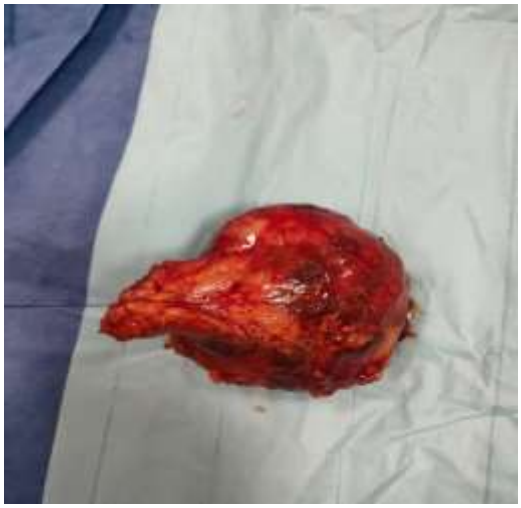

Figure 5: image of caudal pancreatectomy (spotted with wire) carrying the tumor mass

Case 3

A 22-year-old woman, had severe epigastralgia, associated with vomiting, without evidence of clinical cholestasis. The physical examination showed sensitivity in the epigastrium.

An MRI was made, objectifying a large cystic mass of the pancreas of 5x6x7 cm with thin and regular wall, heterogeneous content, with T1 hypersignal area (hemorrhagic); thus, a conclusion to a remodeled hydatic cyst of the pancreas (Figure 6).

The patient underwent surgery, we found a solido-cystic mass of the head of the

pancreas of 7 x 7 cm with respect to the mesenteric vessels and the celiac trunk.

A cephalic duodenopancreatectomy (the procedure according to Whippel) was indicated, an anastomotic assembly according to Child (Figure 7).

The postoperative follow-up was without anomaly and the patient was discharged on the 9th day.

The anatomopathological and immunohistochemical study concluded that it was a pseudopapillary and solid tumour of the pancreas.

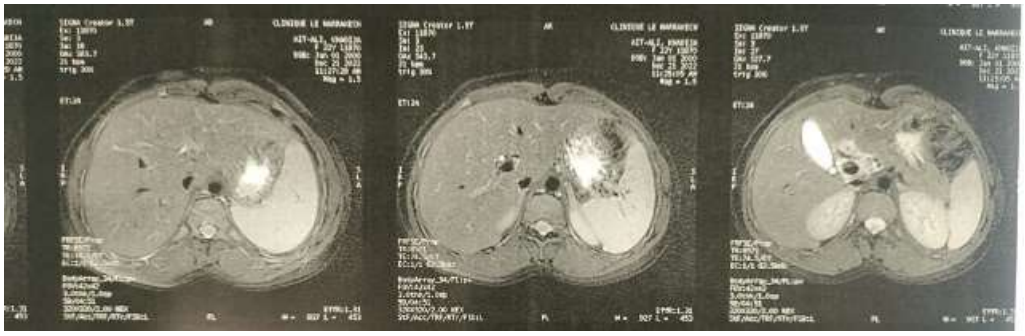

Figure 6: MRI of the cystic mass, the remodeled hydatic cyst of the pancreas

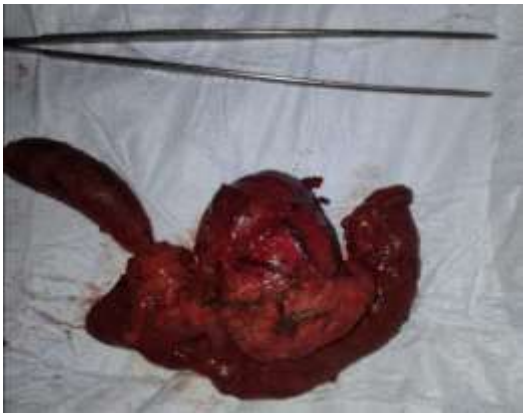

Figure 7: Piece of cephalic dudeno-pancreatectomy for solid tumour of the head of the pancreas

Case 4:

A young woman of 24, with no known pathological history, presented with paroxysmal abdominal pain in the epigastrium and left hypochondrium that had been present for about a year, of moderate intensity, with no other associated signs, all in a context of good general condition. On clinical examination, the patient was in good general condition and had a firm epigastric mass with little pain, fixed posteriorly and about 6 cm long on abdominal palpation.

An abdominal CT scan showed a large pancreatic tumour, caudal to the body, tissue and cystic with small peripheral calcifications measuring approximately 16 cm long, compatible with a solid pseudopapillary tumour of the pancreas. She underwent laparotomy and was found to have a huge mass in the tail of the pancreas measuring approximately 20 x 20 cm, solid cystic in nature with a purplish wall, coming into contact with the pedicle and the splenic hilum without a cleavage plane, the rest of the pancreas was normal in appearance. The operation consisted of a caudal splenopancreatectomy. Anatomopathological examination of the surgical specimen showed a morphological appearance in favour of a solid pseudopapillary neoplasm of the pancreas infiltrating the mesosplenic bone opposite the splenic pedicle and hilum, with healthy pancreatic resection limits; the immunohistochemical complement concluded that the appearance was in favour of a solid pseudopapillary neoplasm of the pancreas. The post-operative course was straightforward and the 6-month follow-up was unremarkable.

Case 5:

The patient was 27 years old, with no specific pathological history, and had been treated for intermittent abdominal pain and asthenia. The clinical examination was unremarkable. Abdominal ultrasound revealed a heterogeneous hypoechoic mass in the left hypochondrium measuring 65x77 mm, poorly circumscribed, vascularised on colour Doppler, in intimate contact with the inferior pole of the homolateral kidney, with respect for the fatty separation line, suggesting a stromal tumour. Abdominal CT scan showed a left flank mass measuring 66x82 mm spontaneously hypodense and heterogeneously enhanced by contrast medium with regular wall, arriving in intimate contact with the stomach, kidney and homolateral renal pedicle, and tail of the pancreas with respect for separating fat interfaces. Laparotomy revealed an 8*7 cm solidocystic tumour, adjacent to the tail of the pancreas, in contact with the splenic vein and the transverse mesocolon, with the presence of a cleavage plane. The mass was removed by caudal pancreatectomy. Pathological examination revealed a morphological appearance of an encapsulated tumour proliferation, with a poorly differentiated papillary appearance and the presence of a vascular embolus, and immunohistochemistry revealed a pseudopapillary solid tumour of the pancreas. Post-operative follow-up was straightforward and the one-year check-up with a CT scan was unremarkable.

Table 1: Patient Demographics and Tumor Characteristics

	Age	**Tumor Location**	**Tumor Size**	**Surgical Intervention**
Cas 1	25	Body and tail	113x85X120mm	Caudal spleno-pancreatectomy with resection of the left 1/3 of the transverse colon
Cas 2	22	Body and tail	66 x 77 mm	Caudal pancreatectomy
Cas 3	22	Head	53x60x70 mm	A cephalic duodenopancreatectomy
Cas 4	24	Body and tail	66x82 mm	Caudal spleno-pancreatectomy
Cas 5	27	Tail	120x110x100,5 mm	Caudal pancreatectomy.
Age, tumor location, tumor size and surgical intervention				

DISCUSSION

Pseudopapillary and solid tumors of the pancreas are rare tumors and predominate in the female sex with a percentage that varies between 1 and 2% of pancreatic tumor lesions. They occur in young people and adolescents in 85-90% of cases [2]. WHO recently classified pseudopapillary and solid tumors of the pancreas as low-grade exocrine pancreatic carcinomas [2].

Symptomatic patients may present with early satiety, abdominal discomfort, vomiting or rarely jaundice. However, most often the diagnosis is incidental [3].

Imaging-wise, pseudopapillary and solid tumors of the pancreas are single heterogeneous lesions with tissue (peripheral) and cystic (central) portions. A hemorrhagic contingent is common. They are usually large (> 3 cm, average 9 cm) and may be located on all segments of the gland. Peripheral calcifications are noted in more than 60% of cases. Pseudopapillary and solid tumors of the pancreas have a low potential for malignancy, however 5-15% are metastatic, and the overall mortality is 3-15% [4].

The diagnosis is based on CT scan and MRI. In case of doubt, an echo endoscopy should be performed. Although essential for preoperative diagnosis, the sensitivity and specificity of cytopuncture for pseudopapillary and solid tumors of the pancreas are not known due to the rarity of this pathology coupled with the lack of quality data [5].

The features on echo-endoscopy are a distinct cystic lesion with a solid component

and the possibility of calcifications within the cyst. Occasionally, these lesions may present as purely cystic or solid [6, 7].

Histologically, pseudopapillary and solid tumors of the pancreas have a heterogeneous appearance with solid cellular areas, pseudopapillary structures, and necrotic or hemorrhagic debris. A degenerative process may result in nuclear atypia; however, the presence of mitoses is rare. Although pseudopapillary and solid tumors of the pancreas are considered low-grade malignancies, more aggressive dedifferentiated variants (sarcoma) have been described [8].

Immunohistochemistry can be valuable in ruling out differential diagnoses, including pancreatic neuroendocrine tumor. However, biopsy should be strongly discussed because of the potential risk of swarming on the puncture site in case of malignant lesion. In current practice, cytopuncture is rarely necessary. Indeed, a review of the literature on 718 patients showed that only 52 of them (7%) had a biopsy before surgery. The remaining patients were operated on according to clinical history, physical examination and radiological data [9].

The treatment is surgical and is consensual. In operable patients, surgical resection should be complete by cephalic duodeno-pancreatectomy or left pancreatectomy with preservation of the spleen if possible. Enucleation, even if technically possible, should be avoided because of the potentially malignant nature of the pseudopapillary solid pancreatic tumor. Local recurrence and metastasis are rarely reported, a review of the literature showed a recurrence rate of 6.63% at ten years of follow-up [10].

CONCLUSION

Pseudopapillary and solid tumors of the pancreas are rare tumors that occur in young subjects and whose treatment of choice is surgical. There is no consensus on the type of surgery or the curage but the resection must be complete.

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