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CASE REPORT

Solid pseudopapillary neoplasm – Management of an extremely rare case of pancreatic tumor in a young patient

Valentin Daniealopol[1], Radu Mircea Neagoe[2], Daniela Tatiana Sala[2], Botond Istvan Kiss[1], Ruxandra Daniealopol[1]*

1. Second Department of Surgery, Emergency County Hospital of Targu Mures, Targu Mures, Mures, Romania
2. Second Department of Surgery, George Emil Palade University of Medicine, Pharmacy, Science, and Technology of Targu Mures, Romania

Introduction: Franz Tumor or the solid pseudopapillary neoplasm is a very rare form of pancreatic cancer, that can be held responsible for approximately 0.2-2% of the exocrine pancreatic tumors. **Case report**: We report a case of a 20-year-old woman, without any comorbidities, admitted to our department accusing mild abdominal pain at the level of the left hypochondrium and epigastrium, that started approximately half a year ago. Computed tomography and magnetic resonance imaging showed a voluminous encapsulated tumor, with mixed structure, apparently adherent to the tail of the pancreas, left kidney and adrenal gland, though being unable to certainly establish its visceral origin. **Management and results**: After analyzing all aspects of the case, we decide in favor of a left subcostal laparotomy approach; the intraoperative aspect is that of a relatively well delineated mass, adherent at the level of the pancreatic tail, therefore a complete excision of the tumor alongside with the pancreatic tissue that came in contact with it was performed. The pathological analysis reveals an encapsulated tumor with solid and pseudopapillary structure, with hemorrhagic and cystic degeneration regions; therefore, we reach the following final diagnosis: pT3 stage pseudopapillary-solid pancreatic neoplasm. **Conclusions**: In spite of its malignant behavior and impressive tumor volume, the surgical intervention was curative with favorable prognosis.

Keywords: solid pseudopapillary neoplasm, Frantz tumor

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Introduction

The solid pseudopapillary neoplasm or Frantz's tumor, known in the past by many names such as "solid cystic tumor", "papillary epithelial neoplasm", "cystic papillary tumor", etc. is an extremely rare tumor, representing 0.2-2% of the exocrine pancreas tumors, affecting almost exclusively women aged 25 to 30[1]. Without having any specific clinical signs, the tumor is usually discovered accidentally, abdominal pain and discomfort appearing only in the presence of compressive phenomena due to its increased size.[2] It is described in literature as having a low degree of malignancy, the metastases being exceptional. Surgery is curative in most cases [3,4].

Case presentation

The 20-year-old patient, with no significant medical history, presents to our department accusing abdominal discomfort, mostly located in the left hypochondrium and epigastrium, with the onset approximately 6 months before. Clinical examination reveals a 10 cm in diameter mass, situated in the epigastrium region; abdominal ultrasound detects a cystic formation with unclear organic origin, probably arising from the caudal portion of the pancreas. In order to clarify the diagnosis and establish the case management, the patient is subjected to further investigations.

Magnetic resonance imaging (MRI) highlights an expansive, voluminous tumor, with mixed structure, with restricted diffusion and gadophilia at the level of the solid component, with irregular septa inside, well delimited by a pseudocapsule. It is in contact with the pancreatic caudal region and the anterior valve of the left kidney, with no visible demarcation rim. Due to its increased size, 72/78/85 mm (ap / tr / cc), it is difficult to assess the organ of origin, raising the suspicion of cystic papillary renal cell carcinoma; the tumor has tangential contact with the lateral side of the left adrenal gland, the great gastric curvature and the splenic vein, which maintains its permeability.

The computed tomography examination performed in addition to the MRI study describes the presence of a voluminous mass, with a pseudomacronodular structure, well delimited, encapsulated, starting from the inferior aspect of the pancreatic tail, developed caudally; there is a sharp angle between the tumor and the pancreatic parenchyma with a typical appearance of "ice cream cone". The formation is heterogeneous native and post-administration of intravenous iodinated contrast agent, with the presence of hypodense, parafluid areas predominantly arranged centrally and dense, iodophilic areas with mostly peripheral disposition. In terms of size, the maximum axial diameters reach 85.3 / 77 mm and about 83 mm cranio-caudal, causing a mass effect on the left kidney and left renal vein, without invading it, and on the pancreatic body-caudal parenchyma. It also comes in close proximity to the lower third of the visceral side of the spleen, but with no invasion of the splenic artery or vein.

* Correspondence to: Ruxandra Daniealopol
E-mail: ruxandradaniealopol@gmail.com

Laboratory examinations show no changes and no signs of endocrine or exocrine pancreatic glandular insufficiency. The urological examination does not detect signs of renal distress, ultrasound shows a left kidney without stones, without stasis at this level; urine summary and biochemistry are within normal limits.

Considering the clinical data, imaging and laboratory examinations, we decide in favor of a left subcostal laparotomy approach; intraoperatively, a tumor formation of approximately 105x95x75 mm is discovered, located at the level of the caudal segment of the pancreas, adherent to it, relatively well delimited by a conjunctive capsule, which allows excision of the encapsulated tumor, en bloc, together with the distal portion of the pancreatic tail. Postoperative evolution is favorable, the patient is discharged on day 6 after surgery (Figure 1).

The pathological analysis of the operative specimen reveals a tumoral mass with solid and pseudopapillary structure, with hemorrhagic and cystic degeneration regions, with a conjunctive pseudocapsule at the periphery, therefore establishing the diagnosis of: solid pseudopapillary pancreatic neoplasm, stage pT3. The immunohistochemical profile expresses β-catenin, CD10, SOX11, vimentin, CD99, CD56 and focal Synaptophysin, being negative for E-cadherin, chromogranin, CTK7, CTK19. The Ki67 proliferation index is <2% (Figure 2).

Discussion

Solid pseudopapillary tumor of the pancreas is a particularly rare entity, usually affecting young women in their second and third decades of life, but cases have also been reported among pediatric patients [5]. Often imaging incidentalomas, these neoplasms are frequently located in the head or tail of the pancreas, but literature also mentions retroperitoneal, mesenteric or adrenal as possible locations [4].

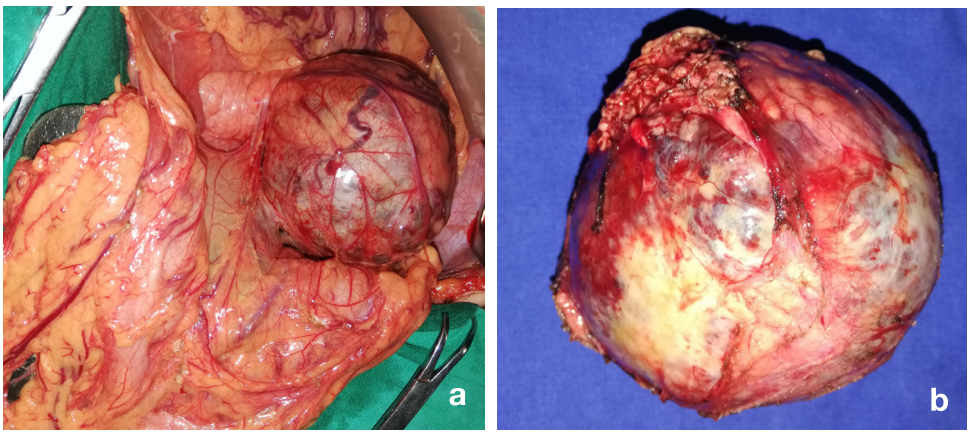

Fig. 1. (a) Surgical in situ aspect; (b) Excised operative specimen

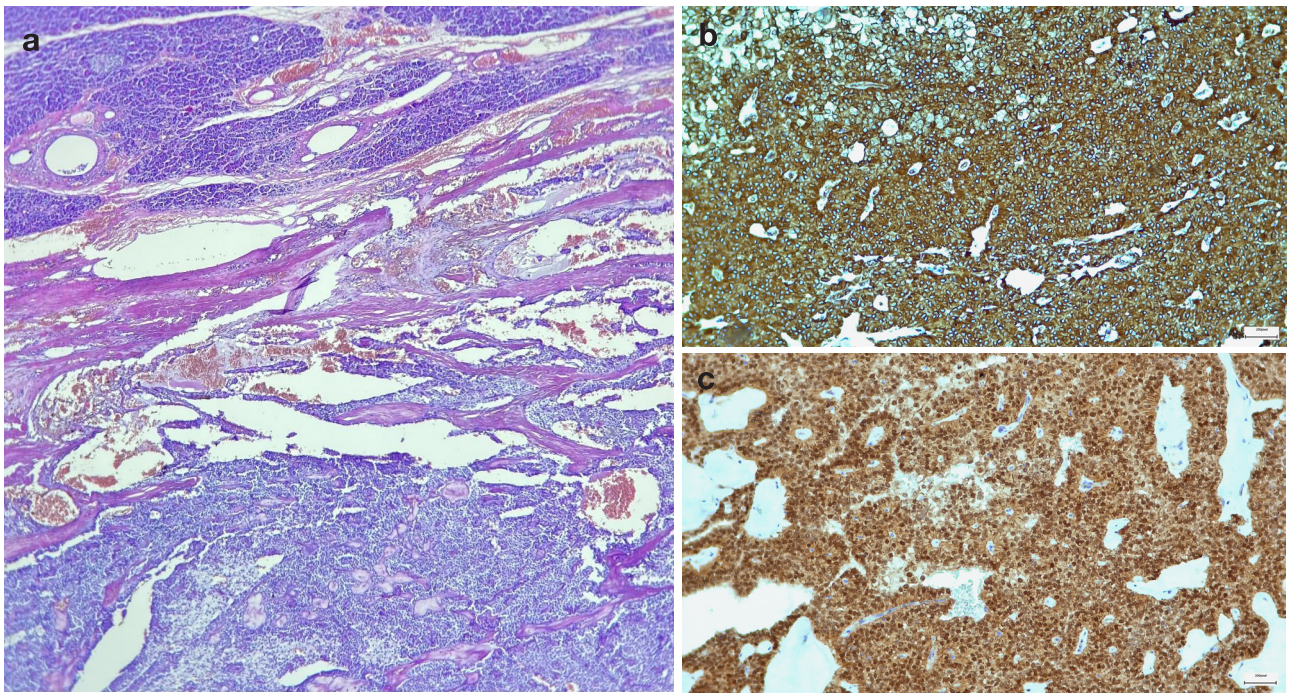

Fig. 2. (a) Hematoxylin-eosin histological aspect 5x; (b) Vimentin immunohistochemistry aspect 5x; (c) β-catenin immunohistochemistry aspect 10x

The clinical picture is poor and nonspecific, in about only a third of cases associating abdominal pain and discomfort. The tumor is palpable in approximately 12% of patients. Preoperative diagnosis is mainly based on imaging examinations, biopsy and fine needle aspiration having a controversial role [6,7].

Regarding the management of these cases, although statistical data are yet insufficient, it is generally accepted that surgery is the only effective curative treatment [8]. In view of the low malignant potential of this tumor, surgical treatment associates a good prognosis with a survival rate of over 95% [4]. Radical resections, duodenopancreatectomies for cephalic tumors and distal pancreatectomies with or without splenectomy for body or caudal tumors are usually performed; In selected cases, interventions aimed at preserving the pancreatic parenchyma, such as targeted resections or enucleations, are an option if they can be performed within safe limits. Despite the often increased size of these neoplasms, tumor volume is not a predictor of resectability [8, 9].

Although adjuvant therapies have been considered, the role of chemo- and radiotherapy is still unclear, given the low degree of malignancy of this tumor [10]. On the other hand, there are authors who take into consideration hormone therapy, given the tumor expression of estrogen receptors [10].

In terms of immunohistochemical profile, most pancreatic solid pseudopapillary neoplasms express vimentin (> 90%), α-1 antitrypsin (> 90%), neuron-specific enolase (> 80%), synaptophysin, cytokeratin, progesterone, estrogen and chromogranin A; positivity for Ki67, can be a predictor of malignant potential and unfavorable outcome [4,11].

Conclusions

Despite the increased dimensions and the malignant character of the tumor, the surgical intervention was curative with complete postoperative recovery and favorable prognosis.

Authors' contribution

DV - Supervision, Investigation, Writing – review & editing
NRM - Supervision, Investigation, Methodology
SDT - Supervision, Investigation, Methodology
KBI - Investigation, Methodology
DR - Supervision, Investigation, Writing – original draft

Conflict of interest

None to declare.

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