Solid Pseudopapillary Tumor of the Pancreas (Frantz's Tumor): Case Report

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Neoplasia Pseudopapilífera Sólida de Pâncreas (Tumor de Frantz): Relato de Caso

Neoplasia Sólida-Pseudopapilar de Páncreas (Tumor de Frantz): Relato de Caso

Carlos Rafael Lima Monção[1]; Levindo Tadeu Freitas de Figueiredo Dias[2]; Cecília Barbosa Alcântara[3]; Thereza Cristina Abreu Versiani[4]; Autieri Alves Correia[5]

ABSTRACT

Introduction: The solid pseudopapillary tumor of the pancreas (Frantz's tumor) is a neoplasm extremely rare that represents less than 1% of all the pancreatic tumors. **Case report:** 48 years old female patient was hospitalized with intra-abdominal mass of etiology hard to define. She was submitted to surgical treatment with successful laparotomy tumor resection and there were no postoperative complications. The patients usually become symptomatic only after long time of evolution, when they have discomfort or abdominal pain or palpation of abdominal masses during physical examination. Complementary exams like ultrasound and computed tomography are necessary to diagnosis and usually they show heterogeneous large intra-abdominal masses. It is necessary the confirmation with anatomopathological study which usually has histological characteristics compatible with low-grade malignancy. The surgical resection is the recommended treatment and even large size tumors has a considerable chance of cure if free surgical margins are obtained. It is known that complete tumoral resection is associated with long-term survival. **Conclusion:** The solid pseudopapillary tumor of the pancreas (Frantz's tumor), although rare, should be considered in differential diagnosis of pancreatic neoplasms because surgical treatment usually has good chances of cure even in large size tumors.

Key words: Pancreatic Neoplasms; Abdominal Neoplasms; Pancreas; Women; Case Reports.

RESUMO

Introdução: A neoplasia pseudopapilífera sólida de pâncreas (tumor de Frantz) é muito rara e representa menos de 1% de todos os tumores pancreáticos. **Relato do caso**: Paciente do sexo feminino, 48 anos, internada para investigação de massa intra-abdominal de etiologia a esclarecer. Foi submetida a tratamento cirúrgico com sucesso na ressecção tumoral e não apresentou complicações no pós-operatório. Os pacientes usualmente tornam-se sintomáticos somente após longos períodos de evolução, quando então apresentam desconforto ou dor abdominal, ou a palpação de massas abdominais no exame físico. Para o diagnóstico, são necessários exames de imagem como ultrassonografia e tomografia computadorizada, que geralmente evidenciam volumosas massas heterogêneas intra-abdominais. É necessária a confirmação com estudo anatomopatológico e comumente estas possuem características histológicas compatíveis com tumores de baixo grau de malignidade. A ressecção cirúrgica é o tratamento recomendado e mesmo os tumores de grandes dimensões possuem chances de curas consideráveis, se for possível obter margens cirúrgicas livres. Sabe-se que a ressecção tumoral completa é associada a longos períodos de sobrevida. **Conclusão:** A neoplasia pseudopapilífera sólida de pâncreas (tumor de Frantz), apesar de rara, deve ser considerada no diagnóstico diferencial de tumores de pâncreas, já que o tratamento cirúrgico apresenta boas chances de cura mesmo em tumores de grandes dimensões.

Palavras-chave: Neoplasias Pancreáticas; Neoplasias Abdominais; Pâncreas; Mulheres; Relatos de Casos.

RESUMEN

Introducción: La neoplasia sólida-pseudopapilar de páncreas (tumor de Frantz) es muy extraordinaria y representa menos de 1% de todos los tumores pancreáticos. **Relato del caso**: Paciente do sexo femenino, 48 años, internada para investigación de masa intraabdominal de etiología a esclarecer. Fue sometida al tratamiento quirúrgico con éxito en la resección tumoral y no presentó complicaciones en el postoperatorio. Los pacientes por lo general se convierten en sintomáticos solamente después de largos períodos de evolución, cuando presentan incomodidad o dolor abdominal o existe palpación de masas abdominales en el examen físico. Para el diagnóstico son necesarios exámenes de imagen como ecografía y tomografía computarizada que generalmente demuestran voluminosas masas heterogéneas intraabdominales. Es necesaria la confirmación con examen anatomopatológico y generalmente tienen características histológicas compatibles con tumores de poco grado de malignidad. La resección quirúrgica es el tratamiento indicado y mismo tumores de grandes dimensiones poseen posibilidades de curas considerables se sea posible obtener márgenes quirúrgicas libres. Se sabe que a resección tumoral completa es asociada a largos períodos de sobrevida. **Conclusión:** La neoplasia sólida-pseudopapilar de páncreas o tumor de Frantz: a pesar de extraordinaria, debe ser considerada en el diagnóstico diferencial de tumores de páncreas y a que el tratamiento quirúrgico presenta buenas posibilidades de cura, mismo en tumores de grandes proporciones.

Palabras clave: Neoplasias Pancreáticas; Neoplasias Abdominales; Páncreas; Mujeres; Informes de Casos.

[1]University Hospital Clemente de Faria (HUCF). Montes Claros (MG), Brazil. Orcid iD: https://orcid.org/0000-0001-9784-9569
[2]HUCF. Hospital Foundation Dilson Godinho. Montes Claros (MG), Brazil. Orcid iD: https://orcid.org/0000-0002-7524-8567
[3]HUCF. Montes Claros (MG), Brazil. Orcid iD: https://orcid.org/0000-0003-0661-003X
[4]HUCF. Montes Claros (MG), Brazil. Orcid iD: https://orcid.org/0000-0002-6649-6550
[5]HUCF. Montes Claros (MG), Brazil. Orcid iD: https://orcid.org/0000-0002-5948-0526
Corresponding author: Carlos Rafael Lima Monção. Rua Tupinambás, 13, Sala 1312 - Melo. Montes Claros (MG), Brazil. CEP 39401-509. E-mail: crmoncao.crm@gmail.com

INTRODUCTION

Firstly described by Frantz in 1959, the solid pseudopapillary tumor of the pancreas is rare (represents 0.3% to 2.7% of all pancreatic neoplasms), most common in young women generally in their third decade of life with low malignancy potential[1,2]. The most common presentation is abdominal mass. The treatment is essentially surgical with good chances of cure as it is a slow growing tumor neoplasm[3].

The methodology utilized was based in the model of elaboration of case report of the Case Report Guidelines (CARE) with the description of clinical findings, diagnostic evaluation, therapeutic intervention and clinical follow up of rare cases or atypical presentation[4].

The present study has the objective of presenting a case report with the description of the clinical history, changes of the physical exam, of the imaging tests in addition to the surgical treatment, of the anatomopathological study and late post-operative follow up of a rare incidence tumor and, for this reason, it is paramount to call the attention of healthcare professionals about the diagnosis of Frantz tumor and the clinical experience learned with this case.

CASE REPORT

Female, 48 years old patient admitted to the University Hospital Clemente de Faria of Montes Claros complaining of abdominal pain for five months. Pain of moderate intensity located in left hypochondrium without irradiation. The condition was followed by nausea, hyporexia and ponderal loss of 5 kg in two months. The abdominal physical exam revealed mass in the left hypochondrium, sore at palpation, movable, stiffened, irregular and apparently flat borders. Abdomen ultrasound revealed extensive heterogenous mass with hypoechogenic areas permeated in close contact with the pancreas tail the computed tomography revealed as voluminous mass of soft parts apparently encapsulated, well bounded and containing parietal calcifications, presenting irregular enhancement through contrast with hypoattenuating areas of diffusion located in the left hypochondrium/flank in close contact with the pancreatic body/tail and gastric chamber measuring around 12.1 x 11.3 x 10.0 cm, compatible with neoplasm (Figure 1 and Figure 2), having Frantz tumor and gastrointestinal stromal tumor (GIST) as diagnostic hypothesis.

Laparotomy with bilateral subcostal incision was performed. In the inventory of the cavity, large tumor lesion was encountered, affecting the pancreas body and tail with close contact with the splenic artery and vein, no peritoneal implant or signs of the advanced disease were detected. Resection of the lesion and ligature of the splenic vein were performed. The pancreatic stump was closed with double suture thread (Figure 3).

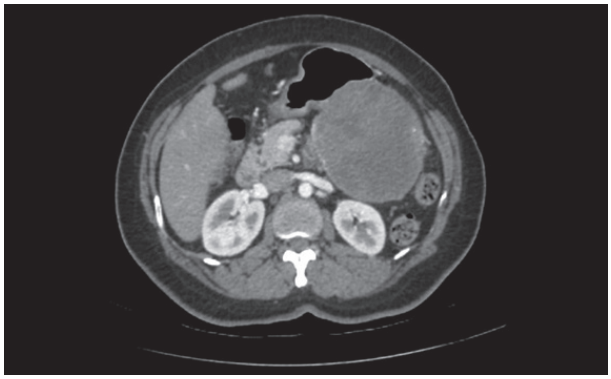

Figure 1. Tumor cross-sectional computed tomography image

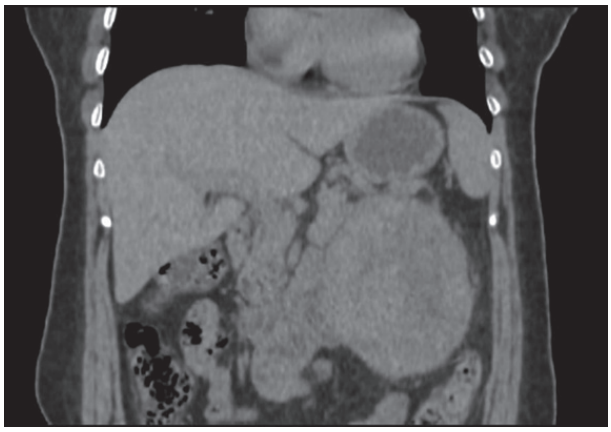

Figure 2. Computed tomography with coronal image of the tumor

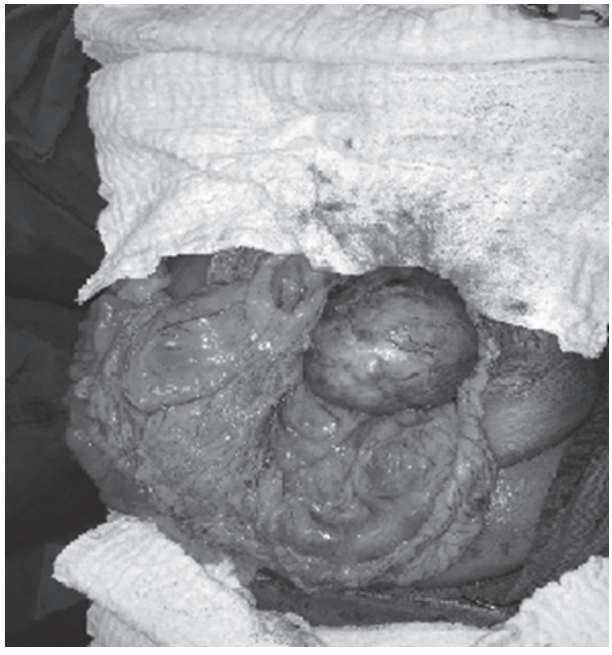

Figure 3. Laparotomy showing neoplastic lesion

At macroscopy the anatomopathological findings revealed tumor formation measuring 12.0 x 11.0 x 8.5 cm and weighing 619 grams; brownish bosselated external surface involved in fibrous pseudocapsule with small area suspected of capsular leaking and at the excisions, brownish lesion, friable, heterogeneous with cystic and hemorrhagic areas.

At the histopathological exam, it was confirmed pancreatic solid pseudopapillary neoplasm (Frantz tumor) with free surgical margins. The patient evolved without postoperative complications. She is being followed up at the clinic oncology of Hospital Dílson Godinho ever since in semester consultations. One year after surgery, she was asymptomatic with unaltered physical and laboratory exams: hemogram, ions, kidney function, LDH, ferritin, CA19-9, CA125 and CEA.

The Institutional Review Board of the "*Faculdades Integradas Pitágoras* of Montes Claros, Brazil, approved the case report CAAE: 28326620,8.0000,5109.

DISCUSSION

In 1959, Frantz described three cases of pancreatic tumors with dissimilar characteristics of the most common pancreatic neoplasms due to solid and cystic areas within the encapsulated tumor most of all.

Henceforward approximately 800 cases were described in the scientific literature. It is estimated that these tumors are responsible for 0.3% to 2.7% of all the pancreatic neoplasms[1,5]. Usually, they affect young female patients in the third decade of life in a proportion of 10:1[6,7].

Patients are often asymptomatic for long periods and the most common presentation is the abdominal mass, most of the times of large dimensions with abdominal pain and rarely with jaundice[2].

The treatment is essentially surgical. This neoplasm has good prognosis because of its low tumor growth[8]. Even tumors of large dimensions can undergo surgical resection with chances of cure[9]. The surgical treatment can involve the necessity of evaluation of vascular resections to obtain free surgical margins. Usually, there is no indication of lymphadenectomy due to the apparent lymph node invasion of this specific type of neoplasm. Surgical laparoscopy is preferable depending on the location of the tumor in the pancreatic parenchyma, of the dimensions of the neoplasm and the staff experience. The most common postoperative complication is pancreatic fistula[5].

Other adjuvant therapeutic modalities as chemotherapy and radiotherapy have also been reported but without strong scientific evidences as it is a tumor with few cases described and good response to surgical treatment alone[10].

After complete surgical resection, nearly 95% of the patients had disease-free survival after five years. The tumor relapses are associated to surgical resections with surgical positive margins mainly and are usually diagnosed by evidences of liver metastases or relapse of the pancreatic parenchyma itself[11].

CONCLUSION

The case report of solid pseudopapillary neoplasm of the pancreas, considered rare with few descriptions in the scientific literature in a patient submitted to surgery successfully is important for other physicians to get acquainted with this type of tumor and its clinical presentation, its characteristic alterations in the imaging exams and the description of the surgical treatment performed, sharing and describing the experience learned with this case.

CONTRIBUTIONS

Carlos Rafael Lima Monção contributed for the conception and/or design of the study, collection, analysis and interpretation of the data, wording and critical review. Levindo Tadeu Freitas de Figueiredo Dias and Cecília Barbosa Alcântara contributed for the wording of the manuscript. Thereza Cristina Abreu Versiani and Autieri Alves Correia contributed for the conception and/or design of the study, collection, analysis and interpretation of the data. All the authors approved the final version to be published.

DECLARATION OF CONFLICT OF INTERESTS

There is no conflict of interests to declare.

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None.

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