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Case Report

Solid-Pseudopapillary Tumor of the Pancreas: Frantz's Tumor

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Introduction

The pseudopapillary solid tumor of the pancreas, also known as Frantz´s tumor, is a rare disease, taking place in approximately 0.17% to 2.7% of non-endocrine tumors of the pancreas. Recently, the increase of its incidence has been noted with more than two-thirds of the total cases described in the last 10 years. A possible explanation is a greater knowledge of the disease and a greater uniformity of conceptualization in the last years. Generally, it affects young adult females. In most of the series, the tumor principally attacks the body and tail of the pancreas.[1]

The objective of the present report is to present the diagnostic and therapeutic option used in this rare pancreatic tumor of low-grade malignancy.

Case Report

A 29-year-old female patient, melanodermic, was referred to our hospital with symptoms of jaundice associated with back pain and accented ponderal loss, with three months of evolution. Patient presented without other comorbities and without significant alterations to the physical examination. A subsequent computerized tomography of the abdomen showed lesion in the uncinate process of the pancreas, tumor mass presenting heterogeneous calcifications. Patient was submitted to laparotomy in February 2007, identifying a tumor in uncinate process of the pancreas, with absence of peritoneal and other organ compromise. A duodenopancreatectomy was performed, with *en bloc* resection. The histologic analysis was compatible with solid-pseudopapillary tumor of the pancreas.The diagnosis was confirmed by immunohistochemical analysis. Patient evolved without complications and observed in regular out-patient follow-up, without any signs of recurrence.

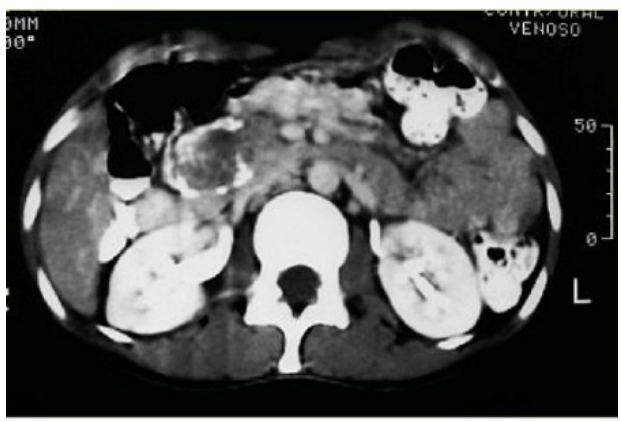

Figure 1- Tumor and Peace of duodenopancreatectomy

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Discussion

In 1959, Frantz reported, as a new entity, cases previously misdiagnosed as nonfunctioning pancreatic islet cell tumors. In 1996, this tumor was named "solid-pseudopapillary tumor of the pancreas" by the World Health Organization.[1]

The pathogenesis and cellular origin are uncertain. Some authors defend the origin of the tumor from embryonic cells. In virtue of the findings not having been compatible with any specific cell line of the pancreas, these authors suggested that some embryonic cells could remain in the pancreas during embryogenesis, subsequently giving rise to solid-pseudopapillary tumor of the pancreas. It is a tumor with a low degree of malignancy. The clinical findings are vague, with the patient generally reporting light abdominal pain and hyporexia.[2]

Image exams, such as ultrasound, computer tomography and magnetic resonance, are fundamental in diagnosis and surgical planning. Vimentin and neuron specific enolase are immunohistochemically positive.[3]

Resection, duodenopancreatectomy or distal pancreatectomy, is the appropriate surgery. Surgeries should be conservatives since the lesion is low grade and encapsulated. Most of the studies agree that extended resections or lymphadenectomies are not indicated. Invasion of veins (portal vein or superior mesenteric artery) do not contraindicate resection.[4]

The rates of resectability are high, due to the tumor growth moving the adjacent structures instead of invading them. More than 95% of the patients with lesion limited to the pancreas are cured by complete resection. Patients with tumors not completely resected can survive for more than 10 years after the operation.[5] Adjuvant chemotherapy and radiotherapy have been proposed in some cases; however, without definite conclusion.[1-2,4]

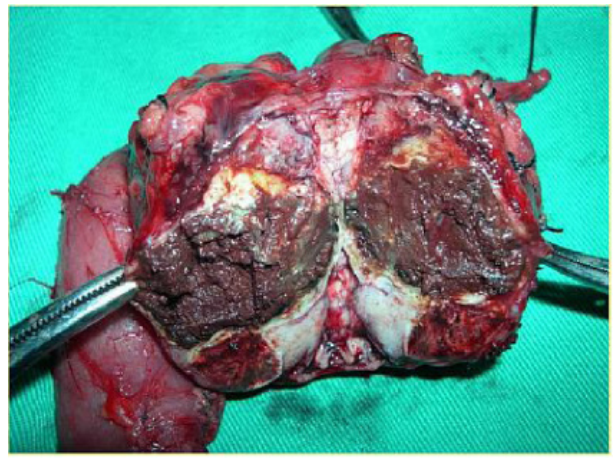

Figure 2 - Tumor specimen

Conclusion

Surgical resection is the best treatment for solid-pseudopapillary tumor of the pancreas, being in most cases, the only sufficient treatment.

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