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CLINICAL CASE

Frantz's tumour. Unusual presentation in two adolescents

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KEYWORDS

Pancreatic cyst; Pancreatic neoplasm; Solid pseudopapillary neoplasm

Abstract

Introduction: Solid pseudopapillary neoplasm of the pancreas is a rare form of cancer with low malignant potential, accounting for 0.17–2.7% of all pancreatic tumours.

Case report 1: A 17-year-old female patient who presented with symptoms six months after a Caesarean section. She was referred to our institution for a $10\,\mathrm{cm}\times10\,\mathrm{cm}$ tumour in the upper right quadrant. Abdominal computed tomography showed a mixed mass at the head of the pancreas compatible with Frantz's tumour. Underwent surgery, in which the classic Whipple procedure was performed.

Case report 2: A 17-year-old female patient with a history of blunt abdominal trauma days before admission. Abdominal computed tomography showed a $10.6\,\mathrm{cm}\times10\,\mathrm{cm}$ heterogeneous and septate tumour at the head of the pancreas; preoperative diagnosis of solid pseudopapillary neoplasm. Underwent surgery, in which the pylorus-preserving Whipple procedure was performed.

Conclusion: Solid pseudopapillary neoplasm of the pancreas is a rare tumour of unknown origin and with good prognosis after surgical resection.

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PALABRAS CLAVE

Quiste pancreático; Neoplasia pancreática; Neoplasia sólida-quística pseudopapilar

Tumor de Frantz. Presentación inusual en dos adolescentes

Resumen

Introducción: Las neoplasias sólidas pseudopapilares del páncreas son una rara neoplasia de bajo potencial maligno, representan del 0.17 al 2.7% de todos los tumores pancreáticos.

Caso clínico 1: Paciente femenino de 17 años de edad, cursando el sexto mes post cesárea. Refieren a nuestra institución por tumoración en cuadrante superior derecho de 10x10 cm. Tomografía abdominal reportó una masa con componente mixto dependiente de cabeza de páncreas compatible con tumour de Frantz. Intervenida quirúrgicamente, realizando procedimiento de Whipple clásico.

Caso clínico 2: Paciente femenino de 17 años de edad, antecedente de traumatismo contuso abdominal días antes de su ingreso. Tomografía abdominal reporta tumoración heterogénea y septada dependiente de cabeza de páncreas de 10.6x10 cm, intervenida quirúrgicamente con diagnóstico preoperatorio de neoplasia sólida pseudopapilar realizando procedimiento de Whipple con preservación pilórica.

Conclusión: La neoplasia solida pseudopapilar del páncreas es una entidad rara, con un origen desconocido con buen pronóstico con la resección quirúrgica.

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Introduction

Solid pseudopapillary neoplasm of the pancreas was first described by Frantz in 1959 as a rare neoplasm of low malignant potential, accounting for 0.17–2.7% of all pancreatic tumours and 3% of all cystic neoplasms of the pancreas. It is also known as Frantz's tumour, solid and papillary tumours, solid and cystic tumour, papillary cystic neoplasm, and solid papillary epithelial tumour. In 1996 the World Health Organisation defined it as solid pseudopapillary tumour of the pancreas, reclassifying it as solid pseudopapillary neoplasm (SPN), and as low-malignant neoplasm of the exocrine pancreas in 2010. 1,2

SPN mostly affects young women in the second or third decade of life, with an average age of 27.2 years, at a female-to-male ratio of up to 9.5:1, while in children the female-to-male ratio is 3:1.^{3,4}

The origin, prognostic factors and natural history of these tumours are unknown, although the tendency to affect young women suggests that sex hormones may be involved in its origin.⁵

Currently surgical resection is the treatment of choice for SPNs even with local invasion or metastasis. The 5-year overall survival rate is about 95%. 1,3

Below we present two cases of SPN in 17-year-old adolescents who were treated at the Hepatopancreatobiliary Surgery and Paediatric Surgery departments of Hospital General de México Dr. Eduardo Liceaga.

Case report 1

A 17-year-old female patient who presented with symptoms six months after Caesarean section, with a history of exploratory laparotomy performed 2 months earlier at another hospital, with a preoperative diagnosis of

cholecystitis. She was referred to our hospital upon observing pancreatic tumour.

Upon admission, the patient reported a sensation of postprandial fullness, nausea and vomiting, with no other symptoms. Physical examination revealed a good general condition, abdomen with a hypertrophic scar in the infraumbilical midline and a right paramedian scar. A $10\,\mathrm{cm}\times10\,\mathrm{cm}$ tumour was felt on palpation in the upper right quadrant, without signs of peritoneal irritation.

An abdominal ultrasound revealed a round, encapsulated heterogeneous lesion measuring $10.6\,\mathrm{cm} \times 9.3\,\mathrm{cm} \times 12.0\,\mathrm{cm}$, with a 3.4-mm wall, regular and well-defined margins, a volume of $629.9\,\mathrm{cc}$, and no dilatation of the main pancreatic duct. Abdominal computed tomography showed a complex mixed mass at the head of the pancreas that did not result in dilatation of the bile duct or the main pancreatic duct (Fig. 1). The

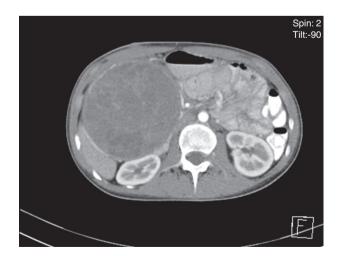


Figure 1 Abdominal computed tomography (axial plane, arterial phase): showing a mixed mass at the head of the pancreas.

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