Duodenum-preserving pancreatic head resection in solid pseudopapillary neoplasm—Report of a case

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ABSTRACT

INTRODUCTION: Solid pseudopapillary neoplasm (SPPN) was first characterized by Virginia Frantz in 1959. The duodenum-preserving pancreatic head resection (DPPHR) has been described as treatment for low-grade malignant tumors of the head of the pancreas including eight cases of SPPN.

PRESENTATION OF CASE: A 16-year-old white female patient presented with abdominal pain and dyspepsia. Computed tomography scan of abdomen showed a 10 × 9 × 10 cm3 lesion on the pancreatic head. After radiological diagnosis of SPPN the patient was submitted to DPPHR. Resection was achieved with clear margins. Immunohistochemical study demonstrated positivity for progesterone receptor, β-catenin, cytoplasmic paranuclear dot-like CD99, negativity for chromogranin and S100 protein and Ki 67 index of 1%.

DISCUSSION: A large encapsulated pancreatic mass with well-defined borders that contains areas of calcifications and intratumoral hemorrhage on CT scan in a young female is virtually diagnostic of an SPPN. A particular dot-like intracytoplasmic expression of CD99 appears to be highly unique for SPPN

CONCLUSION: DPPHR should be considered in cases of SPPN in the pancreas head if there is no compromise with oncologic radicality.

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1. Introduction

Solid pseudopapillary neoplasm (SPPN) was first characterized by Virginia Frantz in 1959 and was defined by The World Health Organization (WHO) as “solid pseudopapillary tumors” of the pancreas in 1996. It is a neoplasm of unknown etiology, low malignant potential and good prognosis that occurs most often in young women. Symptoms of SPPN are often nonspecific but the radiological appearance is distinct from other pancreatic tumors. Complete resection has a favorable prognosis.

Pancreatoduodenectomy is the most described surgical procedure to SPPN in the head of the pancreas. The duodenum-preserving pancreatic head resection (DPPHR) has been described for treatment of low-grade malignant tumors of the head of the pancreas including SPPN, if there is no compromise with oncologic radicality.

We present a case of 16-year-old female patient with solid-pseudo papillary neoplasm submitted to duodenum-preserving pancreatic head resection. The histopathology examination and immunohistochemical study confirmed the diagnosis.

2. Presentation of case

Written informed consent was obtained from the patient for publication of this case report and accompanying images. In October 2012, a 16-year-old white female patient presented with abdominal pain and dyspepsia, which started when she was at age of 12. Computed tomography scan of abdomen showed a 10 × 9 × 10 cm lesion on the pancreatic head (Fig. 1). The radiological diagnosis was of a solid pseudopapillary neoplasm. Serum tumor markers were carcinoembryonic antigen (CEA): 0.8 ng/mL and carbohydrate antigen 19-9 (CA 19.9): 50 U/mL. The patient was submitted to a duodenum-preserving pancreatic head resection (Fig. 2). She had a very good recovery and was discharged on the 9th post-operative day.

Pathologic evaluation showed a well-demarcated encapsulated tumor weighing 201.6 g and measuring 7.0 × 6.5 × 6.0 cm. Sectioning demonstrated a solid mass with soft areas, fibrous bands and hemorrhage focus, without cystic degeneration (Fig. 3). The histological sections showed a neoplasm characterized by polygonal cells, dot-like nuclei, and round, pleomorphic nuclei forming solid nests and papillae with fibrous vascular axes.

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3. Discussion

Solid pseudopapillary neoplasm (SPPN) otherwise known as solid and papillary tumor, solid-cystic tumor and Frantz’s tumor, among others, was first characterized by Virginia Frantz in 1959 as a “papillary cystic tumor of the pancreas”. The patient was a 2-year-old boy who died during an attempted duodenectopancreatectomy. The World Health Organization (WHO) defines it as “solid pseudopapillary tumors” of the pancreas since 1996.

SPPN is an uncommon indolent cancer of unknown etiology, low malignant potential and good prognosis that account for 2–3% of primary pancreatic malignancies. SPPN occurs most often in women during their second and third decades as a slow-growing mass, and can arise at any location within the pancreas (head, body, or tail), without a preferential site. If a complete resection is achieved, most patients have a favorable prognosis, with higher than 90% overall cure rate. However, 10–15% of the patients with SPPN reported developing metastasis and/or recurrence.

Symptoms of SPPN are often nonspecific and include abdominal pain, dyspepsia, early satiety, nausea and vomiting. It is distinct from other pancreatic tumors both in epidemiologic distribution as well as radiological appearance. A large encapsulated pancreatic mass with well-defined borders that contains areas of calcifications as well as intratumoral hemorrhage on CT scan in a young female is virtually diagnostic of an SPPN. Our patient presenting symptoms were comparable with previous literature, with the most common complaint being abdominal pain.

Grossly, the tumor is a solid mass well circumscribed with regions of necrosis, hemorrhage and cystic degeneration with a thick fibrous capsule. Histologically, the solid portions contain sheets, cords and nets of uniform, rather small, and fairly round cells in a rich and delicate vascular network. Many of the cells farthest from the vessels undergo degeneration, causing the remaining cells around the vessels to form pseudorosette or pseudopapillary patterns. Rarely, the SPPN exhibit a high-grade component that consists of diffuse sheets of cells with a high nucleus to cytoplasm ratio, necrosis and elevated mitotic activity suggesting the development of a poorly differentiated carcinoma. These tumors are associated with a rapidly progressive and fatal clinical course.

The tumors show immunohistochemical stain for vimentin, α-1-antitrypsin, neuron-specific enolase, CD96 and CD10. Positivity staining for chromogranin (a precursor of several neuroendocrine peptides) is never seen. The abnormal immunohistochemical nuclear localization of β-catenin staining has been proposed as a diagnostic aid. Corresponding loss of the cell surface expression of E-cadherin is also seen, and these abnormalities likely contribute to reduce cell cohesion that characterizes this neoplasm. A particular dot-like intracytoplasmic expression of CD99 appears to be highly unique for SPPN. CD99 is a glycoprotein encoded by the pseudoautosomal gene MIC2. It is typically present in Ewing sarcomas (ES), primitive neuroectodermal tumors (PNETs), lymphoblastic lymphomas, synovial sarcoma and other solid tumors, but the immunostaining in all those cases is almost exclusively confined to the cellular membrane. The characteristic cytoplasmic paranuclear “dot-like” pattern described in SPPN has also been found in colonic adenomas/adenocarcinomas, pituitary tumors and endometrial serous carcinomas with ES differentiation, but to date it has not been described in any other type of endocrine or
Fig. 4. Histopathology and immunohistochemical study in SPPN. (a) Round cells forming papillae with delicate fibrovascular axes (haematoxylin and eosin; original magnification ×200). (b) Progesterone receptor showing strong and diffuse nuclear positivity in the neoplastic cells (×200). (c) Negativity for chromogranin immunohistochemical stain with positive internal control in a Langerhans islet (×200). (d) Nuclear and cytoplasmic positivity for β-catenin plus membrane staining in normal pancreatic tissue (×400) (e) Cytoplasmic paranuclear dot-like positivity for CD99 stain (×400). (f) Low Ki-67 (MIB-1) labeling index (×200).

exocrine pancreatic tumors included in the differential diagnosis of SPPN.16,17

Several types of procedures have been developed to preserve organ function and reduce complications in patients with low-grade malignancies of the pancreas if there is no compromise with oncologic radicality.9,10,18 The importance of duodenal preservation in pancreatic surgery is that pancreatic enzyme secretion and endocrine function is not reduced after the DPPHR. This surgical procedure preserves the stomach and pylorus and spares the duodenum and the extrapancreatic bile duct, which are important for hormonal control of exocrine and endocrine pancreatic function.9,10

DPPHR was first described by Beger et al. in 198520 for treatment of chronic pancreatitis. Since then several modifications have been reported to remove benign or low-grade malignant lesions in the head of the pancreas. In the conventional DPPHR an incomplete pancreatic parenchymal resection is performed while a complete parenchymal resection, including the groove area, is done in the modified DPPHR.9,10 In the present case we performed a modified DPPHR as described by the previously cited authors, in which only two cases of SPPN was reported.

We have found eight cases of SPPN treated with DPPHR. However because the details of the operation was not described in all publications we cannot affirm exactly how many cases of SPPN were treated by the same technique that we present in our case, apart from the two ones we have mentioned before. In the series of 11 patients21 in which five cases were treated by DPPHR, the procedure performed was the technique first described by Beger et al.,20 that is different from the one we reported in our case, furthermore, there was no report of DPPHR in the recent large series of 64 patients reported by Estrella et al.8

SPPN rarely metastasize to the lymph nodes therefore aggressive regional lymphadenectomy is not routinely recommended,19 but complete resection with negative margins is imperative.5,7,8,19

In conclusion, DPHR should be considered in cases of SPPN in the pancreatic head if there is no compromise with oncologic radicality.

Conflict of interest statement

Vivian Resende and other co-authors have no conflict of interest.
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Nothing to declare.

Ethical approval

Ethical Committee of the Federal University of Minas Gerais, number—CAAE: 23377113.1.0000.5149.

Author contributions

All authors have made substantial contributions to all of the following: (1) acquisition of data, analysis and interpretation of data; (2) revising it critically for important intellectual content and (3) final approval of the version to be submitted.

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