

Solid pseudopapillary tumor of the pancreas: Report of a case after 5-year follow-up

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Key words: *Pancreatic Neoplasms; Pancreatic Pseudocyst; Child; Female*

Arch Oncol 2007;16(3-4):74-6.

UDC: 616.37-006:616-079.1

DOI: 10.2298/A000804074P

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Received: 25.11.2008

Provisionally accepted: 01.12.2008

Accepted: 08.12.2008

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Solid pseudopapillary tumor (STP) of the pancreas is an exceptionally rare neoplasm in children accounting from 1% to 2% of exocrine pancreatic tumors. Frantz ZE first described STP in 1959 as a papillary tumor of the pancreas benign or malignant. This tumor from 1996 has only recently officially been called solid pseudopapillary tumor of the exocrine pancreas, but the past has carried names like solid and papillary cystic tumor, solid and papillary epithelial neoplasm in a child and adenocarcinoma of the pancreas in childhood, as well as Frantz tumor. Various names reflect the "solid cystic" and solid pseudopapillary features. In solid area, the tumor cells are uniform around fibrovascular septa, whereas in pseudopapillary areas show degenerative changes in the cells away from the vasculature.

STP is a tumor of the primitive epithelial cells, which having capacity for exocrine and endocrine differentiation.

The pathogenesis of STP is unknown and nowadays, molecular studies are intensive. It is believed that mutations of β -catenin (S33C; the serine is changed to cystine) can cause disorganization of the E-cadherin causing the loss of adhesion junction with cytoskeleton of the cells and/or overexpression of transcription factors, which to lead to development of STP. STP typically affects young women in their second and third decade of life, especially adolescent girls. Its clinical behavior and its pathologic appearance are consistent with low-grade malignancy with an excellent prognosis if complete surgical excision is performed. Metastases have been described, however, reportedly after long disease-free periods; the lesion's size is not a predictor of unresectability.



Figure 1. CT performed before surgery in December 2002: a) native, b) with contrast medium

Native CT scan shows discreet focal lesion at the transition of the corpus into the tail of pancreas; calcification deposits are minimal. Postcontrast image shows hypodense circular lesion, relatively homogenous, 2 cm in diameter and with clearly margined from surrounding

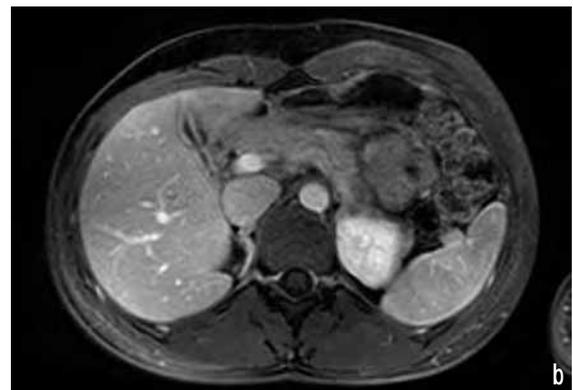
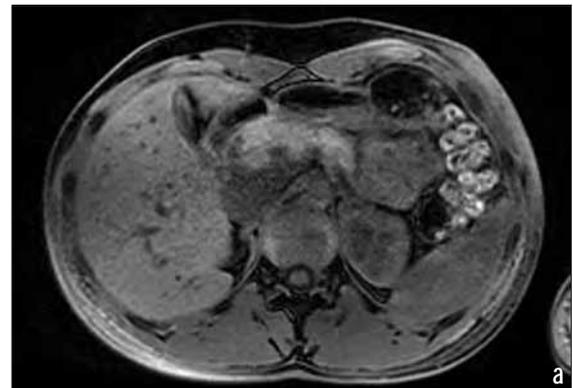


Figure 2. MRI scan performed five years after surgery (November 19, 2008): a) before contrast medium, b) with contrast medium. Both images show normal morphological appearance of the pancreas with no signs of local relapse

The characteristic ultrasound (US) findings of STP are those of well-encapsulated, cystic and solid mass, but sometimes a purely solid mass or internal separation or calcification are seen. By imaging studies, STP is circumscribed by precise margins with a well-vascularized and defined capsule. Calcification and septa are characteristic features of SPTs. The alteration of solid and cystic areas, with a necrotic or hemorrhagic component may also be present.

The usual clinical signs are vague gastrointestinal symptoms like upper abdominal discomfort or pain caused by enlarging and often palpable abdominal mass. STP often remains asymptomatic until the tumor has enlarged considerably and therefore it is sometimes detected incidentally on US imaging for unrelated diseases or after a blunt abdominal trauma. Laboratory parameters including tumor markers are normal.

The pathologic diagnosis of SPT is based on the presence of characteristic light microscopic features. Solid areas alternating with pseudopapillary formations; evidence of cellular degeneration, including cholesterol clefts and aggregates of foamy histiocytes; nuclear grooves and aggregates of hyaline cytoplasmic globules are found.

CASE REPORT

A 10-year old girl came for CT examination because of colic pain in abdomen appearing after meals. The results of ultrasound examination showed cystic lesion in pancreas of unknown origin. CT findings reported the focal capsulated hypodense lesion in pancreas, discretely calcified (Post-traumatic cyst? Echinococcus cyst? Neuroendocrine tumor?).

The patient underwent surgery on July 25, 2003; tumor of 2.5 cm in diameter was enucleated. Surgeon reported the capsulated cystic tumor with partially solid content. Five years after the operation, MR findings (Figures 2a, 2b) were normal and the patient was in good condition.

Tumor consists of oval eosinophilic cells, vascularized cytoplasm, with oval nucleus of dispersed chromatin and usually with one visible nucleus. Cells are organized in solid clusters and numerous abortive papillary formations with discrete fibrovascular stroma. Cystic formations are visible focally with new hemorrhage. Necroses are rare and small.

Immunohistochemical analysis of sampled material showed tumor cells manifesting coexpression of vimentin and sinaptophysin with simultaneous negative reaction to chromogranin and cytokeratin 7 (Figure 3, a-d). Intensely positive nuclear reaction with β catenin indicated possible connection of pathogenesis of this tumor (Figure 3e).

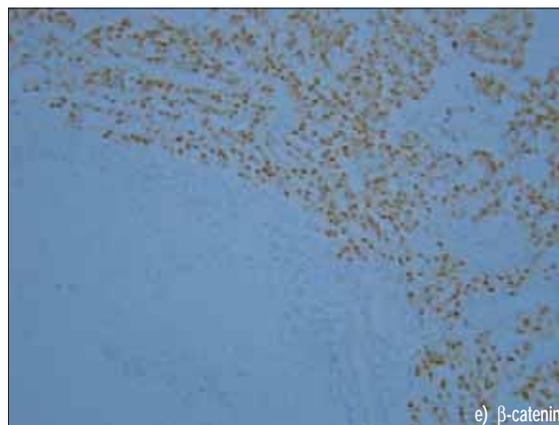
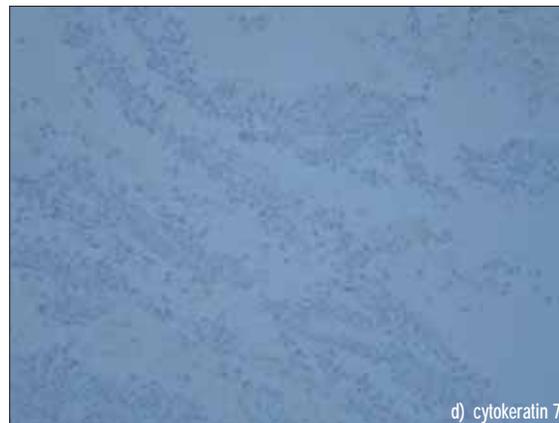
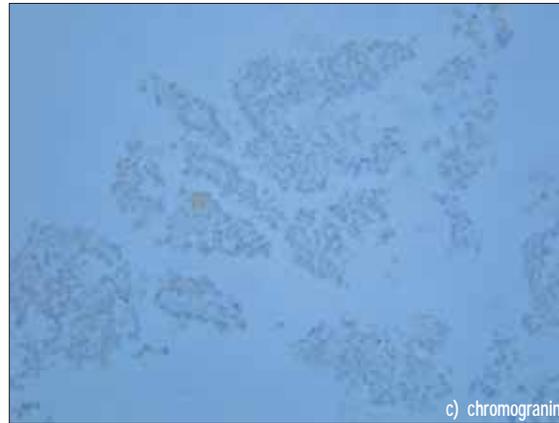
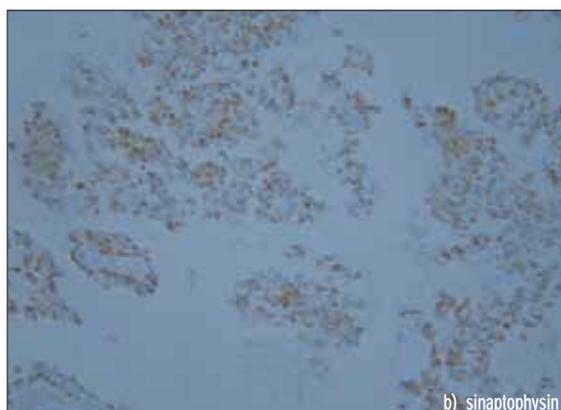
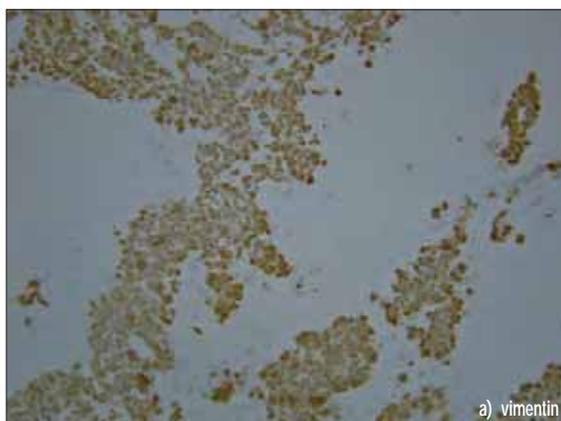


Figure 3. a-b. Positive vimentin and sinaptophysin in cells that line abortive papillary formations with discrete fibrovascular stroma and; c-d. with simultaneous negative reaction to chromogranin and cytokeratin 7; e. intensely positive nuclear reaction with β -catenin

Conflict of interest

We declare no conflicts of interest.

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