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Original article

Solid-Pseudopapillary neoplasm of the pancreas: A clinicopathological review of 20 cases including rare examples

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ABSTRACT

Aims: Solid-pseudopapillary neoplasm (SPN) is an uncommon malignant tumor of the pancreas with a favorable prognosis unlike other pancreatic neoplasms. We investigated the clinicopathological features of 20 patients with SPN in details.
Methods: The patients diagnosed as SPN in Dokuz Eylul University Hospital between January 2005 and March 2016 were reviewed in terms of clinical and histopathological data.
Results: Mean age of the patients was 33. Three of our cases were male and 4 were children. Some patients had synchronuous malignancies. Nine patients were diagnosed by fine needle aspiration cytology (FNAC). One of our tumors had 2 × 1 mm of pancreas endocrine neoplasm in addition to SPN. One case had foci of atypical and multinucleated giant cells. All cases were positive for vimentin and CD10 antibodies and most were positive for PR and β-catenin. The mean follow-up duration was 40 mo (range 2–110 mo). Only one case showed liver metastasis.
Conclusion: Herein we present a series of 20 patients with 3 male and 4 pediatric cases, almost half of which were diagnosed with FNAC findings, and most of which are clinically being followed with one patient showing progression. Our series includes rare examples like collision tumor of SPN and pancreas endocrine neoplasm, SPN with multinucleated giant cells. Also cases with no surgical treatment and no progression, as well as cases with synchronous malignancies are presented. We believe that FNAC findings of any pancreatic mass should be investigated in detail for the designation of a therapy plan especially for the patients with high operation risks. The findings in our series also show that extensive necrosis, angioinvasion, perineurial invasion and larger tumor size might be predictive for worse prognosis and these patients should be more closely followed up.

1. Introduction

Solid-pseudopapillary neoplasm (SPN) is an uncommon and a distinct tumor of the pancreas. It was first observed in 1927 in a 19-year-old woman [28] and first described by Frantz, as "papillary tumor of the pancreas, benign or malignant", in 1959[34]. Until 2010 WHO Classification of Tumors of the Digestive System, the tumor was named as solid-pseudopapillary tumor, solid and papillary neoplasm, solid and papillary epithelial neoplasm, papillary- cystic carcinoma, solid and cystic acinar cell tumor of the pancreas, Frantz tumor, papillary epithelial neoplasm of pancreas in a child and adenocarcinoma of the pancreas in childhood [28,29].These terms reflect the predominant morphologic features; solid, papillary and cystic. Grossly, SPNs appear as solid, cystic, solid-cystic; with or without hemorrhagic foci and microscopically the tumor presents pseudopapillary pattern. Solid areas show cords of small, polygonal, monomorphous cells with grooved nuclei and adjacent cystic areas may demonstrate cholesterol clefts with foreign body giant cells. Occasionally cells may contain PAS-positive cytoplasmic hyaline globules. SPNs mostly affect young women. They may be localized in any part of the pancreas and may rarely develop in other organs, most commonly mesocolon (with ectopic

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pancreatic tissue) and ovary [9]. The incidence is reported as 0.9% to 2.7% of all pancreatic tumors (WHO) and it constitutes about 5% of all cystic pancreatic tumors [28]. Although SPNs are classified as malignant tumors, they have a favorable prognosis unlike other pancreatic neoplasms. In this retrospective study we investigated the clinicopathological features of 20 patients with SPN in details.

2. Methods

The patients histopathologically diagnosed as SPN in Dokuz Eylul University Hospital between January 2005 and March 2016 were reviewed. Twenty patients with sufficient clinical data and available pathology material were included in this study. Medical records to determine the sex, age at initial diagnosis, type of surgery, localization and size of the tumor, progression or metastasis, the date of the last encounter and pathology archival materials in terms of status of surgical margins, angioinvasion, perineurial invasion, pancreatic parenchymal invasion, pleomorphism, mitosis, extensive necrosis and lymph node metastasis of surgical materials as well as cytomorphological findings of fine needle aspiration cytology (FNAC) materials were reviewed. İmmunohistochemical (IHC) staining results with vimentin, CD10, progesterone receptor (PR), synaptophysin (Syn), chromogranin (Chr), cytokeratin (CK), CD56 and beta-catenin antibodies were also documented.

3. Results

The clinical features of the patients are summarized in Table 1. Mean age of the patients was 33 (range 9–63), with 4 patients at pediatric age. Three of our cases were male. Each of the patients had non-specific symptoms like abdominal pain or nausea, and neither of them showed any abnormality in laboratory tests. One of the patients (case 5) was operated for colon adenocarcinoma previously and her pancreatic mass was found incidentally during routine checks. Another patient was screened for staging of a recently diagnosed Hodgkin lymphoma (case 6), and the pancreatic mass was detected by computerized tomography. One of the patients (case 15) had a synchronously diagnosed urachus tumor. Nine patients were diagnosed by FNAC. Cytomorphologic findings included papillary fragments, or solid sheets of uniform tumor cells, some of which also exhibiting nuclear grooves or mild nuclear atypia (Fig. 1). Cell block was also prepared from seven of these cytologic specimens and papillary structures were more prominent in the cell block sections (Fig. 2). The immunocytology panel was restricted to PR, Vimentin and Syn, whereas IHC performed on cell block sections included additional antibodies such as beta-catenin, CD10, CK and Chr (Fig. 3). The results are listed in Table 2. The cytomorphologic findings and IHC staining results were evaluated together with clinical and radiologic findings and the FNAC were reported as “findings suggestive for SPN”.

The pancreatic masses were totally resected in all cases except for 3 patients. Head of pancreas was the most common site. Seven cases had purely cystic tumors, 6 cases had purely solid tumors and 4 of the tumors had both cystic and solid components (Fig. 3). One case had multiple tumor nodules within the pancreas. Although macroscopically being well demarcated, microscopically the tumors had an at least minimally infiltrating pattern and 3 cases had microscopic tumor foci in the pancreatic surgical margin. The tumor cells were small and uniform with eosinophilic cytoplasm and coffee-bean shaped nuclei. They had an arrangement simulating papillary foldings around fibrovascular cores. Mitosis were rare but extensive necrosis was seen in 4 cases. One of our tumors had 2×1 mm of pancreas endocrine neoplasm (PEN) in addition to SPN (Fig. 4). Two cases revealed intracellular hyaline globules. One case

Table 1
Clinicopathologic features of the cases.

Case	Age	Sex	Location	FNAC	Type of bx/surgery	Size (cm)	Gross features	SM	VI	PNI	PPI	Mitosis	Extensive Necrosis	LN met	Additional findings	FUD	outcome
1	20	F	Head	no	Pancreaticoduodenectomi	7,5	cystic	Negative	Present	Present	Absent	0	Present	–	–	88	LFU
2	12	M	Head	no	Excision of the tumor	12	cystic + solid	Positive	Absent	Absent	Absent	1/10 HPF	Present	–	–	107	AWOD
3	9	F	Corpus+tail	no	Distal pancreatectomy+ splenectomy	8	solid	Negative	Absent	Present	Absent	1/10 HPF	Present	–	–	73	LFU
4	30	F	Head	no	Tru-cut bx	4	cystic	N/A	Absent[a]	Absent[a]	N/A	0[a]	Absent[a]	–	–	5	LFU
5	53	F	Head	no	Pancreaticoduodenectomi	4.5	solid	Negative	Absent	Absent	Present	0	Absent	–	Metachronous Colon adenoCA	109	AWOD
6	21	F	Head	yes	FNAC	3.5	solid	N/A	Absent[a]	Absent[a]	N/A	0[a]	Absent[a]	–	Synchronous HL	33	AWD
7	39	F	Head	yes	FNAC	5	cystic + solid	N/A	Absent[a]	Absent[a]	N/A	0[a]	Absent[a]	–	–	62	AWD
8	42	M	Tail	yes	Distal pancreatectomy+splenectomy	5	cystic	Positive	Absent	Absent	Present	2/10 HPF	Absent	–	Hyaline globules	55	AWOD
9	17	F	Corpus	yes	Pancreatectomy	4.5	solid	Positive	Absent	Absent	Present	0	Absent	–	Hyaline globules	56	AWOD
10	29	M	Tail	no	Pancreatectomy	4	cystic	Negative	Present	Present	Present	2/10 HPF	Absent	–	–	54	AWOD
11	57	F	Head	yes	Pancreatectomy	10	cystic	Negative	Present	Present	Absent	1/10 HPF	Present	–	Liver metastasis	49	AWD
12	28	F	Tail	yes	Distal pancreatectomy	2	solid	Negative	Absent	Absent	Absent	0	Absent	–		2	AWOD
13	36	F	Tail	yes	Distal pancreatectomy	5	cystic	Negative	Absent	Absent	Absent	0	Absent	–		41	AWOD
14	18	F	Head	no	Pancreatectomy	3,5	N/A	Negative	Absent	Absent	Absent	0	Absent	–		36	AWOD
15	52	F	Head	no	Pancreatectomy	3	N/A	Positive	Absent	Absent	Absent	0	Absent	–	Synchronous urachus tumor	20	AWOD
16	46	F	Head	yes	Pancreatectomy	2	solid	Negative	Absent	Present	Absent	0	Absent	–	Focal atypia	19	AWOD
17	63	F	Tail	no	Distal pancreatectomy	4	solid	Negative	Absent	Present	Absent	0	Absent	–	Adjacent PEN	12	AWOD
18	19	F	Head	yes	Pancreatectomy	5.5	cystic + solid	Negative	Absent	absent	Absent	0	Absent	–		11	AWOD
19	53	F	Head	yes	Pancreaticoduodenectomi	3	cystic + solid	Negative	Absent	absent	Absent	0	Absent	–		1	AWOD
20	15	F	Head	no	Pancreaticoduodenectomi	5,5	cystic	Negative	Absent	absent	Absent	0	Absent	–		9	AWOD

FNAC: Fine needle aspiration cytology, SM: surgical margin, VI: vascular invasion, PNI: perineurial invasion, PPI: pancreatic parenchymal invasion, FUD: follow-up duration, LFU: lost to follow-up, AWOD: alive without disease, AWD: alive with disease.
[a] Findings limited to tru-cut biopsy.