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Solid pseudopapillary neoplasm of the pancreas: A report of two cases and review of the literature.

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Introduction

In 2018, according to the World Health Organization (WHO) statistics, 5786 malignant pancreatic neoplasms were diagnosed in Poland, making it the ninth most frequently diagnosed cancer. The average age of patients was 49 years. Simultaneously, 5715 patients died due to pancreatic neoplasms, which constituted 5% and was the sixth most frequent cause of death due to cancer. **[1]** Solid pseudopapillary neoplasm (SPN) is a rare pancreatic tumor. It was first described in 1959 by V. Frantz in the publication “Papillary tumors of the pancreas: benign or malignant”. **[2]** SPN is more common in women, and the average age of female patients is approx. 28 years. The leading clinical manifestations include pain or discomfort in the epigastrium and palpable resistance in this region. **[3]** We present two cases of patients suspected of and diagnosed with SPN, with different clinical courses, treated in the Clinic of General, Liver, and Transplant Surgery at University Hospital No 1 named after Antoni Jurasz in Bydgoszcz.

Case reports:

Case 1:

A 23-year-old female patient was admitted to the Clinic in January 2018 due to recurrent epigastric pain. The anamnesis revealed only mild hypothyroidism. Abdominal ultrasonography visualized a 7 cm mass in the head of the pancreas. A computed tomography (CT) image showed a solid-cystic lesion in the head of the pancreas, measuring 7x6x7 cm, and a pancreatic duct dilatation to 5 mm. The levels of tumor markers were as follows: AFP=2.84 [ng/ml], CE < 0.5 [ng/ml], and CA 19- 9=5.6 [U/ml]. The radiological and surgical team decided to treat the patient surgically. The patient underwent surgery on 25.04.2018. A solid-cystic tumor in the pancreatic head, approx. 8 cm in diameter, with heterogeneous, solid-fluid content, was revealed intraoperatively. A section of the common bile duct of approx. 3 cm was adjacent to the tumor; its wall was thickened and had features of macroscopic fibrosis. Multiple, small deposits were detected in the gallbladder. Cholecystectomy was performed and then the tumor was resected, preserving pancreatic parenchyma. The section of the common bile duct affected with fibrosis was also resected. The bile duct was reconnected with end-to-end anastomosis, using a duodenal stent, with single PDS 5-0 sutures.

In the postoperative period, anastomotic leakage of the duct-to-duct biliary anastomosis occurred. Abdominal ultrasonography performed on the third day following surgery revealed a heterogenous hypoechoic area in the tumor bed as well as an anechoic subcapsular lesion in the right liver lobe, divided by multiple echoic bands. A CT scan confirmed the presence of fluid collections. During resurgery, the presence of infected hematoma above the liver was detected, as well as biliary anastomotic dehiscence and ischemia of the distal common biliary duct, with no features of a biliary fistula. End-to-side biliary-enteric anastomosis (a Roux loop), with a silicone drain from the intestines leading outside, was created. The postoperative period was complication-free. The patient was discharged in good general condition on the 27th day following surgery. The histologic examination of the resected lesion revealed solid pseudopapillary neoplasm (SPN), with a preserved surgical margin of approx. 0.1cm. Immunohistochemical staining was positive for β-catenin, CD10, vimentin, as well as (in some cells) NSE and synaptophysin, whereas negative for chromogranin. Features of neural invasion were revealed, whereas features of vascular invasion were absent.

From May to August 2018, the patient was hospitalized four times due to recurrent severe epigastric pain and episodes of fever. Episodes of cholangitis were diagnosed and treated conservatively. In July, an abscess was found in the resected tumor bed. The patient underwent re-surgery during which adhesiolysis was performed, and the abscess was drained. The biochemical test of the abscess content showed a high level of amylases. Endoscopic retrograde cholangiopancreatography (ERCP) was performed, with an attempt at catheterization and stenting of the pancreatic duct. The examination revealed the obstruction of the duct in its proximal section. An

outflow of approx. 300- 500 ml of the content from the drain placed in the abdominal cavity continued. Within a few following days, the levels of inflammatory markers decreased, and no fever was observed.

The last hospitalization occurred on 17-21.08.2018. A slight increase in the levels of amylases and CRP up to 75mg/l was observed. The patient had no fever, and the outflow from the drain ceased. An elective magnetic resonance imaging examination was performed. In the projection of the pancreatic head, a fluid-filled lesion, measuring 11x5 mm, was detected, much smaller than in the previously carried out tests, without features of an active inflammatory process. With the administration of the contrast agent, pancreatic parenchyma was enhanced homogenously, without any features of limited diffusion. Features of a pancreatic fistula were not visualized. During a 3- year follow-up period, tumor recurrence was not observed.

Case 2:

A 23-year-old female patient was admitted in the elective mode to the Clinic of General, Liver, and Transplant Surgery at University Hospital No 1 named after Antoni Jurasz in Bydgoszcz due to a pancreatic tumor. The anamnesis revealed a drop in body mass of 4kg within the last six months, type 1 diabetes, and no other ailments. The results of the physical examination and laboratory tests were within normal limits. An abdominal CT scan revealed an oval heterogeneously dense mass in the tail of the pancreas, measuring 59x38x54 mm, adjacent to the splenic artery, left renal artery, the lesser curvature of the stomach, and the jejunal loop. The levels of tumor markers were as follows: AFP=1.07[ng/ml], CEA=0.81[ng/ml], CA 19-9=7.1[U/ml].

On 08.02.2019, the patient underwent surgery, during which a tumor in the body of the pancreas, with no infiltration to mesenteric vessels, was detected. The tumor was separated from the surrounding tissues; a distal pancreatomy was performed and the entire lesion was resected. A histologic examination revealed SPN with a margin of the lesion resection of 0.1cm, positive immunohistochemical staining for β-catenin, CD56, and CD10, and negative ones for chromogranin and synaptophysin. The postoperative period was complication-free. The patient remains under ambulatory care and does not present any clinical manifestations.

Discussion

The incidence of SPN is assessed as ranging from 2 to 5 % of all pancreatic cancers. **[3]** During the last 12 years, 8 reports were published, analyzing at least 15 cases of SPN each. Most cases (116) were described in 2016 by Cai et al. **[11]** The following table summarizes the results reported in particular publications:

	Salvia et. al. (2007) [8]	**Machado et al. (2008) [9]**	**Yang et al. (2009) [10]**	**Kim et al. (2011) [13]**	**Cai et al. (2014) [11]**	**Carlotto et al. (2016) [2]**	**Lubezky et al. (2017) [12]**	**Zhan et al. (2019) [3]**
Number of cases	31	34	26	114	116	17	32	91
Females	27 (87 %)	27 (79 %)	22 (85 %)	98 (86 %)	100 (86 %)	16 (94 %)	29 (91 %)	78 (86 %)
Males	4 (13 %)	7 (21 %)	4 (15 %)	16 (14 %)	16 (14 %)	1 (6 %)	3 (9 %)	13 (14 %)
Average age [years]	34	23	30	36	35	33	28	29
Number of asymptomatic cases	17 (55 %)	7 (21 %)	11 (42 %)	52 (46 %)	32 (28 %)	1 (6 %)	9 (28 %)	35 (39 %)
The most common clinical presentation	Abdominal pain (32 %)	Abdominal pain (73 %)	Abdominal pain (35 %)	Abdominal pain (37 %)	Abdominal pain (41 %)	Palpable abdominal mass (47 %)	Abdominal pain (47 %)	Abdominal pain (45 %)
The most common tumor location	Body and tail (68 %)	Body and tail (61 %)	Head and neck (54 %)	Body and tail (56 %)	Head and neck (53 %)	Body and tail (65 %)	Body and tail (69 %)	Body and tail (49 %)

Average tumor size (the largest measurement) [mm]	60	72	63	52	64	65	59	69
The most frequent management method	Indirect resection (19.5 %) and PPPD (19.5 %)	Distal pancreatectomy (35 %)	Distal pancreatectomy (38 %)	Distal pancreatectomy (44 %)	Distal pancreatectomy (40 %) - including 24 % splenectomy	Distal pancreatectomy with splenectomy (53 %)	Distal pancreatectomy with splenectomy (53 %)	Distal pancreatectomy (42%) (including 14 % splenectomy)
Complications	-	62 %	27 %	22 %	19 %	41 %	22 %	29 %
The most common complication	-	Pancreatic fistula (56 %)	-	Pancreatic fistula (14 %)	Pancreatic fistula (11 %)	Pancreatic fistula (29 %)	Pancreatic fistula (6 %)	Pancreatic fistula (23 %)

PPPD - Pylorus Preserving Pancreatoduodectomy

The pathogenesis of this neoplasm remains enigmatic. The crucial role is attributed to the modifications in the Wnt/ β-catenin signaling pathway. In nearly all cases of SPN, deposits of β- catenin were found in the cytoplasm and nucleus of neoplastic cells. **[5]**

SPNs are most commonly single, well-demarcated, and well-encapsulated tumors, with a diameter ranging from a few to several centimeters. **[7]** Nearly half of the cases are located in the body and tail and are less frequently found in the head (35 %) and the neck (15 %) of the pancreas. **[3]** Most are benign, and the percentage of malignancies is assessed at 10-15 %. **[4,5]** If malignant, SPN can infiltrate pancreatic parenchyma, the capsule of the spleen or the duodenum, and metastases are found in the liver. **[3,4]**

The most common clinical presentations are abdominal pain (73-32 %) and a palpable abdominal mass. Less frequent symptoms include vomiting, nausea, jaundice, and body mass loss. **[3-6]** A large percentage (55-65 %) of cases is asymptomatic and the tumor is found in imaging tests performed due to other reasons. As in the patients treated in our Clinic, in the majority of cases, the levels of tumor markers CA 19-9 and CEA are not elevated. **[3]** CT remains the most beneficial imaging examination. The radiological appearance of SPN is typically characterized by a well-demarcated mass of solid and cystic components, and features of calcification, necrosis, and hemorrhage. [3,11] Such patterns were found in both cases reported by us. Immunohistochemical staining of the resected tumors most commonly is positive for vimentin, alpha 1-antitrypsin (AAT), CD10, progesterone receptors (PR), neuron-specific enolase (NSE), synaptophysin and negative for chromogranin A and cytokeratin. **[5]** A similar pattern was found in our patients. The method of choice to manage SPN is complete surgical resection. The selection of the procedure depends on the tumor site and its infiltration into adjacent structures. In the case of tumors located in the head of the pancreas, the most common procedure is pancreaticoduodenectomy; whereas when the body or tail is involved – distal pancreatectomy with or without splenectomy. Most surgeries are conducted with the classical laparotomy. The percentage of postoperative complications ranges from 19 to 62 %. The most common complication is a pancreatic fistula of grade B according to the International Study Group for Pancreatic Surgery (ISGPS). **[2,3,9-12]** Features of the fistula occurred in our case 1. However, its presence was not unequivocally confirmed by imaging examinations and during relaparotomy. It needs to be stressed that despite the tumor resection preserving pancreatic parenchyma in case 1, during a 3-year follow-up neither local tumor recurrence nor metastases were observed. Prognosis in SPN is good. In the aforementioned studies, only single cases of deaths were reported. Particularly good prognosis concerns benign tumors. A 5- year survival rate, depending on the publication, ranges from 95 to 97 %. **[3,12]**

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