A SECOND RELAPSE OF A SOLID PSEUDOPAPILLARY TUMOR OF THE PANCREAS: CASE DESCRIPTION

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Solid pseudopapillary tumor (SPT) is a rare pancreatic neoplasm of unspecified origin that occurs most frequently in young women. In most patients, it has a benign character.

In study presented a description of the case of a 36-year-old patient operated due to SPT in 2004. The patient had an atypical clinical course of the disease with a double relapse of the neoplastic process, which was operated on in 2006 and 2007.

The symptoms and clinical course of SPT are characterized by very high interpersonal variation. Therefore, it is necessary to deepen the knowledge of the histogenesis and biology of the tumor. Additionally, the patients who receive surgery due to the tumor must be observed for many years.

Key words: solid pseudopapillary tumor, pancreatic neoplasms

Solid pseudopapillary tumor (SPT) is a very rare pancreatic neoplasm of unknown cellular origin. It is characterized by borderline malignancy and occurs most commonly in female teenagers and young women. The tumor was first described by Frantz (1) in 1959, and it has been referred to as the Frantz tumor, solid-cystic tumor, papillary-cystic tumor, solid and papillary epithelial neoplasm, and solid pseudopapillary tumor of the pancreas. In 1996, the World Health Organization gave it the current name of solid pseudopapillary tumor (SPT) of the pancreas and classified it in the group of exocrine pancreatic tumors (2).

The diagnosis of SPT is based on a number of specific clinical qualities and the decisive diagnostic role of a histopathological examination. In most cases (90%), it occurs in young women; the average age of occurrence is 35 years, but less than 10% of those affected are over 40 (3, 4). This may indicate a relationship between the development of neoplasm and hormonal state. The clinical image, biology, and histopathological qualities of the tumor itself are characterized by remarkable diversity. In most cases, it is a slowly growing neoplasm with low malignant potential. In 15% of cases, however, it results in metastases to the liver and peritoneum (5).

Below we present the case of a patient diagnosed with SPT. This patient experienced a rarely-observed clinical course, in which there was a double relapse of the neoplastic process after radical surgeries.

CASE REPORT

On 13 August 2007, patient S.M., 37, was admitted to the 1st Department of Oncological and General Surgery, Wielkopolska Cancer Center for operative treatment of another SPT relapse in the abdominal cavity. The patient had first been admitted to a hospital near her home in November 2004 for diagnosis due to a palpable, tumorous mass in the left subcostal area.

The computer tomography revealed a solid, heterogeneous, well-bordered, 12.7x8.7 cm-sized tumor located between the spleen, stomach, pancreas, and left kidney. No lesions in the liver, spleen, or local lymph nodes were diagnosed. The body and tail of the pancreas, splenic vein, and left renal vein were strongly pressed by the lesion and retracted. The computer to-
mography did not specify the outlet of the tumor. It was only able to suggest that the tumor might originate from the intestinal mesentery, retroperitoneum, or pancreas (fig. 1). During the surgery, a tumor roughly 12 cm in diameter was diagnosed. The tumor stemmed from the tail of the pancreas, and it infiltrated the pancreas and splenic hilus. The tumor was removed by means of a left-sided resection of the pancreas and spleen.

On the basis of histopathological evaluation of the resected lesion, a diagnosis of neoplasma solidum – pseudopapillare (solid pseudopapillary neoplasm) was made. The neoplasm was 9x15x10 cm in size and locally surrounded by a pseudocapsule. The microscope image revealed a tumor of high cellularity, and cystoid spaces and pseudopapillary proliferations were present. The spleen had hemorrhages present in the hilus and under the organ’s capsule but no other microscopic lesions. The patient was sent to the Wielkopolska Cancer Center, where she remained under the control of the Surgical Outpatient Clinic and Chemotherapy Outpatient Clinic. The follow-up computer tomography examinations carried out in April 2005 and December 2005 did not reveal relapse of the process. Laboratory investigations allowing for neoplastic markers (CEA, Ca19.9) revealed no deviations from the standard. The doctors’ anxiety was aroused by an ultrasound of the abdominal cavity in May 2006. Three solid lesions that could correspond to enlarged lymph nodes on the left side of the epigastrium were described. In the next computer tomography (May 24, 2006), the presence of solid tumorous masses was diagnosed. The masses lay adjacent to one other and were intensively strengthened and interlooped within the left mesogastrium. The biggest of the lesions was 64x60 mm in size and contained a centrally-located hypodense zone of tissue decay. Smaller foci were 35x35 and 35x26 mm in size. No focal lesions were diagnosed in either the other organs of the abdominal cavity or the remaining pancreatic parenchyma after partial resection. No enlarged lymph nodes were observed in the space around large vessels.

On June 22, 2006, the patient was admitted to the 1st Department of Oncological and General Surgery, Wielkopolska Cancer Center for operative treatment. During a laparotomy, a large tumor 10 cm in diameter was found in the mesentery of the transverse colon. The tumor was infiltrating the wall of the prepyloric region of the stomach. Two other tumors 8 and 6 cm in diameter were diagnosed in the splenic flexure. Additionally, numerous implants up to 3 cm in diameter were found in the mesentery of the small intestine, rear wall of the stomach, and lesser omentum. The liver showed no focal lesions in palpation. The transverse colon with the prepyloric part of the stomach was resected. An implant 1.5 cm in diameter was resected from the region of the hepatoduodenal ligament. A tumor 3 cm in diameter was resected from the mesentery of the splenic flexure. Similar lesions were resected from the mesojejunum.

The histopathological examination confirmed the diagnosis of solid pseudopapillary tumor of the pancreas. The incision lines and intestinal wall were free from neoplastic lesions, and no metastases were diagnosed in the lymph nodes.

In July 2006 and January 2007, the state after the resection of neoplastic lesions was described in a follow-up computer tomography examination. No signs of relapse of the neoplastic process were noted.

However, progression of the disease was diagnosed in another computer tomography carried out in April 2007. At the level of the navel on the right intraperitoneal side, a 42x42 mm solid mass was observed bound to the wall of abdominal integuments. An image of the pan-
creatic region showed no change when compared with the previous examination. No enlarged lymph nodes were diagnosed.

Due to the symptoms of progression in the computer tomography, the administration of chemotherapy was suggested after consultation with a clinical oncologist. The patient was qualified for treatment in ambulatory conditions. A typical scheme for the treatment of pancreatic cancer (Gemcitabine + Cisplatin) was applied.

Another computer tomography examination in July 2007 revealed no positive reaction to the chemotherapy. The observed lesion grew to the size of 52x44 mm (fig. 2). No enlarged lymph nodes were observed in the space around large vessels. The patient was qualified for operative treatment. During the surgery (16 August 2007), a tumor 7 cm in diameter that originated from the mesentery of the small intestine and had grown into the parietal peritoneum in the left iliac fossa was found. A fragment of the small intestine with the tumor was resected. In addition, a polycyclic tumor invisible in the computer tomography was diagnosed between the head of the pancreas and stomach and was also resected. In the histological image, the solid pseudopapillary tumor was diagnosed again.

Presently, the patient is receiving care from the Surgical Outpatient Clinic, Wielkopolska Cancer Center. Her last check-up on October 25, 2007 included an ultrasound of the abdominal cavity and did not reveal any suspicious lesions. The patient feels well and does not report any ailments.

**DISCUSSION**

Solid pseudopapillary tumor is a very rare neoplasm that comprises about 1-2% of exocrine pancreatic tumors (2). Until 2000, only about 450 cases of the neoplasm had been described; two-thirds of these were described in the last decade. This type of tumor mainly occurs in women in the second and third decade of life. In many cases, the tumor is diagnosed incidentally during an examination carried out for an unrelated reason. In some patients, the following symptoms occur: pain in the abdomen, palpable mass, dyspeptic symptoms, and mechanical jaundice (6). The course of the disease does not show typical deviations in laboratory investigations. Additionally, routine neoplastic markers like Ca19.9, Ca125, CEA, and AFP do not exceed the standard limits.

The diagnostics of SPT find examination via imaging methods, such as ultrasound, CT, and MRI, most useful. In an ultrasound of the abdominal cavity or computer tomography, we usually obtain an image of a well-separated polycystic mass filled with a fluid alternating with solid lesions (7, 8). An MRI additionally reveals regions of hemorrhage inside the tumor (9).

In a macroscopic image, the tumor looks like a large lesion about 10 cm in diameter. It is typically well-separated and surrounded by an incomplete capsule, and very rarely does it infiltrate adjacent organs of the abdominal cavity. In most cases, the tumor primarily develops in the head or tail of the pancreas. In the literature, however, there are descriptions of cases of atypical multifocal or extrapancreatic occurrences in the mesocolon, extraperitoneally, in the omentum, or even intrahepatically (10, 11). In a histopathological image, two characteristic types of cellular organization forming alternating solid and pseudopapillary regions can be seen (12).

Differentiation of SPT is a very important problem. In differential diagnostics, the following need to be taken into consideration: endocrine pancreatic tumors, which is morphologically similar but with a much worse prognosis; acinar cell carcinoma, which mainly occurs in men and can be differentiated based...
on the absence of pseudopapillary structures and positive immunohistochemical examinations for the presence of pancreatic enzymes and cytokeratins; pancreatoblastoma, which occurs in children less than 10 years in age.

In most cases, the prognosis in SPT is favorable; 97% of patients experience a 5-year period free from relapse after a radical surgery (13). According to the data from the literature, however, metastases of SPT mainly to the liver, peritoneum, and rarely regional lymph nodes can be seen in 15% of patients. However, the mere presence of metastases in SPT does not dramatically worsen a patient’s prognosis. Some authors suggest that while the infiltration of vessels, nerves, and neighboring organs, cellular diversity, and a large mitotic index may be related to the metastatic and relapsing character of the neoplasm, the absence of these qualities does not exclude a malignant course for the disease. Therefore, all patients need observation for many years.

The treatment of choice for SPT involves surgical resection the tumor itself, its metastases, and its local relapses (14). Some authors suggest that adjuvant treatment is unnecessary and does not improve the prognosis (15). Taking into consideration the rarity of the neoplasm’s occurrence, it is impossible to explicitly define the usefulness of non-surgical treatment. In the literature, there are reports of individual cases treated by means of radiotherapy (16). Additionally, there is information about the effectiveness of preoperative chemotherapy with the application of cisplatinum and 5-FU or gemcitabine (14, 18).

Forty-eight years have passed since Frantz described the neoplasm, but its histogenetic origin still arouses controversy. Different origins, ranging from the lobulated cells of the pancreas to the outlet ducts to endocrine cells or even neuroectodermal cells, have been suggested (14, 19). However, none of these hypotheses explains the strong relationship between the occurrence of the neoplasm and female sex. Kosmahl and coll. observed the similarity between the SPT and ovarian theca cells, and the proximity of the sex crests and pancreas during embryogenesis could explain the origin of SPT from the cells of sex crests incorporated into the pancreas during organogenesis. Furthermore, the presence of progesterone receptors in the tumorous tissue in many cases indicates the probable contribution of hormones to the pathogenesis of SPT. In contrast, no estrogen receptors are found.

In summary, the available literature shows that SPT is a rare neoplasm with low malignancy that predominantly occurs in young women. There is great diversity in both the clinical presentation of the neoplasm and the course of the disease process. Surgical resection remains the only effective and proven method of treatment for both the original tumor and metastatic lesions or consecutive relapses.

REFERENCES


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COMMENTARY

The presented study demonstrated a rare case of pancreatic tumor. Solid pseudopapillary pancreatic tumor is only diagnosed in young females, and it is associated with benign character and good prognosis. Radical surgery is the therapeutic method of choice. The authors of the study presented a rare form of the tumor. In spite of excision during a three year observation period, this tumor produced local recurrence and distant metastases. Such a course for the disease is rarely observed, but it should be considered in 15% of patients. We presently have two such patients, although one at the time of diagnosis presented with distant liver metastases. Unfortunately, there is no data concerning the efficacy of chemotherapy or radiotherapy in the treatment of advanced forms of solid pseudopapillary pancreatic tumors. Admittedly, the course of the disease is slow even in case of advanced forms, and the condition of patients surprisingly good. However, the final prognosis is unfavorable. The presented case provides a good illustration of the current knowledge concerning the problem. Despite the good prognosis, the authors justly suggest the need for both long-term observation of patients diagnosed with solid pseudopapillary pancreatic tumors and surgical intervention in case of disease recurrence.

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