SOLID PSEUDOPAPILLARY TUMOR OF THE PANCREAS IN A 39-YEAR-OLD WOMAN – CASE REPORT

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Solid pseudopapillary tumor (SPT) is a rare pancreatic tumor with low malignancy, constituting 1-2% of exocrine pancreatic tumors, occurring mostly in young women. SPT, despite the achievement of large size gives scant clinical symptoms. The authors present the case of 39-year-old woman with SPT, diagnosed at intraoperative biopsy. The aim of this study is to describe the clinicopathological feature, diagnosis and surgical treatment of SPT.

Key words: solid – pseudopapillary tumor, diagnosis, treatment

Solid pseudopapillary tumor (SPT) is a rare neoplasm of the pancreas with a low malignant potential, first described in 1959 by an American pathologist Virginia Frantz (1). In 1996, this neoplasm was officially named “solid pseudopapillary tumor” (SPT) by the World Health Organization (WHO). The SPT is most prevalent among young females (82% of cases) in their 20s or 30s (2). SPTs constitute approximately 5% of pancreatic neoplasms and approximately 1–2% of endocrine neoplasms (3). The SPT is located primarily in the body of the pancreas, but can also occur in the spleen or duodenal wall. The tumor can measure up to 30 cm in diameter. It is usually enveloped by a fibrous capsule and well demarcated from the normal pancreatic tissue (4). Despite growing to a large size, the tumor causes few clinical symptoms. The histological structure of SPT is characterized by solid spaces built of pseudopapillary cells that stain positive for Vimentin, neuron-specific enolase (NSE), and alfa-1-antitripsin, as well as express progesterone receptors (5). The treatment of choice for a SPT is surgery.

CASE REPORT

A 39-year-old previously healthy woman (B.S.), with a month’s history of pain in the left upper quadrant radiating to the back and a several-day history of fever, was admitted to the hospital. Physical examination revealed a tender mass in the epigastrium measuring approximately 15 cm. Abdominal ultrasonography showed a mixed solid/cystic lesion of approximately 13.5 cm in the tail and, partly, in the body of the pancreas. Endoscopic retrograde cholangiopancreatography (ERCP) showed an apparent “cutoff” of the pancreatic duct (of Wirsung) within the body of the pancreas. An abdominal magnetic resonance imaging (MRI) scan revealed a proliferative lesion, measuring 120 x 93 x 123 mm, of mostly cystic unhomogenous structure with “septa” and hemoglobin degradation products, showing evidence of marked enhancement within its solid parts, mostly peripherally and in the “septa”, located in the left hypochondrium. The stomach, the left adrenal gland, and the left kidney were compressed by the tumor; the left kidney was also pushed back. Retroperitoneal lymph nodes on the left side were found to be enlarged, up to 15 mm. The head and the body of the pancreas were normal. The anterior surface of the tumor displayed the stretched distal part of the pancreatic tail. The posterior surface of the tumor compressed splenic vessels (fig. 1, 2). Laboratory tests showed no
abnormalities. The levels of cancer antigens Ca 19-9 and CEA were also within normal limits.

The patient was qualified to undergo surgery. Intraoperatively, the tumor was assessed to be of approximately 15 cm in diameter and involve the body and tail of the pancreas. The tumor was excised together with part of the body and tail of the pancreas, as well as the spleen. The remaining part of the pancreas was sutured shut with a double running suture (Prolene 3.0) and additionally covered with a sponge containing thrombin and human fibrinogen. An intraoperative macroscopic assessment revealed a mass measuring 14 cm in diameter, enveloped by a fibrous capsule that contained several foci of visible neoplastic tissue; the center of the tumor contained extensive hemorrhagic/necrotic areas. An [intraoperative] microscopic examination yielded a diagnosis of solid pseudopapillary tumor. This diagnosis was confirmed in the final histological examination. Splenic tissue was free from pathological lesions. On day 16 after the procedure, the patient was discharged home in good general and local condition.

**DISCUSSION**

The SPT is a rare primary pancreatic neoplasm with a low malignant potential. As of 2010, there have been 951 case reports of SPT diagnosed mainly in Europe, the USA and Japan (2, 6). In comparison, there were 553 cases of this neoplasm reported in China between 1996 and 2009 (4). To date, there have been no publications on this disease in Polish literature.

The SPT most commonly affects young women. The mean age of those affected ranges from 27.2 to 30.3 years (2, 4). There have been reports of the SPT occurring also in children (14). The prevalence of the SPT varies between men and women, with a ratio of 1 : 9.5, respectively (5). SPT cells express progesterone receptors, therefore a particularly high incidence of this tumor is observed among women with polycystic ovary syndrome (2, 7).

According to Chinese reports, the SPT is most commonly found in the head of the pancreas (39.8%), whereas European, American and Japanese reports show that this tumor is most commonly found in the tail of the pancreas (35.9%) (4, 6).

The SPT can be found also outside the pancreas, mainly in such structures as the spleen, duodenal wall, left adrenal gland, small-bowel mesentery, and retroperitoneum. Despite its low malignant potential, the neoplasm metastasizes to the liver, omentum, and retroperitoneum in 15% of patients (2, 4). The SPT tends to proliferate locally, which was the case here, but it can also infiltrate surrounding structures (70% of cases), especially the vena cava, splenic vein, spleen, diaphragm, stomach, omentum or retroperitoneum. The size of reported tumors ranges from 0.5 to 34.5 cm (6). The most common clinical symptoms are: an uncharacteristic abdominal pain, sometimes radiating to the back (>46%), abdominal distension, loss of appetite, nausea, and vomiting. In approximately 15.5% of patients the SPT is asymptomatic and is detected incidentally (4, 6).

Fever, as in the case presented here, is rare
(1.24%) (6). Due to a non-characteristic clinical presentation, imaging studies, such as ultrasound, CT, and MRI, play a key role in SPT diagnostics. Abdominal ultrasound typically reveals non-homogenous lesions within the pancreas. Abdominal CT scans exhibit a sensitivity of over 70%.

A typical CT finding in SPT patients is the presence of a rather large, heterogeneous, mixed solid/cystic, encapsulated lesion with visible peripheral calcifications (2, 4, 5). The inhomogeneous low signal intensity on T2-weighted MRI images of the abdomen is characteristic for hemoglobin degradation products within the tumor, which allows for the differentiation of the SPT and other pancreatic tumors (10, 11). This characteristic image was observed in our patient’s MRI scan. The sensitivity of abdominal MRI scans in diagnosing SPTs is 90% (5, 12) and MRI is the examination of choice in pregnant women (11). A PET scan, apart from a higher F-FDG uptake by tumor cells, did not show any features characteristic for the SPT (5). Fine-needle aspiration (FNA) biopsy was not performed. The use of FNA in SPT diagnostics is quite controversial. Some researchers believe that this technique helps diagnose the SPT in 75% of cases and constitutes an essential part of SPT diagnostics (2, 13, 14). Others believe the contrary and are of an opinion that due to the danger of neoplastic cell implantation and the risk of complications such as bleeding or pancreatic and biliary fistulas, FNA is a high-risk procedure and should be reserved for those cases where the SPT cannot be diagnosed by abdominal CT or MRI scans (4, 15). Kibil, Kulig et al. emphasize the importance of endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA). The diagnostic accuracy of this technique in pancreatic adenocarcinomas reaches 90%. However, one disadvantage of EUS-FNA is difficulty in microscopic examination of the material, as EUS-FNA aspirates tend to be contaminated with gastric epithelial cells (16).

One characteristic microscopic feature of the SPT is the presence of cysts surrounded by solid tissues, with pseudopapillary structures penetrating into the cyst lumen (2). Although the SPT is an exocrine neoplasm, immunohistochemical examinations show, apart from a positive reaction for alpha-1-antitrypsin and alpha-1-antichymotrypsin, a positive reaction for neuron-specific enolase, characteristic for endocrine tumors. Other characteristic immunohistochemical staining features of the SPT are the positivity for CD10 antigen, progesterone receptors, and Vimentin (2, 4).

Despite the low malignant potential of the disease, an estimated 15% of adult and 13% of pediatric SPT cases are malignant (9). Malignant potential of the SPT is a result of mitotic index >30%, DNA aneuploidy, chromosome 3 trisomy, unbalanced chromosomal translocation (13:17), cell polymorphism, and vascular penetration into the tumor (4, 5). Malignant forms of the SPT, which metastasize and recur, have a higher incidence in men and in people over 30 years old (9).

The treatment of choice in the SPT is surgery (5). The type of procedure depends on the tumor’s location within the pancreas. If the tumor is located in the body/tail of the pancreas, distal pancreatic resection is conducted, most often with splenectomy, and if the tumor is located in the head of the pancreas, pancreaticoduodenectomy using Whipple or Traverso-Longmire procedure is conducted (2, 13). In the case of small, up to 1.5-cm, tumors located far from the pancreatic duct, the treatment of choice is tumorectomy (2, 6, 7). Jabłońska et al. point out the type and incidence of complications following such procedures (17). In order to reduce the risk of local recurrence, SPTs should be excised with a surgical margin of 3 to 5 mm, and any hepatic metastases should be excised with a surgical margin of 1 cm (2, 9). Unlike with other malignant pancreatic tumors, the presence of metastases in the SPT is not a negative prognostic factor and these lesions should be removed (2, 6). Routine extensive lymphadenectomy is not recommended, as the SPT very rarely metastasizes to lymph nodes (4, 5).

Despite the fact that laparoscopic resection of SPTs is not recommended, due to the risk of the “spray effect,” there have been reports of distal pancreatectomy performed via the laparoscopic approach in children (9, 15). Recent years have seen attempts at chemo- and radiotherapy. There have been reports on the successful use of 5-FU, gemcitabine, cisplatin, VP-16, cyclophosphamide, doxorubicin, and vincristin in neoadjuvant chemotherapy (3). 5-FU and cisplatin have been used post pancreatectoduodenectomy, in the
treatment of recurrent SPTs (2). The effect of chemo- and radiotherapy on 5-year survival rate is not known.

The 1-, 3-, and 5-year survival rates following radical surgery are 99.4%, 97.5, and 96.5%, respectively (4). Patients who, apart from primary tumor resection, undergo metastasectomy, survive from 6 months up to 17 years, hence the importance of radical surgical procedures in SPT treatment (6).

REFERENCES


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