SOLID PSEUDOPAPILLARY TUMOR OF THE PANCREAS IN A YOUNG WOMAN – CASE REPORT

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We report a case of pancreatic solid pseudopapillary tumor that was diagnosed in a 36-year-old female patient. This neoplasm usually occurs in young women. Solid and cystic areas form a characteristic appearance of this tumor. Surgical resection is the mainstay of treatment and is possible in the majority of cases. Neoplasm is associated with a low-grade malignancy and a very good outlook.

Key words: pseudopapillary tumor, pancreas, surgical resection

Solid pseudopapillary tumor of the pancreas is a rare neoplasm that occurs significantly more often in women, especially young (1-7). It is thought to have neither definite exocrine nor endocrine differentiation (1). Niu et al. on the basis of their immunohistochemical analysis proved that this tumor is usually exocrine with accompanying neuroendocrine differentiation (6). Its histogenesis is unclear, but it can derive from the primordial cells or pluripotent embryonic pancreatic cells with multipotential differentiation (1, 6). This tumor can be hormone-dependent because progesterone receptors are present in most of the female patients (6). It is usually encapsulated (2-6, 8, 9). It is also well-defined, well circumscribed and well-demarcated (2, 4-9). Characteristic solid and cystic portions are present in the lumen of the tumor and they can be observed on radiological examinations, such as USG, CT and MRI (2-8). Their mutual proportions can be various and the tumor can be differentiated from entirely solid to completely cystic (4, 6, 7).

CASE REPORT

A 36-year-old woman was transferred to The Department of Endocrine and General Surgery of Medical University in Łódź from The 2nd Department of Urology of Medical University in Łódź, to which she had been admitted due to the suspicion of neoplastic process within the left kidney. On abdominal CT a mass measuring 17x11x13 cm, located in the left mid abdomen and having smooth and distinct margins was detected. The mass was distinctly compressing the stomach, pancreas, spleen, left kidney and loops of the small intestine together with the splenic flexure of the colon and displacing the latter almost into the pelvis. Neither infiltration of those organs nor the origin of the mass were found. Strong, but heterogenous enhancement occurred. Near the posterior margin of the mass, below the left renal vein, a vascular pedicle was observed – probably vascularization originating from the superior mesenteric and splenic artery. Few blood vessels were present inside the mass. In abdominal organs other abnormalities were not observed. Adenopathy was not found. The suspicion of desmoid tumor or GIST was created (fig. 1). The kidney was not definitely indicated as the point of origin of the mass so the patient was transferred to The Department of Endocrine and General Surgery.

On physical examination palpable mass in the left lumbar and palpable cervical lymph nodes were the only abnormalities. The patient was qualified to the scheduled surgical proce-
The midline incision was performed. In the lesser sac the huge mass originating probably from the pancreatic tail was found. The mass seemed to be a neoplasm with hemorrhages in its lumen (fig. 2). After suction of the bloody fluid the tumor was resected completely and the region of the pancreatic tail was managed using the manual suture and the omental pack. The drain was inserted into the lesser sac. The peritoneal cavity was checked for metastases, but dissemination was not found. The abdominal layers were closed separately. After the surgical procedure 4 units of packed red blood cells and 1 unit of fresh frozen plasma were transfused. Total parenteral nutrition was not undertaken. On the third day after surgery the gruel diet was initiated and gradually improved during the next days. The postoperative period was uneventful. During the hospital stay an ultrasound guided fine needle aspiration biopsy of the mass within the right lobe of the thyroid gland had been also performed and a benign lesion was diagnosed. The patient was discharged in a good general condition 9 days after the surgical procedure.

On the histopathological examination macroscopically the encapsulated, solid-cystic mass originating from the pancreatic tail, measuring 17x13x8 cm and filled with brittle, cauliflower-like structures was observed. Microscopically solid cystic pseudopapillary neoplasm was diagnosed. Immunohistochemical staining: PR (+), CD 10 (+), synaptophysin focally (+), chromogranin focally (+), Ki 67 (+) in < 1% cells. Infiltration of the capsule by tumor cells was focally present. During postoperative follow-up of 6 months recurrence did not occur.

**DISCUSSION**

Clinical presentation of solid pseudopapillary tumor of the pancreas is various. Abdominal pain can be the most common symptom (1, 7), but the majority of patients can be also asymptomatic and a tumor is then detected incidentally (2). The other symptoms, that occur more rarely, are nausea and vomiting (5, 6, 7), discomfort in the epigastrium (4, 6), weight loss (7) or jaundice (3, 5). Even if abdominal pain does not occur in most of the patients, it is present in many of them (2, 6). Machado et al. found that this neoplasm among male patients occurs in the older age than in women and is more aggressive, what was confirmed by more frequent involvement of the portal vein or the presence of metastases (7). The authors did not observe a correlation between tumor aggressiveness and its size or patients’ age.

Surgical excision is the basis of treatment for solid cystic pseudopapillary tumor of the pancreas and offers the best chance for cure (1, 4). In the suspicion of the presence of this neoplasm surgical procedure should be attempted, even if extensive resection is required (1). Radical resection is the procedure of choice even in the presence of metastases (7), when complete removal of a primary and metastatic lesion is indicated (2). When the pancreatic neoplasm is advanced and involves adjacent organs, a surgeon ought to tend to perform radical extensive resection with complete excision of the tumor (en bloc), optimally with microscopically free margin (7). Depending on the tumor location within the pancreas, its size, local invasion and involve-
ment of the contiguous organs or structures, various intentionally radical surgical procedures are performed, such as local excision of the tumor, enucleation, spleen-preserving distal pancreatectomy, distal pancreatectomy with splenectomy, subtotal distal pancreatectomy with splenectomy, central pancreatectomy with distal pancreateojunostomy, duodenum-preserving pancreateoduodenectomy, pylorus-preserving pancreateoduodenectomy and Whipple procedure (1-9). When the tumor is more advanced, the pancreatic procedure can be extended with for example resection and reconstruction of the veins forming the portal system (7), right hemicolectomy (1) or excision of liver metastases (2). If the tumor is initially unresectable, cytoreductive surgery can extend life (6). Commonly found a large size of a tumor or its direct contact with adjacent organs and surrounding vessels do not confirm unresectability, while complete resection is usually achievable and curative (1-9). Reports describing adjuvant therapy are rare. Machado et al. presented a case of the patient who developed an unresectable local recurrence after previous pancreateoduodenectomy (7). He underwent chemotherapy receiving 5-fluorouracil (5-FU) and cisplatin. He was still alive 39 months after surgical procedure. Niu et al. undertook postoperative transcatheter hepatic arterial chemoembolization due to liver metastasis, achieving stable condition of the patient’s body (6). The other patient with an unresectable tumor contiguous to the superior mesenteric artery had undergone radiotherapy (γ radiation) and the size of tumor reduced.

Solid pseudopapillary neoplasm is characterized by a low-grade malignancy and the excellent outlook, what is supported by a high amount of initially resectable patients (even despite local advance or the presence of metastases) and those who are free of a recurrence at postoperative follow-up (1-7). Besides relatively long survivals are observed in patients who were initially managed by cytoreductive therapy or with a recurrence (6, 7). This tumor can reoccur and cause death even 14 years after the surgical procedure (4). However, deaths due to a recurrence are rare (4-7).

CONCLUSIONS

1. Solid cystic pseudopapillary neoplasm is usually found in young women.
2. Even if the tumor is locally advanced and large, radical surgical resection is realizable and should be performed.
3. If a large pancreatic tumor suggesting the presence of advanced neoplastic disease is found in a young woman, the diagnosis of solid cystic pseudopapillary neoplasm should be considered in differential diagnosis.

REFERENCES