Beger procedure in a 20-year-old female with solid pseudopapillary tumour of the pancreas

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Solid pseudopapillary tumour is a rare indolent neoplasm of the pancreas (< 2% of exocrine pancreatic tumours), which predominantly affects young women at reproductive age, without significant clinical symptoms.

We report a case of a 20-year-old Caucasian female who presented with upper abdominal pain of one-year duration. Ultrasound scans of the abdomen demonstrated enlarged pancreatic head and body containing a poorly separated mass (52 × 41 × 36 mm) with a multi-cystic component 20–24 mm in diameter. Laboratory tests including, tumour markers levels, were normal. She underwent complete resection of the tumour using a Beger procedure. By immunohistochemistry, the case stained strongly for CD10 and CD56 and was negative for cytokeratin-7 (CK-7), synaptophysin and chromogranin A. The proliferation index (Ki-67) was < 1%. The patient is being followed-up and remains healthy.

Conclusions: Solid pseudopapillary tumour is a tumour with low potential of malignancy and with generally favourable prognosis; surgical resection is usually curative.

Key words: pancreatic tumour, solid pseudopapillary tumour, surgery, tumour markers.

Contemp Oncol (Pozn) 2014; 18 (3): 211–214
DOI: 10.5114/wo.2014.41387

Introduction

A solid pseudopapillary tumour (SPT) of the pancreas is a rare type of neoplasm, which was first observed in 1927 in a 19-year-old woman, and it was described for the first time by Virginia Franz in 1959 [1, 2]. Solid pseudopapillary tumour represents less than 2% of exocrine pancreatic neoplasms, with marked adolescent predominance [3]. This tumour occurs mainly in young women in their second or third decade, but it is observed also in men in less than 10% of cases [4]. So far, around 700 cases have been reported in the English-language literature [5]. Most of them were diagnosed in Europe, the USA and Japan during the last 10 years [6]. Before its inclusion in the World Health Organization (WHO) classification of pancreatic tumours in 1996, this entity had different descriptive names in the literature ("papillary-cystic tumour", "adenocarcinoma of the childhood pancreas", "solid-and-papillary neoplasm", "Frantz tumour" or "papillary epithelial neoplasm of the pancreas") emphasizing its various biological and histopathological features [7]. According to the most up-to-date definition, introduced by the International Agency for Research on Cancer (IARC), SPT is a low-grade malignant neoplasm of the exocrine pancreas [8]. The cells of SPT are polygonal and discohesive in nature. It has histopathological features distinctive from other pancreatic tumours, comprising pseudopapillary, cystic and solid changes merging with one another [9]. Moreover, cut sections of SPT demonstrate cystic calcification, variable degrees of internal haemorrhage and necrotic zones. Immunochemistry shows expression of progesterone receptors and reveals positive staining for α1-antitripsin, CD10, CD56, Vimentin and neuron-specific enolase (NSE) [10, 11]. Neuroendocrine differentiation in occasional cases is regarded as one of the possibilities, rather than as evidence of neuroendocrine origin [12]. Interestingly enough this tumour shows a tendency to increase in malignancy with age [13]. Recurrence and metastases develop in approximately 15–20% of patients, most of which are hepatic [14, 15]. However SPT has favourable prognosis and is confined to the pancreas in 85% of patients [16]. Slow and less invasive growth means that most patients experience a long-term survival after complete resection [17]. Unfortunately, the diagnosis is frequently delayed due to its unusual presentation and rarity. Solid pseudopapillary tumour should be differentiated from more aggressive pancreatic tumours such as adenocarcinoma and endocrine tumours. In contrast to pancreatic ductal adenocarcinoma, SPT is not associated with changes in p16, K-ras, p53 or SMAD4/DCP4 genes. However, mutations in the β-catenin gene are observed in most SPTs, leading to nuclear and cytoplasmic accumulation of this protein [18]. Its extrapancreatic location is extremely rare and is related mainly to the ectopic pancreatic tissue in the omentum or the mesocolon [19].

Most available data on SPTs comes from small series and case reports, resulting in scant accurate information on optimal management.

Case report

A 20-year-old female, who presented with intermittent upper abdominal pain of one-year duration was admitted in September 2012 with an abdominal mass. She had had no previous medical history. Diagnosis was based on clinical symptoms and radiological findings. Physical examination revealed only mild epigastric tenderness. Routine laboratory test results including tumour marker levels (1.31 ng/ml, 6.84 U/ml, 10.46 U/ml and 8.5 U/ml for CEA, CA 125, CA 19-9 and CA 15-3, respectively) in peripheral blood were normal. Ultrasound scans of the abdomen demonstrated enlarged pancreatic head and body containing poorly separated mass (52 × 41 × 36 mm) with a multi-cystic component 20–24 mm in diameter. This lesion was adjacent to portal and splenic veins, but without unambiguous features of their invasion or compression. The remaining pancreatic tissue was unchanged together with undilated pancreatic duct. At laparotomy, exploration revealed an encapsulated mass 7 cm in diameter located on the borderland between the pancreatic head and body, without involvement of surrounding tissues. Tumour resection during the Beger procedure was performed. This operation was first described in 1972 by Hans Beger and involves subtotal resection of the pancreatic head with preservation a small rim of the pancreas on the duodenum as well as the posterior branch of the gastroduodenal artery [20]. During reconstruction, two pancreaticojejunal anastomoses (one to the rim of the pancreatic head and another to the pancreatic body) are performed, usually with use of a Roux loop. The procedure in our case was made as described above. Moreover, during surgery, before manipulation in the vicinity of the tumour, a blood sample from the portal vein was taken to determine tumour marker concentrations and, again, CEA, CA 125, CA 19-9 and CA 15-3 levels were within normal limits (1.72 ng/ml, 4.14 U/ml, 6.86 U/ml and 5.5 U/ml, respectively). Diagnostic laboratory tests after surgery showed high levels of amylase and lipase in serum (maximum values of 1166 U/l and 2058 U/l on the first postoperative day, respectively), which was interpreted to be proof of pancreatic origin. The morphology of the tumour was determined during a routine histopathology analysis, which was carried out at the Pathomorphology Department of the Medical University in Lodz. The microscopic study revealed resection R0 (defined as tumour-free surgical margins) of solid pseudopapillary tumour. Immunohistochemical staining was negative for cytokeratin-7 (CK-7), synaptophysin and chromogranin A and strongly positive for CD10 antigen and CD56 antigen (Figs. 1, 2). The fraction of Ki-67-positive tumour cells was below 1% (Fig. 3). The postoperative course was uneventful, and the patient was discharged after nine days with analgesics, antibiotics and low-molecular-weight heparin. Abdominal computed tomography (CT) performed one month after the surgery did not demonstrate any pathological lesion within the abdominal cavity, including the remaining pancreatic tissue; the physical examination and laboratory tests did not elicit any infectious process. The patient has been followed-up as an outpatient for 10 months and remains healthy.

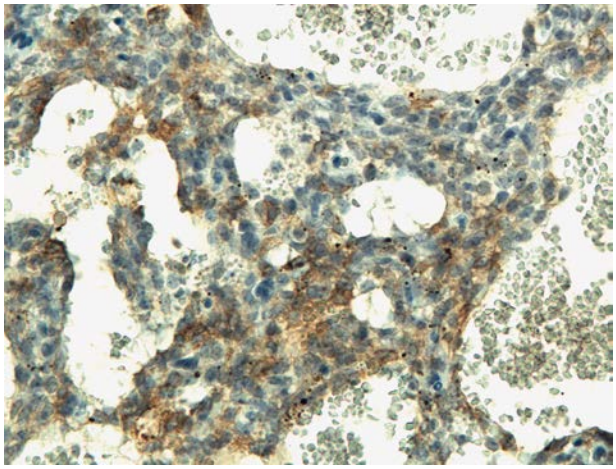

Fig. 1. Focal cytoplasmatic CD10 immunoexpression in the tumor cells. Magnification 200×

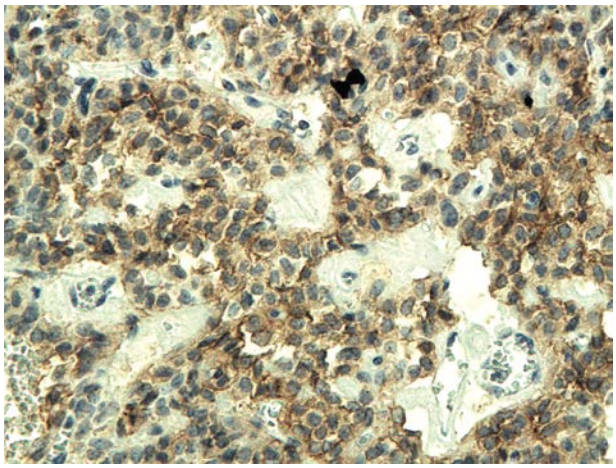

Fig. 2. Tumor cells displaying strong cytoplasmatic CD56 immunoexpression. Magnification 200×

Discussion

Solid pseudopapillary tumour accounts for 5% of cystic pancreatic neoplasms, and its origin has not yet been

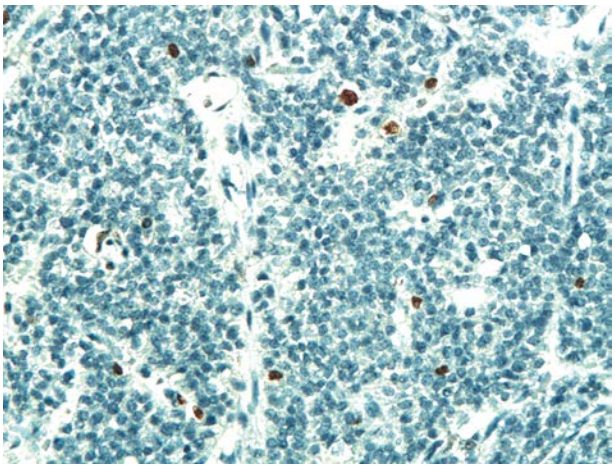

Fig. 3. Weak (below 1%) nuclear immunoexpression of Ki67 antigen in the tumor cells. Magnification 200×

clarified (it may originate from primitive pancreatic cells, e.g. acinar cell, ductal epithelium or from genital ridge angle-related cells). The size of the tumours ranges usually from 1.5 to 30 cm in diameter, and SPTs are located mainly in the pancreatic tail and body [21]. Some researchers suggest that the sex hormone may play a role in the development of this neoplasm, because the prevalence of SPT especially concerns women at the beginning of the reproductive period and progesterone receptors are present in more than 80% of cases, whereas oestrogen receptors are absent [22]. Machado *et al.* found that the onset of the disease occurs later in time and the tumour aggressiveness is greater among males [23]. The criteria indicating the tumour's malignancy are: angioinvasion, perineural invasion and deep invasion of the surrounding pancreatic parenchyma [21]. Recently, some other factors such as high mitotic rate, massive necrosis and sarcomatoid areas have been proposed as indicators of aggressive tumour behaviour [14]. Most patients present with unclear symptoms such as appetite reduction, abdominal discomfort and mild pain or nausea [24]. However, the tumour is asymptomatic and is found incidentally in 15% of cases during complementary imaging studies (CT or ultrasound scans of the abdomen) [5]. Laboratory values usually are not contributory, but a few reports show a raised level of CA 19-9 [25]. Endocrine and pancreatic enzyme markers are absent. Cytokeratin expression may be found only focally in less than 30% of cases [26].

The treatment of choice is aggressive resection, which may provide an overall 5-year survival rate of 95–98% [1]. Solid pseudopapillary tumour recurs or develops metastases after resection only in a small number of cases. Martin *et al.* report one recurrence in 17 patients who had complete resection, at median follow-up of 8 years [27]. Surgery should be considered even in the presence of local recurrence or metastases, and the resection of them should be performed at the time of primary resection [28]. However, there are available reports on a favourable response to radiotherapy in locally advanced unresectable disease and on successful chemotherapy in SPT with multiple hepatic metastases [29]. Solid pseudopapillary is usually large but rarely has invasion into adjacent organs, which is why the surgeon should aim for complete, en bloc resection including adjacent structures.

In conclusion, SPT is a largely benign tumour of relatively indolent behaviour. However, SPT of the pancreas is uncommon but should be considered in the differential diagnosis. Because patients may survive for a long time after surgery, up-to-date recommendations support targeting complete resection of the primary tumour as well as metastases. In cases of unresectable tumour or recurrent disease, adjuvant radio- or chemotherapy should be considered.

The authors declare no conflict of interest.

This study was supported by grant no. 503/8-145-01/503/01 from the Medical University of Lodz.

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Submitted: 30.07.2013
Accepted: 11.03.2014