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CASE REPORT

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Solid pseudopapillary neoplasm of the pancreas: a case report

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ABSTRACT

This paper presents a case report of a solid pseudopapillary neoplasm (SPN) of the pancreas, a discussion on the clinical-pathological, histopathological, and immunohistochemical picture, and a review of the literature regarding the occurrence of this type of cancer. The case of a 61-year-old woman with the presence of a lithium-cystic lesion of the body and tail of the pancreas was initially assessed by MRI as a pathological mass with the presence of abscesses. Double biopsy under EUS control was non-diagnostic. The patient underwent surgery to remove the body and tail of the pancreas together with the tumour and spleen.

Tumour size 15×10×8 cm lithium-cystic, grey-brown, with the presence of numerous calcifications and bone metaplasia, and stones in the pancreatic duct. In the histopathological picture, solid woven with the presence of pseudodimplants and pseudocystic areas with haemorrhages. Positive tests for NSE, vimentin, PR, CD56, and cyclin D1 were obtained in immunohistochemical (IHC) tests. The patient was discharged from the hospital in good general condition and is under gastroenterological control.

SPN is a rare cancer with low malignancy. The tumour most often occurs in teenagers or young women. Initially, it runs without ailments, until it is large. Then there is pain, nausea, and fever. The histopathological and cytological picture is suggestive, but it should be supported by research. SPN should be differentiated with neuroendocrine tumours (NENs) and acinar cancer and pancreatoblasoma. SPN generally has a good prognosis. Local relapses and distant metastases are rare.

Key words: pancreas, SPN, EUS, MRI, IHC

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Introduction

The classification of pancreatic tumours is based on the type of cell differentiation line, e.g. ductal, acinar, neuroendocrine, etc. [1] SPN is a pancreatic cancer in which there is no differentiation into a specific cell line, and it is considered an undefined tumour histogenesis. [3] Most, because about 90% of pancreatic cancers occurring in adults are ductile infiltrating cancers. Cystic and intracystic tumours constitute about 4–5%, neuroendocrine tumours about 3–4%, acinar and other cancers up to 3%, and SPN 0.9–2.7% [1]. The introduction of imaging tests has increased the detection of small solid pancreatic tumours and cystic. Like other pancreatic cancers in the early stages of hyperplasia is an incidental find. Symptoms are non-specific; usually discomfort or nausea. Tumour haemorrhage after injury can give a sign of acute abdomen. All cancerous markers are normal. There are also no endocrine disorders. 90% of SPN occur in teenagers and young women — the average age is 28 years. This cancer is rare in men. Other acceptable names for SPN include: solid-pseudopapillary tumour, solid-cystic tumour, papillary cystic tumour, solid and papillary epithelial neoplasm, and Frantz tumour [1].

Case report

A 61-year-old patient was admitted to the Gastroenterology Clinic because of abdominal pain increasing for three months, loss of appetite, and weight loss of 8 kg in two months, as well as low-grade fever. In the abdominal ultrasound performed, a hypoechoic area of approximately 3.5 cm in diameter was found. CT scan revealed a lithium-cystic lesion within the body and tail of the pancreas up to 14 cm in diameter. The performed laboratory tests revealed anaemia (E 3.98 × 10 ^ 6/uL, Hb 10.8 g/dL, Fe 22 ug/dl, TIBC 194 ug/dl, hyperuricemia during treatment, and CRP 17.0 mg/dl. Other results were within the norms. Comorbidities included: chronic pancreatitis, gout, epilepsy, condition after surgical

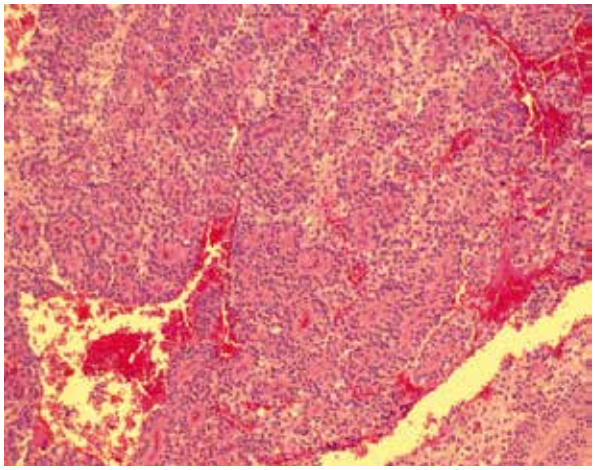

Figure 1. SPN solid weaving 10×

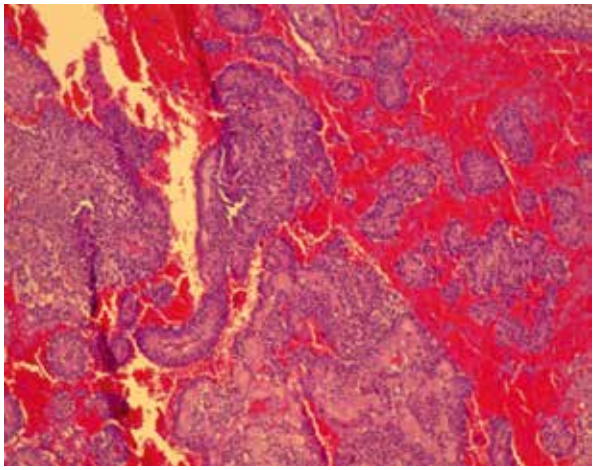

Figure 2. SPN haemorrhages and pseudocysts 10×

treatment of meningioma in 2013. The family history of malignancies included: breast, stomach, and lung in parents, grandparents, and siblings. During the stay, two endosonographic examinations were performed, during which material for cytological examination was collected using PCI. The material proved to be barely diagnostic, and no proliferative change was found. Due to persistent elevated inflammatory markers and abdominal pain, an MRI examination of the pancreas was performed. MRI showed a pathological suppurative mass up to 15 cm in diameter. There were no metastases to lymph nodes or adjacent organs. The patient was qualified for surgery. The course of surgery was without complications. The body and tail of the pancreas with the tumour, spleen, and peri-lymph nodes were removed. Macroscopically, a cross-sectioned 15×10×8 cm tumour with fragments of pancreatic parenchyma and spleen was seen in sections. A greyish-red tumour with solid fields and cysts as well as numerous calcifications and bone foci was observed.

The remaining pancreas and spleen tissue and lymph nodes remained unchanged. In the microscopic image, the SPN depicted the structures of lite (Fig. 1), pseudocystic (Fig. 2), and pseudopapillary (Fig. 3–5). The solid and pseudocystic part was composed of monomorphic cells with eosinophilic or clear cytoplasm with vacuoles, nuclei with small nucleoli, and evenly distributed chromatin. Mitosis was sporadic. The cells were characterised by poor cohesion. The cells entwined the glazing and myxoid stroma. Bone elements (Fig. 4), calcifications (Fig. 5), histiocytes, and multinucleated giant cells were seen. Part of the weaving comprised abscesses and necrotic lesions. The tumour was well delimited from pancreatic parenchyma. There was no vascular invasion or nerve invasion. The tumour weaving gave positive IHC for cyclin D1, CD56, CD10, PR, vimentin, and NSE (Fig. 6–13).

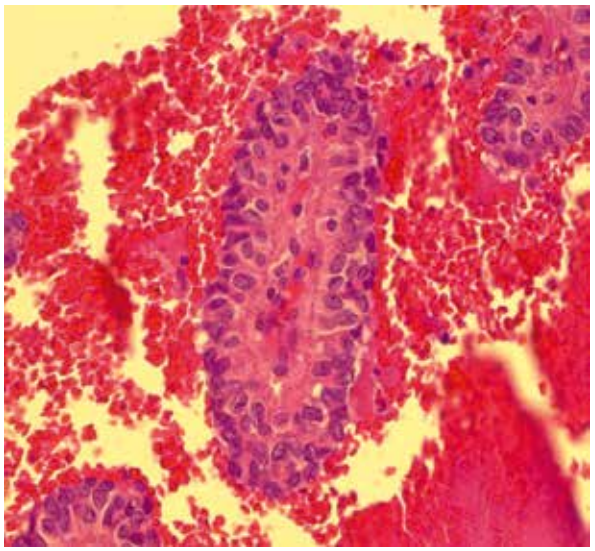

Figure 3. SPN calcified necrotic masses

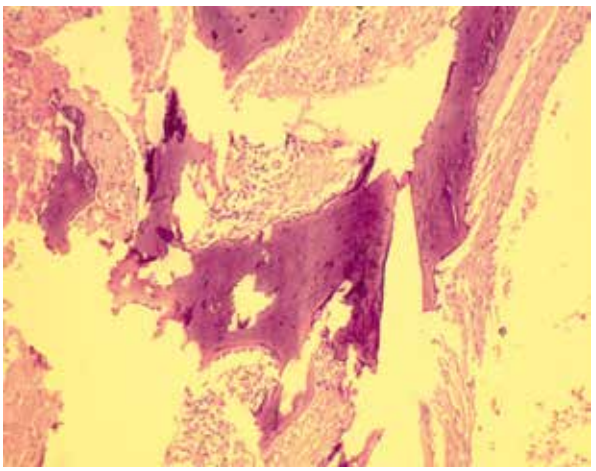

Figure 4. SPN bone elements 10×

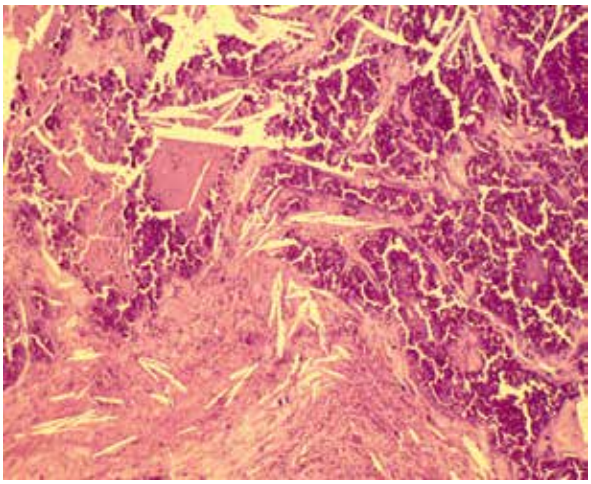

Figure 5. SPN pseudo wart 40×

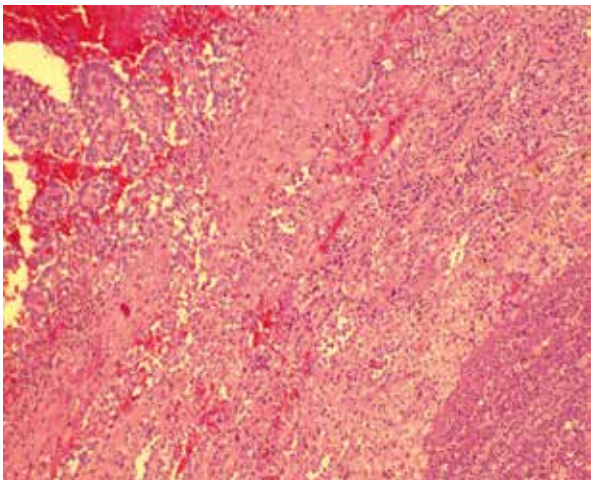

Figure 6. SPN necrotising lesions 10×

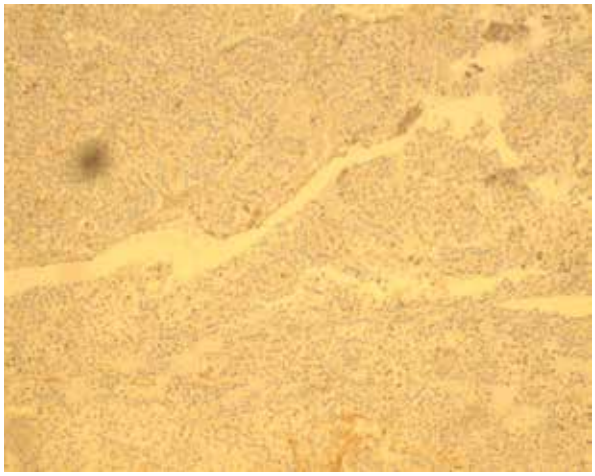

Figure 7. SPN CD 117 10×

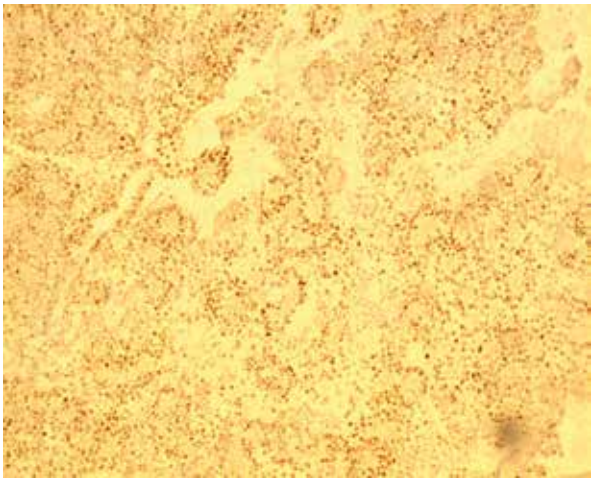

Figure 8. SPN Cyclic D1 10×

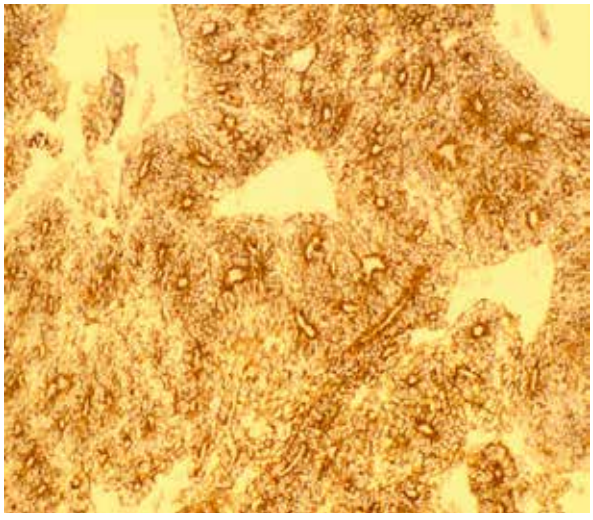

Figure 9. SPN CD 56 10×

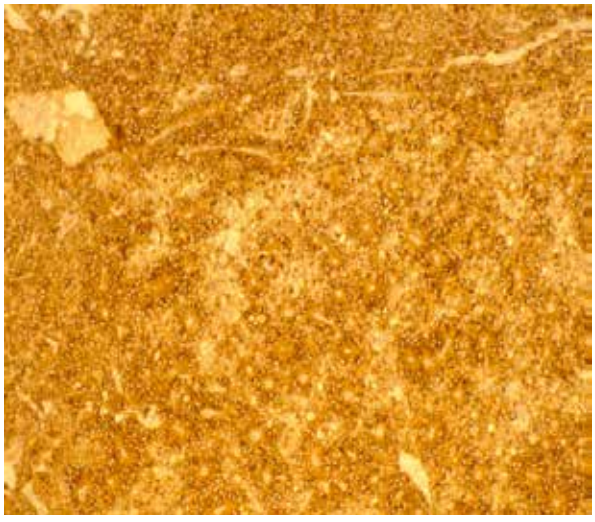

Figure 10. SPN CD 10 10×

Figure 11. SPN PR 10×

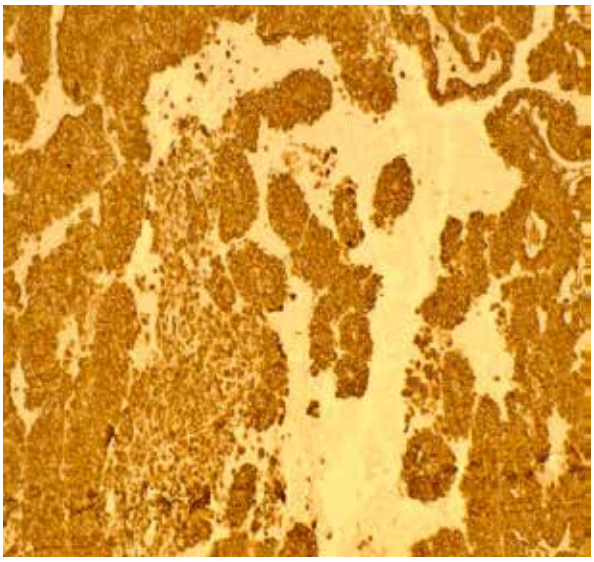

Figure 12. SPN Vimentin 10×

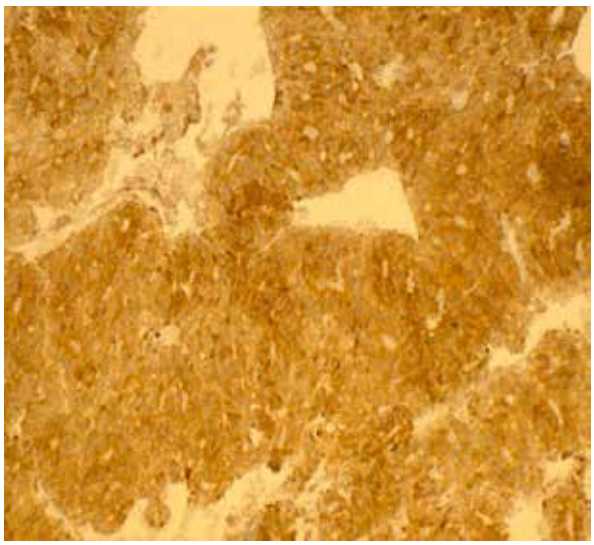

Figure 13. SPN NSE 10×

The tumour was removed entirely with a healthy tissue margin. Lymph nodes and spleen were without tumour invasion. The following diagnosis was made: solid pseudopapillary neoplasm of the pancreas. pT2N0

Sixteen days after the surgery, the patient was discharged home in good condition. Due to the histopathological diagnosis of pancreatic cancer, the patient remains under the control of an oncologist.

Discussion

Better imaging methods allow detection of pancreatic tumours in a stage of low advancement, when their size is 2-3 cm. Such small pancreatic tumours can be removed by laparoscopic method [2–12].

Solid pseudopapillary neoplasm (SPN) of the pancreas, also called solid and cystic pancreatic neoplasm (SCPN) or Frantz's tumour, is a form of pancreatic cancer of unknown aetiopathogenesis.

It is a cancer of the pancreatic exocrine portion, and although it can occur at different ages, it

mainly develops in adolescents and women between 20 and 30 years of age. It is very rare in men [5]. A relationship with gender and age suggests a hormonal effect on tumour development, but such disorders have not yet been reported in its course. The tumour development is affected by the CTNNB1 somatic mutation (B-catenin coding site). CTNNB1 somatic mutation on exon 3 is the only known mutation in SPN to date. Like most tumours, when they are small, they are detected incidentally in imaging (USG, MRI, CT) [13–18]. Large tumours cause abdominal pain, abdominal discomfort, low-grade fever weight loss, and anaemia, and haemorrhage to the tumour may present a symptom of acute abdomen. The cytological picture of cell smears obtained by thin-needle biopsy via endoscopic ultrasound fine-needle aspiration allows for precise diagnosis of SPN [9]. Typically, the cytological picture contains numerous cohesive, non-coherent, small monomorphic tumour cells lining thin-walled vessels. Testicles with evenly distributed chromatin, a small nucleolus, sometimes with a convex, bent nuclear membrane. Sometimes there are bizarre testicles. Similarly, to the histopathological picture, the cells have eosinophilic or clear cytoplasm. Sometimes high-grade foci with pleomorphic cells and numerous mitoses are present within the tumour, which indicates transformation into a highly aggressive tumour. The tumour may directly infiltrate the

duodenum, stomach, spleen, or portal vein. Sometimes there are hyaline globules that in ultrastructural image contain zymogen grains.

Conclusions

Solid pseudopapillary pancreatic neoplasm is a rare cancer of the pancreas. The cancer occurs in patients of a certain age and predominantly in females. The histopathological picture of the tumour and cytology are characteristic of the formation of solid fields, pseudopapillary and pseudocystic cysts, with the presence of inflammatory exudate, calcification, and multinucleated giant cells. Immunohistochemistry in the form of a reaction panel constitutes a complementary element of diagnostics. Currently, SPN is classified as a low-grade malignancy. The prognosis after complete tumour resection, even in the case of metastases and recurrences, is very good for young patients but slightly worse for older ones. Aneuploid SPNs are promising worse than diploid. Cancer detection at an early stage of development during research imaging may suggest a simple cyst or inflammatory lesions, createing the possibility of 100% cure with less invasive laparoscopic surgery.

Acknowledgments

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