medicinska revija
medical review

UDK: 616.34-006

Živojinov M. et al. ■ MD-Medical Data 2015;7(3): 249-252

Prikazi bolesnika/
Case reports

SOLID PSEUDOPAPILLARY NEOPLASM OF THE PANCREAS: FRANTZ'S TUMOR

SOLIDNI PSEUDOPAPILARNI TUMOR PANKREASA: FRANTZ-ov TUMOR

Mirjana Živojinov[1,2], Jelena Ilić Sabo[1,3], Tamara Bošković[1,3], Zorana Budakov[4,5]

Correspondence to:

Asist. dr **Jelena Ilić Sabo**
Bulevar Cara Lazara 5/52
21 000 Novi Sad;
+38163-303-585
jelena.ilic1705@gmail.com

[1] Center for Pathology and Histology, Clinical Center of Vojvodina, Novi Sad
[2] Department of Pathology, Medical Faculty, Novi Sad
[3] Department of Histology and Embryology, Medical Faculty, Novi Sad
[4] Center for Transfusiology, Clinical Center of Vojvodina, Novi Sad
[5] Department for Internal medicine, Medical Faculty, Novi Sad

Ključne reči

solidni pseudopapilarni tumor, pankreas, patohistološki nalaz, imunohistohemija;

Key words

solid pseudopapillary tumor, pancreas, histopathology analysis, immunohistochemistry

Abstract

Solid pseudopapillary tumor (SPT) of the pancreas is a very rare neoplasm, first described in 1956 by Frantz. It occurs predominantly in adolescent girls and women. The paper showed a case in 22-year-old female with solid pseudopapillary tumor of the pancreas. By macroscopic examination, it was described encapsulated fragment, size 10x6 cm, shiny and smooth surface with haemorrhage zones. As observed the cut section, there was soft tissue, well demarcated from the surrounding pancreas, in a form of cystic formation. On serial sections there were solid areas and cystic structures filled with coagulated blood. Histologicaly, tumor tissue was built of round cells with ovoid nuclei and clear cytoplasm. Nucleoli were inconspicuous. Mitotic figures were rare (less than 2 on 10 HPF). In some places papillae were observed, composed from partially hyalinised connective tissue. Groups of foamy macrophages were accumulated, and there were clusters of lipid crystals surrounded by foreign body giant cells. There was a haemorrhage in connective hyalinised capsule and signs of old haemorrhages. Positive immunoreactivity were for chromogranin, vimentin, CD56 and S100 and solid pseudopapillary cystic tumor of the pancreas with neuroendocrine differentiation is diagnosed.

INTRODUCTION

Solid pseudopapillary tumor (SPT) of the pancreas is a very rare neoplasm. It was first described in 1956 by Frantz, incorrectly diagnosed as a tumor of cells of Langerhans islets.(1) Until 1996, when the World Health Organization gave it the current name of solid pseudopapillary tumor of the pancreas, it has been referred as: „solid cystic tumor", „papillary cystic tumor", „solid and papillary epithelial tumor", „the Frantz tumor".(2) It is usually a low malignant potential neoplasm, with the incidence 1-2% of all exocrine pancreatic tumors. In approximately 13% of surgically resected cystic lesions of the pancreas, Solid pseudopapillary tumor was the cause.(3) It occurs predominantly in adolescent girls and women (mean age 35 years), while it occurrence in males is very rare.(4) Basically, the etiology is unknown. But, there are views that the tumor is connected to endocrine disbalance like overproduction of estrogen and progesterone and long-term taking of oral contraceptives. There is also view that tumor origins from acinar pancreatic cells.(5) The body and tail of pancreas are affected more frequently, than the head.(6) Grossly, it presents as large, round, solitary mass average size 3-10 cm, often fluctuant. It is usually encapsulated and well demarcated from the surrounding tissue. The cut surfaces reveal light brown solid areas, with zones of hemorrhage and necrosis, and cystic spaces filled with necrotic debris and blood. Occasionally, the hemorrhagic-cystic changes involve almost the entire lesion so that the neoplasm may be mistaken for a pseudocyst. (7) Histopathological analysis reveals solid monomorphic and

locally pseudopapillary patterns. In both patterns, the uniform round tumor cells, with oval/round nucleus and clear eosinophilic cytoplasm are arranged around hyalinized fibrovascular stalks. The cystic areas between papilla are often filled by blood. Also, groups of foamy macrophages that accumulate could be found, as well as clusters of lipid crystals surrounded by foreign body giant cells.[7,8] Immunohistochemical profile consider staining for: alpha-1-antitrypsin, vimentin, chromogranin, synaptophysin, S-100, CEA 19.9, cytokeratin. Metastases are rare, but common metastatic sites include regional lymph nodes, the liver, peritoneum and grater omentum. Presence of perineural invasion, angioinvasion, high grade of nuclear atypical cells and high mitotic index indicate malignant behavior.[9]

In general, the prognosis is good. Timely diagnosis and complete removal of the tumor leads to hilling in more than 95% patients.[9]

CASE REPORT

After clinical and MR diagnoses (Figure 1.) of the tumor of pancreas, the female 22-year-old patient had the part of the pancreas surgically removed. By grossly examination it is described as encapsulated round fragment, size 10x6 cm, glossy and smooth surface, bruised by blood. At the cut section, soft, pallid, cystic, well demarcated formation could be seen. On serial sections there are solid tissue areas and small cystic forms filled by the coagulated blood. Pathohistological analysis reveals tumor tissue composed of solid areas which consist of round tumor cells with oval nucleus, without nucleolus, faintly acidophilic cytoplasm. Mitosis are rare (less than 2 per 10 HPF). Occasionally papilla could be seen (Figure 2.) composed of fibromuscular stroma with partly hyalinized tissue, surrounded by tumor cells. Cholesterol crystals are present (Figure 3.), as well as the foamy macrophages and the clusters of lipid crystals surrounded by foreign body giant cells. There is a hemorrhage in connective hyaline capsule that encloses tumor tissue from the surrounding pancreas tissue, as well as signs of old hemorrhage (hemosidherophages). We obtained positive immunohistochemical profile of cromogranin, vimentin (Figure 4.), CD56 and S100.

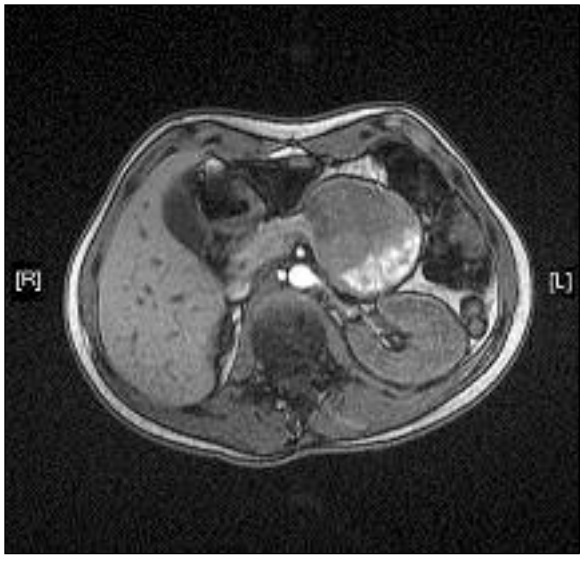

***Figure 1**. MRI MRCP of abdomen;*

Solid pseudopapillary cystic tumor of the pancreas with neuroendocrine differentiation (Figure 5.) is diagnosed.

DISCUSSION

Solid pseudopapillary tumor of the pancreas is very rare neoplasm with low malignant potential, by 2009 was recorded only 718 cases.[2] There is a noticeably higher prevalence in Asians and African Americans.[9,10] The tumor occurs predominantly in women. The most common ages at which the tumor occurs ranges from 22 to 35 years.[2,4,10] Our patient was 22 years old, which coincides with the majority of literature data.[2,4,10] However, there are presented the cases of SPT in children aged between 11 and 14,[9] which is slightly younger age than the average age of incidence of the neoplasm.

The etiology of the tumor is not known with ceirtanty. One view is that an disbalance in the production of estrogen and progesterone leads to a solid pseudopapillary tumor of the pancreas.[10] Recent studies indicate a mutation in exon 3, β-catenine gene could be cause of SPT.[11] A similar mutation occurs in pancreatoblastoma, and there is a hypothesis that it is possible that these two tumors originate from stem cells.(11)

Size of the tumor in our case is 10x6 cm. Studies on tumor size showed that it vary between 2,5 cm,[12] and 24 cm,[3] while the average size is between 5 and 10 cm.[2] Size is an important factor. Larger tumor in addition to structure and histopathological features indicates SPT, while the small size of the tumor may indicate a wrong diagnosis. [12]

The most common location of tumors, as reported in the literature, is the body and tail of the pancreas.[3,7] In our case the tumor was localized in the area of the body towards the tail of the pancreas.

Grossly, shape of tumor was round in more than half of cases,[7] which is also the result of grossly inspection in our case.

The typical histological picture of pancreatic SPT reveals a solid areas constructed of round cells with regular, oval nuclei and clear cell cytoplasm.[7,13] The presence of papillae built of fibrovascular stroma with hyalinized tissue and cholesterol crystals are almost regular finding.[7,13] Rare cases of this tumor with spindle cells were found, and it may significantly affect the setting misdiagnosis and faulty therapeutic procedures.[12] A significant contribution in such cases has immunohistochemical findings, which may indicate the origin of the tumor. SPT is characterized by positive immunostaining with vimentin, CD10, CD56 i alpha-1-antitrypsin, and may also show positive staining for cytokeratin, neuron specific enolase and synaptophisin.[1,2] If it is difficult to make a definitive diagnosis based on histological findings, immunohistochemistry is the one that leads to it.[12]

In general, SPT of the pancreas is a low-grade malignancy, rarely present with local invasion of adjacent organs, perineural and/or intravacular invasion and metastasis in lymph nodes.[3,14] Mitotic index is low, and nuclear atypia is rare.[3,15] However, although low malignant tumor, with untimely diagnoses, fatal cases were reported.[3]

SPT of the pancreas can also occur in association with other tumors (e.g. kidney hamartoma) or methabolic disorders,[5] which further complicates the situation and treatment of the patient.

There are recorded cases of relapse of SPT of the pancreas.[4] Although they were considered possibilities of further chemotherapy and radiotherapy, the final statement was a radical surgical procedure with postoperative monitoring of patient.[4]

Solid pseudopapillary neoplasm of the pancreas is very rare neoplasm. It is predominantly low malignant potential. Proper diagnosis of these tumors is extremely important, considering well-timed and complete resection, the healing of this neoplasm is possible. The progression of the tumor as directly spreading and metastasis are associated with malignancy. Aggressive surgical in early disease, leads to complete healing in over than 95% of patients with solid pseudopapillary tumor of pancreas.

Declaration of interest

The authors have no conflict of interest to declare.

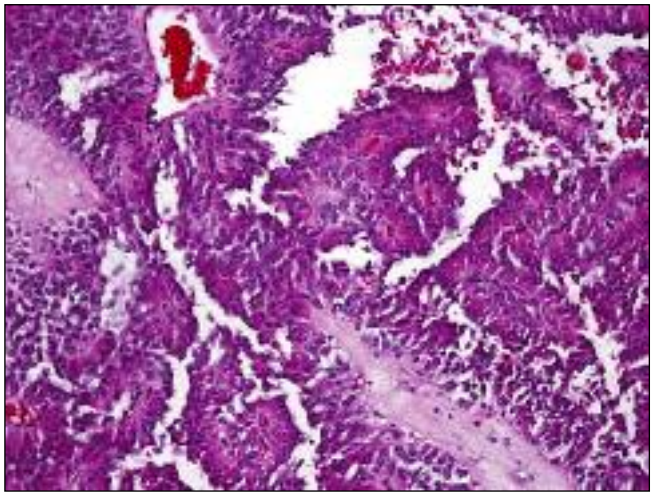

***Figure 2**. Solide Pseudopapillary tumor of pancreas, papillary part (Hematoxylin-eosin stain ; magnification x200)*

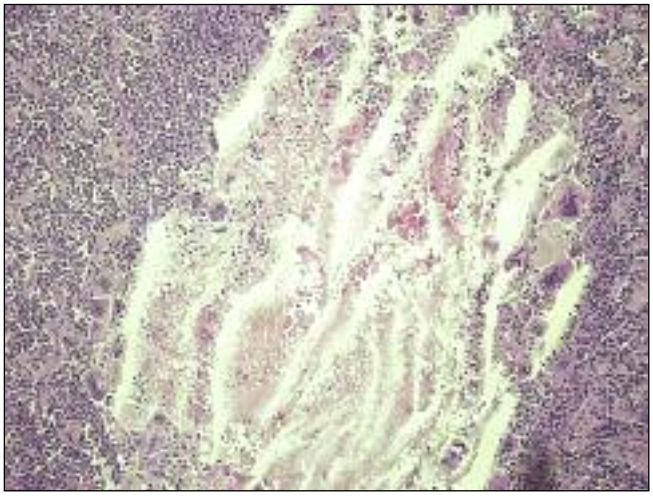

***Figure 3**. Solid Pseudopapillary tumor of pancreas, cholesterol crystals (Hematoxylin-eosin stain ; magnification x200)*

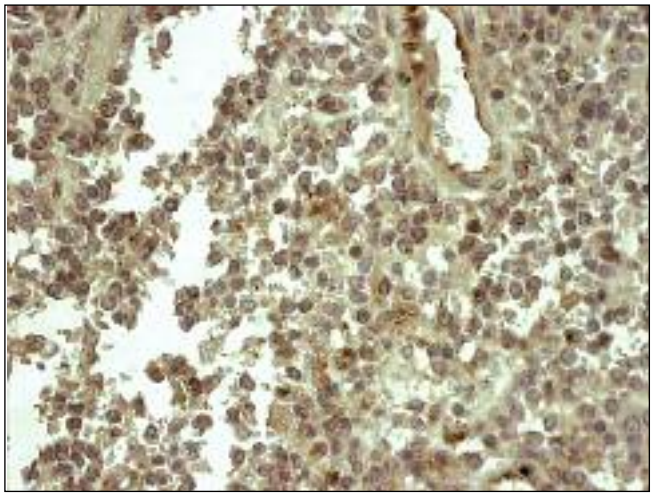

***Figure 4**. Solid Pseudopapillary tumor of pancreas,(imunochistochemical stain, vimentin; magnification x200)*

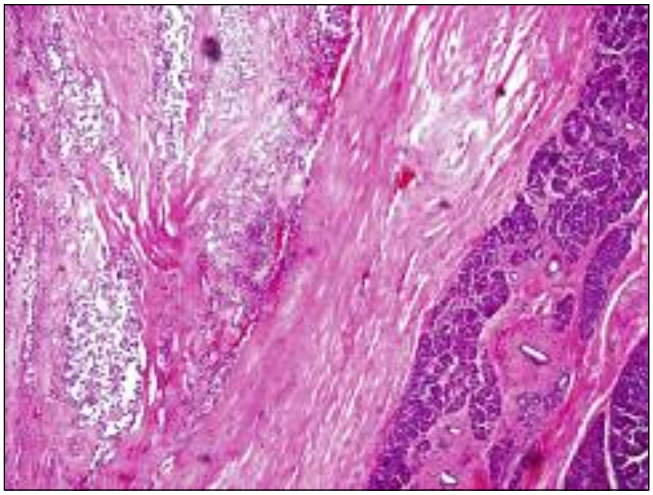

***Figure 5.** Solid Pseudopapillary tumor of pancreas, (Hematoxylin-eosin stain, neuroendocrine component of tumor; magnification x50)*

Sažetak

Solidni pseudopapilarni tumor pankreasa je veoma retka neoplazma opisana od strane Frantz-a 1956. god. Najčešće se javlja kod adolescentkinja i žena starosti oko 35 godina. U radu je prikazan slučaj pacijentkinje starosti 22 godine sa solidnim pseudopapilarnim tumorom pankreasa. Makroskopskim pregledom opisan je inkapsulisan fragment veličine 10x6 cm, sjajne i glatke površine, podlivene krvlju. Na preseku se uočava meko tkivo, jasno ograničeno od okolnog tkiva pankreasa u vidu cistične formacije. Na serijskim rezovima se nalaze solidna područja tkiva i manje cistične strukture ispunjene koagulisanom krvi. Patohistološkom analizom tumorsko tkivo je sagrađeno od solidnih plaža koje čine okrugle tumorske ćelije ovalnih, pravilnih jedara, bez uočljivih jedaraca ,oskudne acidofilne citoplazme. Mitoze su retke (manje od 2 na 10 HPF). Mestimično se uočavaju papile čija je stroma sagrađena iz vezivno-vaskularnog, delimično hijalinizovanog tkiva. Prisutni su holesterolski kristali, makrofagi penušave citoplazme, džinovske ćelije tipa oko stranog tela kao i okruglasta hijalina područja amorfnog izgleda. Prisutno je krvarenje u vezivnoj hijalinoj kapsuli koja tumorsko tkivo ograđuje od okolnog tkiva pankreasa, kao i znaci starog krvarenja Dobijen je pozitivan imunohistohemijski profil na chromogranin, vimentin, CD56 i S100, te je postavljena dijagnoza solidnog pseudopapilarnog (cističnog) tumora pankreasa sa neuroendokrinom diferencijacijom.

REFERENCES

1. Li L, Li J, Hao C, Zhang C, Mu K, Wang Y,et all. Immunohistochemical evaluation of solid pseudopapillary tumors of the pancreas: The expresion pattern of CD99 is highly unique. Cancer letters. 2011; 310:9-14.

2. Bostanoglu S, Otan E, Akturan S, Hamamci EO, Bostanoglu A, Gokce A, et all. Frantz's tumor (solid pseudopapillary tumor) of the pancreas. A case report. J Pancreas. 2009; 10(2):209-11.

3. Goh BKP, Tan YM, Cheow PC, Chung YFA, Chow PKH, Wong WK, et all. Solid pseudopapillary neoplasm of the pancreas:an updated experience. J Surg Onc. 2007; 95:640-4.

4. Murawa D, Zasadowski Z, Kruszyna L. A second relapse of a solid pseudopapillary tumor of the pancreas: case description. Polish Jurnal of Surgery. 2008; 80(1):46-50.

5. Chen X, Zhou GW, Zhou HJ, Peng CH, Li HW. Diagnosis and treatment of solid-pseudopapillary tumors of the pancreas. Hepatobilliary Pancreat Dis Int. 2005; 4(3):456-9.

6. Morgan DE. Cystic lesions of the pancreas. Seminars in Roentgenology. 2009; 255-65.

7. Lee JH, Yu JS, Kim H, Kim JK, Kim TH, Kim KW, et all. Solid pseudopapillary carcinoma of the pancreas: differentiation from benign solid pseudopapillary tumour using CT and MRI. Clinical Radiology. 2008; 63:1006-14.

9. Qahtani SA, Gudinchet F, Laswed T, Schnyder P, Schmidt S, Osterheld MC, et all. Solid pseudopapillary tumor of the pancreas in children: typical radiological findings and pathological correlation. Clinical Imaging. 2010; 34:152-6.

10. Shabbir A, Hai-Yi CT, Leow CK. Review of solid-pseudopapillary tumour of the pancreas with case illustration. Surgical practise. 2006; 10: 148-53.

11. Graziano A, Campisi M. Tumour (Frantz's) solid-pseudopapillary of the pancreas:review and case report. Acta Chirurgica Mediterranea. 2009; 25:37-43.

12. Hyang JG. A spindle cell predominant pancreatic solid-pseudopapillary tumor. Yonsei Med J. 2008; 49(4):672-5.

13. Kamat RN, Naik LD, Joshi RM, Amrapurkar AD, Shetty TS. Solid pseudopapillary tumor of the pancreas. Indian J Pathol Microbiol. 2008; 51(2):271-3.

14.Karamarković AR, Micev M, Ćulafić Đ, Popović N, Đukić N. Solidni cistični pseudopapilarni tumor pankreasa. Srp Arh Celok Lek. 2004; 132(11-12):431-4.

15. Mirković D, Karamarković A. Solidni pseudopapilarni tumori pankreasa. Vojnosanit Pregl. 2005; 62(10):763-7.

The paper was received on 23.06.2015. Revised on 24.06.2015. Accepted on 26.06.2015.