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Single center experience in the diagnostics of solid pseudopapillary (Gruber-Frantz) tumor of the pancreas – description of two cases and literature review

Doświadczenia własne w diagnostyce guza Gruber-Frantza – opis dwóch przypadków i przegląd piśmiennictwa

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Key words

pancreas, solid pseudopapillary tumor, Gruber-Frantz tumor

Słowa kluczowe

trzustka, lity pseudobrodawkowaty guz, guz Gruber-Frantza

Summary

Recent development of radiological techniques and growing number of patients studied by different scanners results in a great number of asymptomatic tumors accidentally discovered what can input that radiological symptomatology of some very rare tumors should be overworked.

The retrospective review of the medical records and images of 2 patients who underwent surgery for solid pseudopapillary tumor (SPT) of the pancreas between December 2002 and December 2013 was performed.

In both patients with no or mild symptoms the pancreatic lesions were revealed in the screening abdominal ultrasound followed by computer tomography (CT). In 54-year-old woman the pancreatic body lesion (17 x 17 mm) has presented as hypoechoic, homogeneous solid tumor, isodense, non-enhancing mass in the CT study, mimicking malignant tumor.

In 30-year-old woman lesion in the pancreatic head presented as inhomogeneous cystic tumor (35 x 23 mm) with peripheral nodule-like enhancing solid component in CT study (more typical for Gruber-Frantz tumor).

Intraoperative pathomorphological examinations in both cases excluded pancreatic cancers and the local excisions of the tumors were performed. In both cases the final diagnosis was SPT (Gruber-Frantz tumor).

Streszczenie

Rozwój technik radiologicznych, który się ostatnio dokonał, wraz z rosnącą liczbą pacjentów badanych powodują wzrost wykrywalności bezobjawowych incydentalnych guzów trzustki, co może spowodować nowe podejście do symptomatologii radiologicznej niektórych rzadkich guzów.

Opracowanie przedstawia retrospektywny przegląd opisów medycznych i obrazów radiologicznych dwóch pacjentek poddanych pomiędzy grudniem 2002 i grudniem 2013 roku zabiegom chirurgicznym z powodu litych pseudobrodawkowatych guzów trzustki.

U obu pacjentek z miernie wyrażonymi objawami klinicznymi po badaniu usg ujawniającym zmiany w trzustce wykonano tomografię komputerową (TK). U 54-letniej kobiety zmiana w trzonie trzustki (17 x 17 mm) prezentowała się jako jednorodnie hipoechogenny, lity guz, niewzmacniający się w badaniu TK, imitujący guz złośliwy.

U 30-letniej kobiety zmiana w głowie trzustki prezentowała się jako niejednorodny echogenicznie torbielowaty guz (35 x 23 mm) w usg z brzeżną, guzkową wzmacniającą się komponentą w badaniu TK, mogący odpowiadać guzowi Gruber-Frantza.

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W obu przypadkach śródoperacyjne badanie histopatologiczne wykluczyło raka trzustki, umożliwiając wykonanie regionalnego wycięcia guzów. Także w obu przypadkach ostateczną diagnozą był lity pseudobrodawkowaty guz trzustki (SPT, guz Gruber-Frantza).

INTRODUCTION

Rapid developement of radiology and growing number of patients studied by US, MR and CT scanners results in a great number of asymptomatic tumors accidentally discovered. One can suppose that radiological symptomatology of some very rare tumors should be overworked. Solid pseudopapillary tumor (SPT) or neoplasm (SPN), also named papillary epithelial neoplasm or Frantz tumor (also Gruber-Frantz or rarely Hamoudi tumor) is a low-grade or borderline epithelial pancreatic malignancy and has been reported in 0.9-2.7% of all neoplasms of the pancreas often characterized as a rather large one, presenting mixed morphology (1).

PATIENT 1 – CASE DESCRIPTION AND IMAGING

In 54-year-old woman presenting no symptoms nor clinical disorders ultrasound of the abdomen revealed a 17 x 17 mm solid hypoechoic lesion in the body of pancreas. The absence of typical sonographic hiperechogenity behind the lesion could suggest a solid character of the tumor (fig. 1). Routine blood tests, including liver function studies and ESR, were essentially normal. Clinical laboratory tests concerning various tumor markers were unremarkable.

CT (Hi Speed, General Electric) including native pre-contrast scans and two phases after contrast administration confirmed the presence of the lesion in the body of pancreas depicting hypodense mass well-demarcated from pancreatic parenchyma and retroperitoneal tissue surrounding the pancreas (fig. 2). There was no significant enhancement of the lesion after intravenous administration of contrast medium in arterial phase (fig. 3). The attenuation of the tumor varied from +17HU to +40HU. On the basis of imaging the suspicions of malignancy was concluded. There was neither peripancreatic lymph nodes enlargement nor liver metastases. A fine needle aspiration biopsy was not made and the patient was sent to surgery.

In December 2002 local excision of the tumor was performed. In the pathomorphologic examination the solid-cystic-papillary epithelial neoplasm was found with the expression of chromogranin, synaptophysin and without signs of cytological atypia, suggesting Gruber-Frantz tumor. Postoperative follow-up with US and CT examinations revealed neither evidence of local reccurence of the tumor nor metastases and the patient remains asymptomatic 11 years after operation.

PATIENT 2 – CASE DESCRIPTION AND IMAGING

A 30-year-old woman in a good overall health with no remarkable past family or personal medical history, was admitted to hospital due to mild epigastric discomfort for a few weeks after ultrasound examination performed in the ambulatory service disclosing lesion in the region of pancreatic head. She reported nausea, diarrhea and headache. Clinical examination revealed

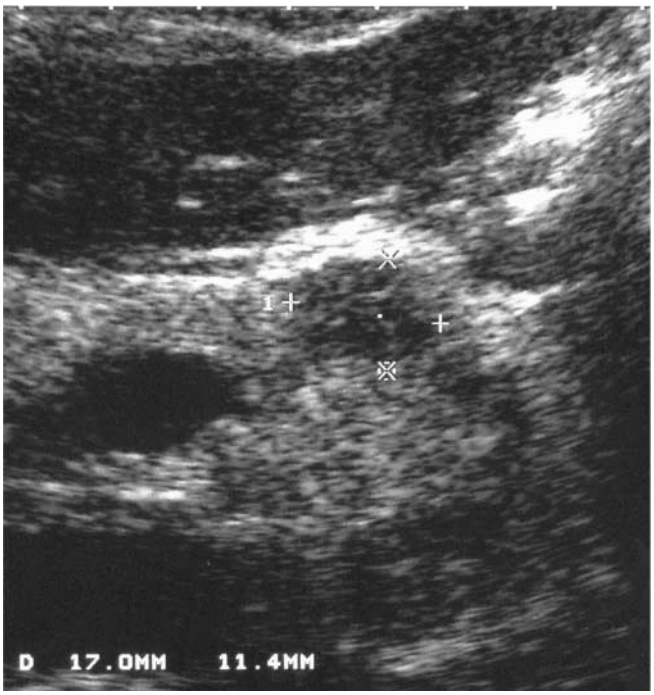

Fig. 1. Solid hypoechoic mass of 17 x 11 mm diameter with no distal enhancement, known to be typical for cystic tumors.

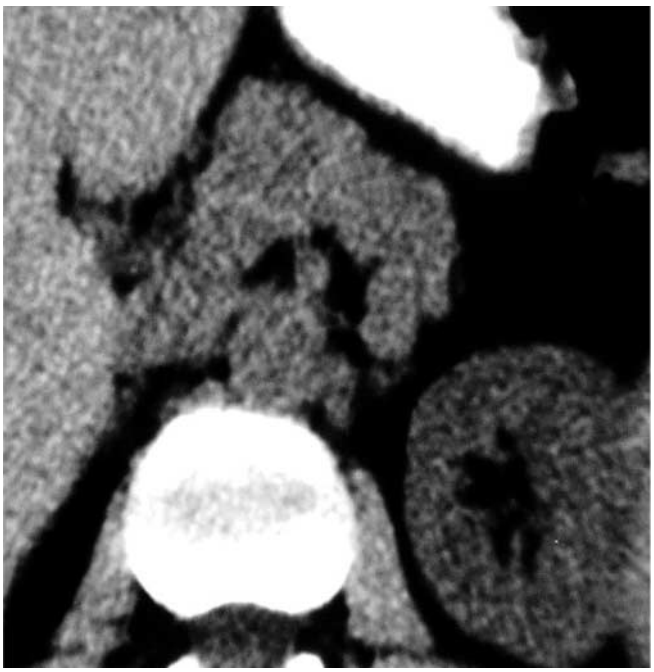

Fig. 2. Native CT scan presents small deformation of pancreatic margins with the same density as the parenchyma.

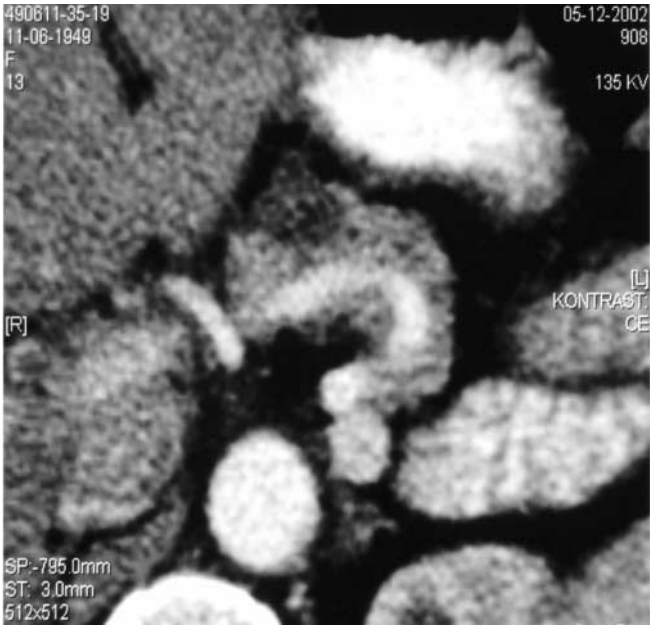

Fig. 3. Post contrast CT scan demonstrates small hypovascular tumor of the pancreas.

no fever, pallor, icterus, lymphadenopathy or a palpable mass. All laboratory investigations including tumor markers were within normal limits.

Besides hepatomegaly within the abdominal cavity and uterine myofibromas in the pelvic cavity, all other abdominal and pelvic organs were normal. The chest X ray was normal.

Pre-contrast CT (64 MDCT General Electric scanner) confirmed the localization and presence of the abnormal hypoattenuating mass (25-35 HU) with a size approximately 35 × 23 mm with focal peripheral calcification (fig. 4). Inhomogeneous, mainly nodule-like peripheral enhancement (80-100 HU) was found. The strongest enhancement was observed in the area adherent to the unchanged part of the pancreatic body (fig. 5, 6).

No dilatation of the biliary duct nor pancreatic duct, vessel invasion, calcification, infiltration of adherent pancreatic parenchyma and surrounding vessels was observed. The margin between the lesion and the pancreas and the surrounding fatty tissue was unclear.

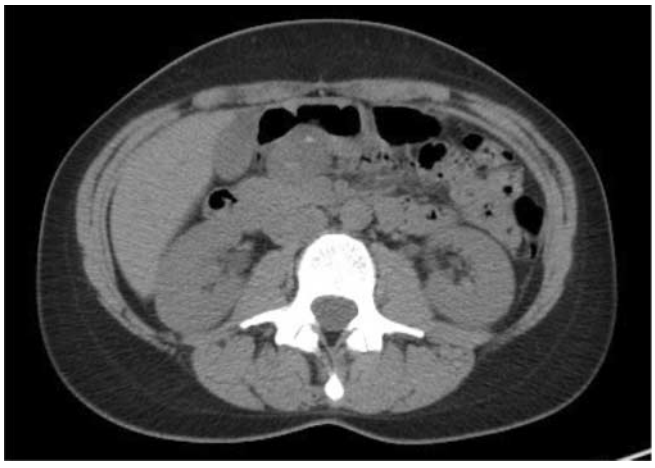

Fig. 4. Pre-contrast CT.

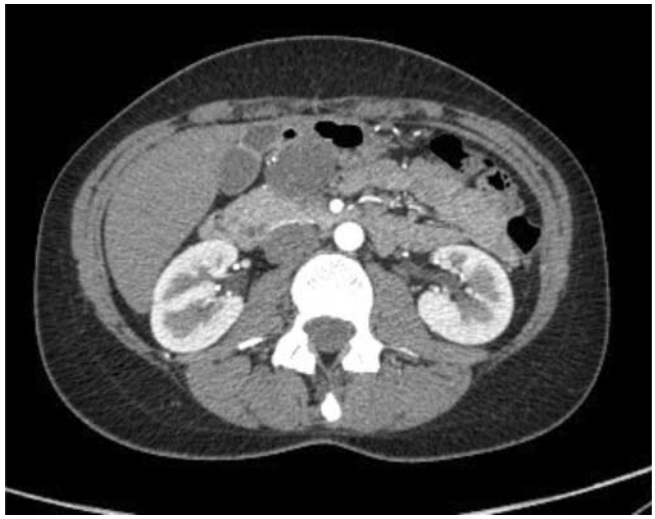

Fig. 5. Post-contrast CT – arterial phase.

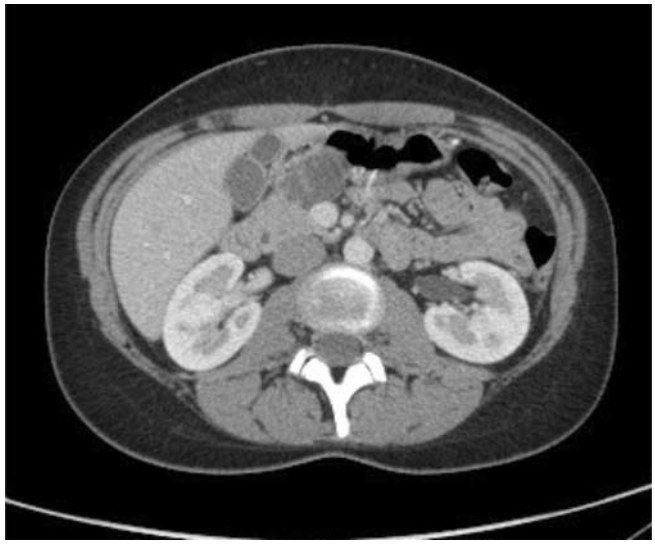

Fig. 6. Post-contrast CT – venous phase.

Postoperative follow-up with US and CT examinations revealed neither evidence of local regrowth of the tumor nor lymphadenopathy, nor liver metastases and the patient remains asymptomatic 3 years after surgery.

DISCUSSION

Solid pseudopapillary tumor of the pancreas (SPT) was first described in 1959 by Frantz (2) as papillary tumor of the pancreas, and is also called solid and cystic tumor or solid and papillary epithelial neoplasm of the pancreas. It is an uncommon primary low grade malignant neoplasm of the pancreas that constitutes less than 1% of all pancreatic tumors and occurs in great majority of cases in young non-Caucasian women (between the ages of 20 and 30 years the ratio of females to males is 14:1) with predilection for Asian and black race according to some authors (3). Ninety percent of cases present in individuals between the age of 15 and 35 years (3, 4). The etiology of SPT still remains unclear. Some evidence suggest that SPT actually rep-

resent an embryonic tumor that arises from pluripotential pancreatic stem cells capable of endocrine or exocrine differentiation because of the variety of markers from various pancreatic cell types (5). It was reported that the high proliferative index assessed by immunohistochemical staining for Ki-67 may predict poor outcome of malignant SPT (5). In literature more than 700 cases of SPT have been reported so far (2-4). Despite the underlined fact that pathologically SPTP are fairly common misdiagnosed as nonfunctional islet cell tumors, cystadenomas, or cystadenocarcinomas (6), it remains no doubts that SPT is a very rare neoplasm. Due to its slow and asymptomatic growth the tumor is usually large at the moment of diagnosis with a mean diameter of 9.3 cm (1, 3).

In patients with SPT, usually complaining of non-characteristic upper abdominal discomfort or pain, an examination may reveal a palpable large mass typically situated in the epigastrium or left upper quadrant. Rarely an acute abdomen may be present when there is an hemorrhage from or into the tumor (7). Solid pseudopapillary tumors are localized in the body-tail region of the pancreas in up to 64%. The remaining 36% of the tumors are found in the head (2), however a few cases of the tumor located outside the pancreas, usually arising from heterotopic pancreatic tissue have also been described (8).

Radiologic imaging techniques like a plain film radiography, an US, a contrast enhanced CT and MRI of the abdomen are performed for establishing the diagnosis. The radiologic findings in SPTP are not specific. Plain film radiographs may reveal displacement of adjacent organs (if the tumor is large enough) and occasionally calcification which is not common. Sonography and CT present well-demarcated large masses in the pancreas containing variably sized cysts with a honeycomb-like pattern. The structure of the mass most frequently is mixed solid and cystic, sometimes mostly cystic and rarely only solid, generally depending on the degree of hemorrhage or necrosis. The CT numbers of cystic parts, analysed by Friedman et al. (9), were higher than CT numbers typical for fluid, suggesting the presence of blood which allows to differentiate SPT from mucinous cystic neoplasms (9). MR usually reveals a well-demarcated lesion with areas of high signal intensity on T1- or T2-weighted images corresponding to regions of hemorrhagic necrosis and cystic parts respectively (1).

Calcifications are secondary to previous necrosis or hemorrhages. Non-encapsulated lesions have also been reported, but they were probably detected in an early stage, before formation of the capsule (1). The capsule composed of fibrous connective tissue has a unique morphology in all radiological techniques. It is usually hypoechoic in ultrasound examination, hypo- or isodense in CT and hypointense in T1- and T2-weighted MR images (1, 4). According to their name, tumors are usually solid but often, especially in large lesions, cavities filled with necrotic masses or blood as a result of acute or remote hemorrhages are also noted. In an extreme situation they may imitate a classic cystic pancreatic lesion and histopathological evaluation is crucial to prove their true nature (1). Due to their soft consistency, tumors rarely cause obstruction of bile and pancreatic ducts, which may be examined by MRCP. Despite the relatively benign nature of the tumor, solid parts are usually enhanced after contrast injection in both CT and MR, with an early “wash-in” effect during the dynamic phase (10). Progressive, heterogeneous enhancement in portal venous and parenchymal phases but less than the surrounding and unchanged part of the pancreas has been also observed (10). Unfortunately it can be stated only for the typical, rather large size of Gruber-Frantz tumor. The smaller tumor the less specific image presents. In our case SPT was discovered as a 17 mm solid tumor. This suggest, that each lesion starts as a small solid focus and then degenerates during its growth. In differential diagnosis any malignant pancreatic tumors could not be excluded (neither primary nor secondary cancer) considering the patient age. A decision of performing guided percutaneous biopsy was not made because, neither the histopathology results from the fine needle biopsy, most of the patients with pancreatic tumors when no infiltration was found in imaging methods, will go to surgery (6). It usually consists of a distal pancreatectomy when the tumor is located in the body or tail of the pancreas or duodenopancreatectomy if the lesion arises from the pancreatic head. Local excision with complete removal was seemed to be reasonable as an initial procedure only in pediatric-aged patients due to good prognosis in the contrary to the older patients when an initial pancreaticoduodenectomy is recommended (6, 7, 10).

Because high survival rates can be achieved in most cases, the aggressive treatments even in metastatic disease are performed. Even the selective internal radiotherapy was reported to be used after extensive tumor resection when the hepatic recurrence was diagnosed (6).

Preoperative diagnosis in small, hormonally non-functional pancreatic tumors is difficult and made usually in histopathological examination. In large inoperable tumors biopsy can be useful. Nowadays, due to significant continuous progress in diagnostic imaging one can find as small focal lesions as few milimeters. It is possible even in such diagnostically difficult organ as pancreas. It can change the strategy of treatment in elderly patients in the favor of local excision of pancreatic solid tumors. But first any radiological criteria should be overthink again.

BIBLIOGRAPHY

1. Kim SY, Park SH, Hong N et al.: Primary solid pancreatic tumors: recent imaging findings updates with pathology correlation. Abdom Imaging 2013; 38: 1091-1105.

2. Vassos N, Agaimy A, Klein P et al.: Solid-pseudopapillary neoplasm (SPN) of the pancreas: case series and literature review on an enigmatic entity. Int J Clin Exp Pathol 2013; 6: 1051-1059.

3. Mao C, Guvendi M, Domenico DR et al.: Papillary cystic and solid tumors of the pancreas: a pancreatic embryonic tumor? Studies of three cases and cumulative review of the world's literature. Surgery 1995; 5: 821-828.
4. Papavramidis T, Papavramidis S: Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. Journal of the American College of Surgeons 2005; 6: 965-972.
5. Yang F, Jin C, Long J et al.: Solid pseudopapillary tumor of the pancreas: a case series of 26 consecutive patients. Am J Surg 2009; 2: 210-215.
6. Krug S, Bartsch DK, Schober M et al.: Successful selective internal radiotherapy (SIRT) in a patient with a malignant solid pseudopapillary pancreatic neoplasm (SPN). Pancreatology 2012; 5: 423-427.
7. Jeng LB, Chen MF, Tang RP: Solid and papillary neoplasm of the pancreas: emphasis on surgical treatment. Arch Surg 1993; 128: 433-436.
8. Tornoczky T, Kalman E, Jakso P et al.: Solid and papillary epithelial neoplasm arising in heterotopic pancreatic tissue of the mesocolon. Journal of Clinical Pathology 2001; 3: 241-246.
9. Friedman AC, Lichtenstein JE, Dachman AH: Cystic neoplasms of the pancreas: radiological-pathological correlation. Radiology 1983; 149: 45-50.
10. Horisawa M, Niinomi N, Sato T et al.: Frantz's Tumor (Solid and Cystic Tumor of the Pancreas) With Liver Metastasis: Successful Treatment and Long-Term Follow-Up. Journal of Pediatric Surgery 1995; 5: 724-726.

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