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Papillary-cystic neoplasm of the pancreas – a case report

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Papillary-cystic neoplasm of the pancreas is rare. It affects young women in the third decade of life. The prognosis is good even in advanced cases, in contrary to other pancreatic neoplasms. Radical surgical resection is the treatment of choice. The authors present a margins 20-years old woman with papillary-cystic neoplasm of the pancreas. The tumor was completely resected with its capsule and with clear – of surrounding tissue. The patient is under regular follow-up and has no evidence of disease after 22 months.

Brodawkowato-torbielowaty nowotwór trzustki – opis przypadku

Brodawkowato-torbielowaty nowotwór trzustki występuje rzadko. Dotyczy głównie młodych kobiet w trzeciej dekadzie życia. Choroba często rozpoznawana jest przypadkowo, podczas badań lub zabiegów operacyjnych przeprowadzanych z innych powodów. Guzy w momencie rozpoznania są duże, średnio o średnicy 10 cm. Objawy chorobowe, z którymi zgłaszają się chorzy, są niespecyficzne; głównie jest to obecność niebolesnego wyczuwalnego guza w śródbrzuszu. W badaniach obrazowych stwierdza się zwykle dobrze ograniczony lity, lito-torbielowaty lub torbielowaty twór, zlokalizowany najczęściej w trzonie lub ogonie trzustki. Histogeneza tego typu nowotworów jest niejasna i nie udało się do tej pory jednoznacznie określić linii komórkowej, stanowiącej punkt wyjścia. W odróżnieniu od innych, częstych nowotworów trzustki rokowanie jest dobre, nawet w przypadkach zaawansowanych. Na około 340 opisanych przypadków tylko u 40 chorych stwierdzono naciek okolicznych narządów lub przerzuty odległe. Mimo stwierdzenia inwazyjnej postaci choroby okres przeżycia był długi, a tylko 3 pacjentów zmarło z jej powodu. Leczeniem z wyboru jest radykalny zabieg operacyjny. Autorzy przedstawiają przypadek choroby u 20 letniej chorej, u której na podstawie przedoperacyjnych badań obrazowych rozpoznano guza głowy trzustki. Wykonano całkowite wycięcie guza z marginesem tkanek zdrowych. Chora pozostaje pod stałą kontrolą w naszym ośrodku i w okresie 22 miesięcy od operacji nie stwierdza się cech nawrotu lub rozsiewu choroby.

Key words: pancreas, papillary-cystic neoplasm
Słowa kluczowe: trzustka, brodawkowato-torbielowaty nowotwór

Introduction

Papillary-cystic neoplasm of the pancreas (PCNP) is a rare tumor with a favorable prognosis. It mainly affects young women. It was first described by Frantz in 1959 [1]. Approximately 340 cases were reported in the world literature up to this day. It's pathogenesis remains unclear.

Case report

A 20-year old woman was admitted with 5-months history of abdominal pain with occasional vomiting and nausea.

She was in good general state. A tender mass was felt during deep palpation of her upper abdomen slightly above navel. The abdominal ultrasound showed solid, well-circumscribed 5 cm – diameter tumor of the head of pancreas (Fig. 1). The computerized tomography revealed a pathological solid structure, size 5x4x4 cm, (density 40jH) intensifying after contrast injection, projecting on the pancreatic head (Fig. 2). There was no evidence of extrapancreatic spread of the disease.

Hematological and biochemical tests were normal. Patient was qualified for surgery. A round tumor was discovered in the head of the pancreas during laparotomy. There was no evidence of metastatic disease in the abdomen. Tumor was removed completely together with it's surrounding capsule and with clear margins of normal tissue. The post-operative course was uncomplicated and the patient was discharged on the 8-th day after surgery. Pathology report revealed encapsulated papillary cystic neoplasm, 4x4x2.5 cm. The patient has no evidence of the disease 22 months after surgery and is still under regular follow-up.

Papillary-cystic neoplasm of the pancreas was first described in 1959 [1]. The nomenclature is confusing and

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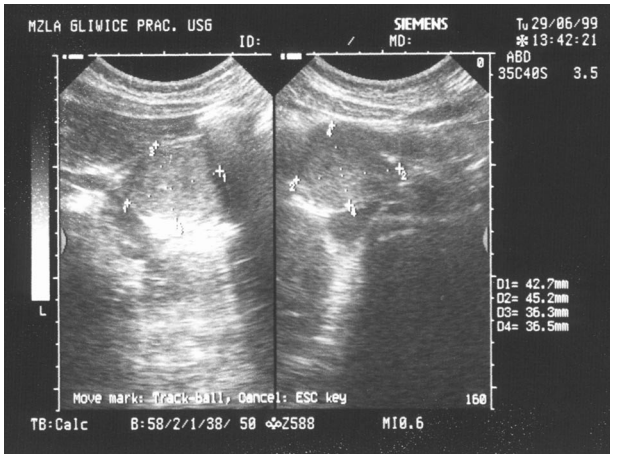

Fig. 1. Preoperative ultrasound image

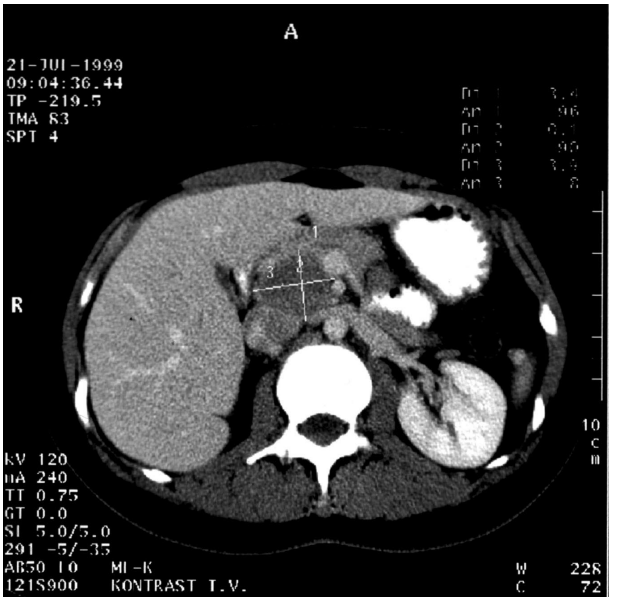

Fig. 2. Preoperative image by computerized tomography

different synonyms have been used to describe and name this lesion like: papillary cystic neoplasm/tumor of the pancreas, solid and papillary neoplasm, solid papillary epithelial neoplasm, solid-pseudopapillary tumor [2, 3]. Over 300 cases have been reported in the literature so far [4]. PCNP occurs predominantly in young women. Mean age is 25 years [4]. Men to female ratio is 1:11. At the time of diagnosis tumors are usually large, with mean diameter more than 10 cm [5]. In younger patients, under 20 years, tumors are slightly smaller. Mostly they are located in the tail of pancreas but in 1/3 of cases occur in the head of organ [4-6]. There was also report of PCNP developing outside the pancreas, the tumor was located retroperitoneally beneath tranverse mesocolon [7]. The symptoms are non specific. Mainly it is a palpable painless abdominal mass, sometimes associated with nausea and vomiting. Abdominal pain can also occurs. Tumor is often detected accidentally during routine physical examination or evaluation after an injury or other purposes. There were also reports of rapid symptoms caused by rupture of the tumor [2, 5, 8]. Pre-operative diagnosis and differentiation between other pancreatic tumor is difficult. Radiologically, a lesion can be solid, solid-cystic or cystic [2], and usually well circumscribed [9]. Presence of the sharp capsule is similar to the image of pancreatic pseudocysts [10]. Smaller tumors are generally solid and look like endocrine neoplasm. Magnetic resonance images may show areas of hemorrhage inside the tumor [11]. Tumoral calcification may also be recognized [3, 12]. There are no tumor markers present in the serum, and no report of endocrine or exocrine dysfunction [4,12]. Cytological diagnosis by fine needle aspiration is possible and smear is characteristic, but final pathologic diagnosis is usually made after examining the entire resected lesion [12, 13]. The majority of tumors have well-defined fibrous capsule. The tumor mass shows necrosis and hemorrhage. Cystic character observed in larger lesions is caused by degenerative changes in fundamentally solid tumors [12]. Cell lineage from which PCNP may origin is still unknown. Immunohistochemical and ultrastructural findings lead to divergent results. Some theories consider its histogenesis from ductal cells, others suggest acinar or endocrine origin. According to some authors these neoplasm arise from totipotential primitive stem cells of the pancreas [3, 5, 8, 12]. Despite of the confusing and mysterious origin of PNCP, all literature reports show its indolent character. Among 338 reported patients only 40 had invasive (infiltrating adjacent organs or important anatomical structures) or metastatic disease [4]. Only 3 patients died of this disease [4]. They all had hepatic metastases and died 131, 467, 72 month after first presentation. These patients were older then the mean age in the entire group: 36, 47 and 60 years. Metastases are usually presented at the time of diagnosis, rarely after first surgery [12]. Low grade or even only potential malignancy suggest the possibility of local excision of the tumor, although it leads to relatively high – 15% – risk of local recurrence [4]. Most of authors suggest radical resectional surgery [2, 4, 5, 13]. When the tumor is located in the tail or body of the organ (most cases) distal pancreatectomy is a safe procedure. Pancreatoduodenectomy for a potentially benign process in young patients when the head of pancreas is the site of tumor remains controversial. In the presented case we decided to perform local excision of the tumor from pancreatic perenchyma because of the lack of strict preoperative diagnosis and small magnitude of the lesion. Considering slow dynamic of primary tumors and small or even potential risk of distant metastases, the therapeutic decisions should depend on the site and diameter of the tumor as shown by radiology and cytology. Multiple unresectable lesions can be treated by chemo- or radiotherapy [5]. In spite of a few promising information a role of this kind of treatment has not been established, mainly because of a limited number of cases.

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