Turk J Gastroenterol 2006; 17 (4): 316-319

Solid pseudopapillary tumor of the pancreas: A case report

Solid psödopapiller pankreas tümörü: Vaka sunumu

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A 29-year-old woman presented with an abdominal mass existing for 10 years and abdominal pain for one year. Physical examination revealed an abdominal mass about 10 cm in diameter between the epigastrium and right upper quadrant. Abdominal ultrasonography and multi-slice computerized tomography showed a well-demarcated solid mass containing cystic and calcified areas (97-80 mm in diameter) located on the head and uncinate process of the pancreas. Percutaneous ultrasonography-guided tru-cut biopsy was performed and the pathologic diagnosis of biopsy material was solid pseudopapillary tumor of the pancreas. The patient then underwent surgery and exploration revealed an encapsulated mass of 10 cm in diameter that was retracting the portal vein and was adherent to mesentery of the transverse colon. Pancreaticoduodenectomy was performed preserving the pylorus. Histopathologic diagnosis of the mass supported the tru-cut biopsy findings. At 12th months of follow-up, physical, laboratory and radiological examinations were all normal. Although solid pseudopapillary tumor is a rarely seen low-grade malignant tumor, it is important to differentiate it from other pancreatic tumors because of its benign course.

Key words: Solid pseudopapillary tumor, pancreas, tru-cut biopsy, pylorus-preserving pancreaticoduodenectomy

29 yaşındaki kadın hastanın 10 yıldır karında gittikçe büyüyen kitle ve son bir yıldır karın ağrısı şikayeti vardı. Muayenede epigastriumdan karın sağ üst kadrana uzanan yaklaşık 10 cm büyüklüğünde kitle mevcuttu. Abdominal ultrasonografi ve bilgisayarlı tomografide pankreas baş-uncinate proses kısmında 97x80 mm boyutlarında, içerisinde kistik ve kalsifiye alanlar bulunan iyi sınırlanmış solid kitle saptandı. Ultrasonografi eşliğinde yapılan perkütan tru-cut biyopsinin histopatolojik incelemesinde pankreasın solid psödopapiller tümörü saptandı. Ameliyatta pankreas baş-uncinate proses kısmında yaklaşık 10 cm çapında, kapsüllü, portal veni kendisine çekmiş, transvers kolon mezosuna yapışık kitle görüldü. Pilor koruyucu pankreatikoduodonektomi yapıldı. Kitlenin histopatolojik incelemesi tru-cut biyopsinin histopatolojik inceleme bulgularını doğruladı. 12. ayda yapılan kontrolde fizik muayene, laboratuar ve radyolojik tetkikler normaldi. Pankreasın solid psödopapiller tümörü nadir görülen, düşük dereceli malign bir tümördür. Ancak bu tümör genellikle benign klinik seyir gösterdiği için diğer pankreas tümörleri ile ayırıcı tanısının yapılması önemlidir.

Anahtar kelimeler: Solid psödopapiller tümör, pankreas, tru-cut biyopsi, pilor koruyucu pankreatikoduodonektomi

INTRODUCTION

Solid-pseudopapillary tumor (SPPT) is a very rare primary neoplasm of the pancreas (2-3% of primary pancreatic tumors occurring at all ages [1]). Franz first described it in 1959. It is usually seen in young females (2). In spite of possible histological findings of malignancy, SPPT typically shows a benign clinical course and a low malignant potential (3). The pathogenesis of these tumors is still controversial. It has been suggested that it might originate from ductal and acinar pancreatic cells, endocrine cells or pluripotential stem cells (4-7).

CASE REPORT

A 29-year-old woman presented with abdominal mass existing for 10 years and abdominal pain for one year. The patient had never been examined for these complaints before. Physical examination revealed an abdominal mass about 10 cm in diameter between the epigastrium and right upper quadrant. The mass was smooth-surfaced and partly mobile. Laboratory findings revealed increased lactate dehydrogenase (LDH) level [302 U/I (normal range: 100-210 U/I)] and erythrocyte sedimentation rate [38 mm/h (normal range: 0-20)] and mild anemia (hemoglobin: 11.3 g/dl). Tumor mar-

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Manuscript received: 19.04.2006 **Accepted:** 27.09.2006

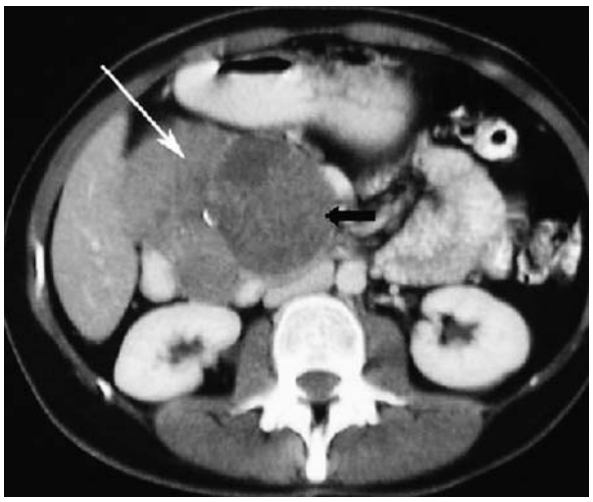

Figure 1. Abdominal computed tomography showed an enhancing, well-demarcated mass on pancreatic head and uncinate process, with solid (white arrow) and cystic (black arrow) components

kers (alpha-fetoprotein, carcinoembryonic antigen, CA-125, CA15-3, CA19-9, CA72-4) of serum and other biochemical analyses were within normal limits. Abdominal ultrasonography (USG) and multi-slice computed tomography (MSCT) showed a solid mass containing cystic and calcified areas (97-80 mm in diameter) located on the head and uncinate process of the pancreas (Figure 1). Celiac and superior mesenteric angiography was performed to exclude vascular invasion. Superior mesenteric angiography and arterial portography revealed some lateral displacement of the superior mesenteric artery and tumoral enhancement on arterial and late arterial phase; the portal vein was displaced superiorly. There was no vascular invasion. After the angiographic evaluation, percutaneous USG-guided tru-cut biopsy was performed. Pathologic examination of biopsy material revealed solid islets of small cells with uniform spherical nuclei and tumoral cells forming papillary structures. Neuron specific enolase (NSE), chromogranin, CD10 and synaptophysin antibody stainings were negative; however, the tumoral cells were stained remarkably with progesterone receptor antibody. In light of these findings, primary pathologic diagnosis was SPPT. The patient then underwent surgery and exploration revealed an encapsulated mass of 10 cm in diameter that was retracting the portal vein and adherent to the mesentery of the transverse colon. The mass was dissected from the mesentery and

Figure 2. Resection material

portal vein, and pancreaticoduodenectomy was performed preserving the pylorus of the stomach (Figure 2). Totally 21 lymph nodes (13 peripancreatic and 8 periduodenal) were resected.

Pathologic examination supported the primary diagnosis. The tumor was largely infiltrating pancreatic tissue, and invading the pancreatic parenchyma (Figure 3). No vascular invasion or lymph node metastasis was detected. Postoperative period was complicated with pneumonia and the patient was discharged with complete recovery on the 11th day postoperatively. In the 12th month after the operation, physical examination was normal and the patient had no symptoms. MSCT of thorax and abdomen revealed that there was no tumoral recurrence or metastasis.

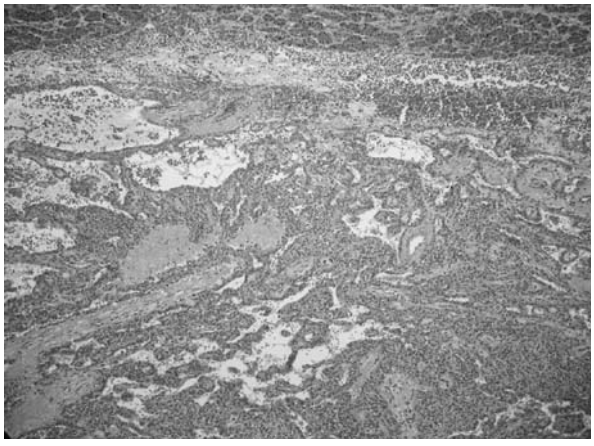

Figure 3. Pathologic specimen of mass; tumoral tissue near the normal pancreatic tissue (H&E, x40)

DISCUSSION

Solid-pseudopapillary tumor is almost exclusively (90%) encountered in young females between the 2nd and 4th decades (8, 9). The body and tail of the pancreas are more frequently affected (64%) than the head (3). These tumors are detected incidentally in most cases; however, abdominal pain may be the sole and important sign of the tumor. Additionally, abdominal mass, anorexia, weight loss, and symptoms due to the compression of adjacent organs can be present (3, 10, 11). Rarely, acute abdomen and/or hypovolemic shock may develop due to intra-tumoral or intra-abdominal hemorrhage (10). Our patient had abdominal mass and pain. Biochemical analyses usually do not help in diagnosis (11). Although the tumor markers (CA19-9, CA242, CA-50, CA72-4) have diagnostic value in malignant pancreatic cancers, serum levels of these markers are all usually normal in SPPT (11, 12). In our patient, erythrocyte sedimentation rate and LDH were slightly increased and there was mild anemia.

Cysts of different origin (dysontogenetic, post-traumatic, inflammatory or hydatid cysts, and retention cysts) and exocrine (pancreatoblastoma, ductal cell or nonductal acinar cell carcinoma), endocrine (islet cell hyperplasia, insulinoma, gastrinoma and VIPoma) or connective tissue tumors (e.g., sarcoma, lymphoma, teratoma, hemangioendothelioma) are included in the differential diagnosis of a mass located in the region of the lesser sac (3, 11, 13).

Solid-pseudopapillary tumor is rarely malignant, and it usually has a benign clinical course (95%) (6). Common sites of metastasis include liver (42%), peritoneum (42%) and lymph nodes (25%) (14). In 16% of cases, it invades adjacent tissues and vascular structures (3). There was no metastasis in our case.

Surgery is the mainstay of treatment. Complete resection of the tumor with distal pancreatectomy or pancreaticoduodenectomy is usually achieved. Tissue-sparing surgery is possible and justified owing to the favorable biological characteristics of the tumor and since the mass is usually surrounded by a dense fibrous capsule. In most studies, extensive lymphatic dissection or more radical local approaches are not indicated (3, 15). In our case, we performed pylorus-preserving pancreaticoduodenectomy operation because of the location of the tumor. Zinner et al. (16) reported that 10-year survival was 100% in a case series.

The lack of an exact preoperative diagnosis poses the greatest problem for therapeutic decision making in these cases (9). In diagnostic work-up, abdominal and/or endoscopic USG, CT, magnetic resonance imaging (MRI), fine needle biopsy, pancreatic ductal brush cytology, and intraoperative frozen section pathologic examination are all valuable (3, 9, 10). A CT scan of the abdomen usually shows a well-encapsulated mass with both solid and cystic components. There may be calcification and also contrast enhancement of solid components (17). However, none of these radiological features are diagnostic (18). MRI could demonstrate the solid papillary and cystic components of tumor better than CT (15). In the setting of typical clinical and imaging findings, an accurate preoperative diagnosis of pancreatic SPPT can be established by aspiration cytology and immunochemistry with or without concomitant tru-cut biopsy, on the basis of which clinicians decide treatment (19).

Pathologic findings of SPPT are characteristically cystic areas and pseudopapillary solid cellular clusters (9). It was reported that immunohistochemical examination is usually positive for vimentin, keratin, alpha-1-antitrypsin, alpha-1-antichymotrypsin, NSE, and sometimes reactive for S-100 (3, 10). This tumor sometimes has estrogen or progesterone receptors (3, 20).

In conclusion, SPPT of the pancreas, which has a benign clinical course, is rarely observed as a low-grade malignant tumor. Because of benign characteristics of the tumor, preoperative diagnosis is very important for the surgical strategy. Because the radiologic findings are not specific, radiologists and surgeons should take this tumor into consideration in the differential diagnosis of pancreatic malignancies, especially in young female patients; preoperative USG or CT guided tru-cut biopsy could provide the exact diagnosis.

REFERENCES

1. Morohoshi T, Held G, Kloppel G. Exocrine pancreatic tumors and their histological classification: a study based on 167 autopsy and 97 surgical cases. Histopathology 1983; 7: 645-61.

2. Frantz VK. "Tumors of the pancreas" Atlas of Tumor Pathology, section VII, fascicles 27 and 28. Washington, DC, US Armed Forces Institute of Pathology, 1959.

3. Rebhandl W, Felberbauer FX, Puig S, et al. Solid-pseudopapillary tumor of the pancreas (Frantz tumor) in children: report of four cases and review of the literature. J Surg Oncol. 2001; 76: 289-96.
4. Boor PJ, Swanson MR. Papillary-cystic neoplasm of the pancreas. Am J Surg Pathol. 1979; 3: 69-75.
5. Benjamin E, Wright DH. Adenocarcinoma of the pancreas of childhood: a report of two cases. Histopathology. 1980; 4: 87-104.
6. Von Herbay A, Sieg B, Otto HF. Solid-cystic tumour of the pancreas. An endocrine neoplasm? Virchows Arch A Pathol Anat Histopathol. 1990; 416: 535-8.
7. Pezzi CM, Schuerch C, Erlandson RA, et al. Papillary-cystic neoplasm of the pancreas. J Surg Oncol. 1988; 37: 278-85.
8. Ferlan-Marolt V, Pleskovic L, Pegan V. Solid papillary-cystic tumor of the pancreas. Hepatogastroenterology. 1999; 46: 2978-82.
9. Bektas H, Werner U, Kaaden S, et al. Solid-pseudopapillary tumor of the pancreas-a rare and frequently misdiagnosed neoplasm. Langenbecks Arch Surg. 1999; 384: 39-43.
10. Meshikhes AW, Atassi R. Pancreatic pseudopapillary tumor in a male child. JOP. 2004; 5: 505-11.
11. Salvia R, Festa L, Butturini G, et al. Pancreatic cystic tumors. Minerva Chir. 2004; 59: 185-207.
12. Jiang XT, Tao HQ, Zou SC. Detection of serum tumor markers in the diagnosis and treatment of patients with pancreatic cancer. Hepatobiliary Pancreat Dis Int. 2004; 3: 464-8.
13. Kosmahl M, Pauser U, Peters K, et al. Cystic neoplasms of the pancreas and tumor-like lesions with cystic features: a review of 418 cases and a classification proposal. Virchows Arch. 2004; 445: 168-78.
14. Mao C, Guvendi M, Donenico 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; 188: 821-8.
15. Kheng Hong NG, Puay Hoon T, Choon Hua T, et al. Solid pseudopapillary tumor of the pancreas. ANZ J Surg. 2003; 73: 410-5.
16. Zinner MJ, Shurbaji MS, Cameron JL. Solid and papillary epithelial neoplasms of the pancreas. Surgery. 1990; 108: 475-80.
17. Dong PR, Lu DS, Degregario F, et al. Solid and papillary neoplasm of the pancreas: radiological-pathological study of five cases and review of the literature. Clin Radiol. 1996; 51: 702-5.
18. Procacci C, Graziani R, Bicego E, et al. Papillary cystic neoplasm of the pancreas: radiological findings. Abdom Imaging. 1996; 20: 554-8.
19. Bhanot P, Nealson WH, Walser EM, et al. Clinical, imaging, and cytopathological features of solid pseudopapillary tumor of the pancreas: a clinicopathologic study of three cases and review of the literature. Diagn Cytopathol. 2005; 33: 421-8.
20. Kloppel G, Maurer R, Hofmann E, et al. Solid-cystic (papillary-cystic) tumours within and outside the pancreas in men: report of two patients. Virchows Arch A Pathol Anat Histopathol. 1991; 418: 179-83.