

Vaka Takdimleri

Papillary Cystic Tumor of The Pancreas: A Case Report

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Papillary cystic tumor of the pancreas (PCTP) is a distinct clinicopathologic disease (8). PCTP is a rare, low-grade malignancy occurring primarily in a young woman. Its histogenesis is still controversial (2,6,18,31,35). Because of its gross and histologic features, PCTP has been called solid and papillary epithelial neoplasm (6), solid and cystic tumor (5,20), papillary epithelial neoplasm (2), and papillary epithelial tumor (8,21). Although the biologic characteristics of the tumor generally indicate a good prognosis, recently a malignant form has been reported (3,6,7,11,14,18,25,27,30,32,33,35,36). The tumor has favorable prognosis after surgical resection (6,14,26). Since Frantz described the first case in 1959, approximately 130 cases have been reported in the literature. We describe a case in which PCTP was found on the pancreatic tail of a 36 year old woman.

CASE REPORT

A 36 year old woman was admitted to the hospital with complaint of left quadrant abdominal mass and pain. This was associated with nausea and sometimes vomiting. Her medical history was essentially unremarkable. She didn't smoke and not used alcohol. Physical examination; Auscultatory findings of the heart and lungs were unremarkable. A large mass was palpated in the left upper quadrant to midepigastic region of the abdomen. The abdomen was soft, nontender and displayed normoactive bowel sound.

At the time of admission, routine serum laboratory results were within normal limits for example SGOT, SGPT, alkaline phosphatase, prothrombine time, blood glucose level, amylase, lipase, total bilirubine, HCG. The erythrocyte sedimentation rate was elevated up to 60 mm/h.

Abdominal computed tomography scan and ultrasonography revealed a 9x9x5 cm heterogen cystic mass on the tail of the pancreas. The liver was unre-

markable. No kidney abnormality was noted and no retroperitoneal adenopathy was seen. A chest roentgenogram and endoscopic retrograde cholangiopancreatography (ERCP) were unremarkable. Total excision of the mass and distal pancreatectomy was performed.

Gross inspections of the specimen demonstrated a mass of 9x9x5 cm in diameter. On gross examination, PCTP occurred as a well-circumscribed mass generally demarcated from the pancreas by a peripheral pseudocapsule. On cross section, a variegated appearance was evident with variable combinations of solid haemorrhagic and cystic necrotic areas. Solid viable portions were more frequently grayish brown, whereas cystic-necrotic areas were red-yellow. The capsule of each lesion was intact. The intervening pancreatic tissue was grossly unremarkable.

Microscopy: The tumor grossly appeared to have large collagenous capsule and large haemorrhagic necrotic areas (Fig 1). There were sheets of relatively uniform small cells with dark round nuclei and clear cytoplasm in the solid areas (Fig 2). In the papillary areas, there were papillae with a central fibrovascular core (Fig 3). There was a slight neutrophilic and monocytic inflammatory infiltrate in the tumor. In the papillary areas and microcystic areas, there were empty spaces.

DISCUSSION

PCTP is a rare tumor of low grade malignant potential occurring predominantly in young woman (8,9). Because of PCTP's striking female predominance, some authors have suggested hormonal participation in its pathogenesis (7). Although some studies have found evidence of estrogen and progesterone receptor positivity, other studies have failed to confirm any hormone-receptor positivity (19). There is evidence that tumors occurring in older person are more likely to be-

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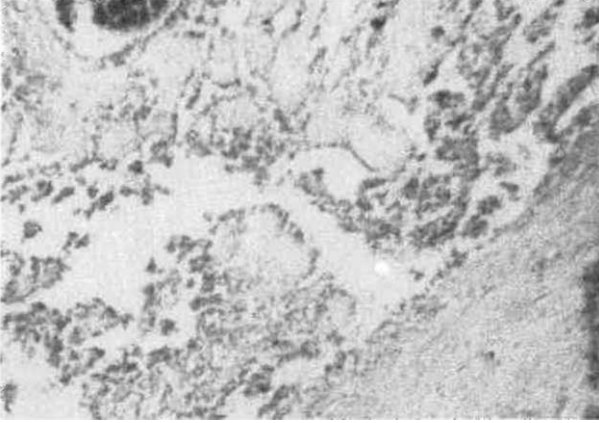


Figure 1. Papillary, haemorrhagic and microcystic areas (HEEx200)

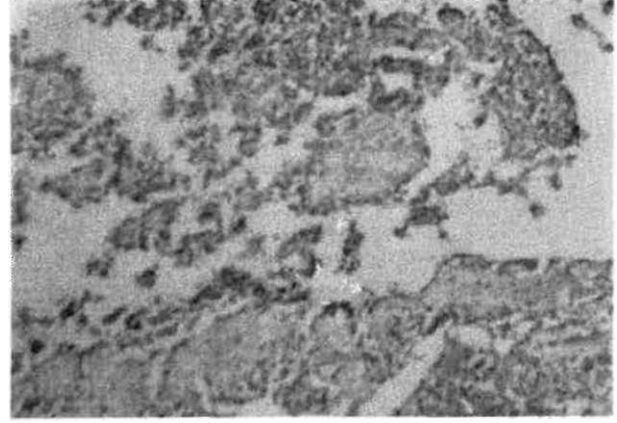


Figure 2. Microcystic and solid areas (HEEx200)

have aggressively (19). Only eight cases of PCTP in male patients have been reported (4,9,10,12,13,27,34). Two thirds of the tumors occur as a slowly enlarging abdominal mass, whereas one third is accompanied by abdominal pain or discomfort (2) like our case. The diagnosis of PCTP is usually not suspected until laparotomy because of the lack of specificity of laboratory tests, ultrasound and radiologic appearances. A preoperative fine needle aspiration (FNA) cytologic diagnosis under ultrasound guidance in the appropriate clinical setting may be rendered with confidence so that the physician may know in advanced the nature of the abdominal mass and plan adequate treatment. Bonderson and Colleagues (1) first described the preoperative cytological diagnosis under ultrasound guidance, emphasizing the conclusion of branching papillary structures with delicate fibrovascular stalks lined by one or more layers of monotonous, bland appearing cells (2). We don't have any experience in this issue. Preoperative ERCP would be beneficial in some instance where available, intraoperative ultrasound would be most useful in detecting an occult second pancreatic mass (5). This tumor may occur anywhere in the pancreas (9,22) and most PCTPs occur within the tail of the pancreas. The mass of our case was measured 9x9x5 cm in diameter on the tail of the pancreas. PCTPs were describe; the mean diameters of these tumors were usually 8 to 10 cm. The neoplasm displaces surrounding structures instead of invading them and complete removal of the tumor is usually possible in every case example our case. Most tumors behave as a benign or very low-grade malignant fashion and the prognosis after surgical excision is excellent (24). Aggressive behavior with extension of the tumor into surrounding organs or vessels, local recurrence or distant metastases, have been documented in 21 patients (3,8,23, 26,30,32,33). Oertel and associates reported that in

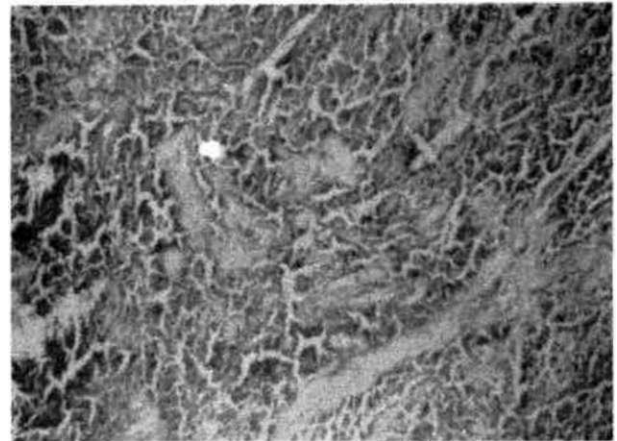


Figure 3. Papillary areas with a central fibrovascular core (HEX100)

their study "several patients over 20 years of age with neoplasm have developed peritoneal and hepatic metastases and at least one died" (22). The presence of unrecognized lesions within the residual pancreas may explain the recurrences that were seen in partial pancreatic resections (8,15,23); however there is evidence that local recurrence and even distant metastases of PCTP may be excised successfully with subsequent long patient survival times (18,32). Another metastatic PCTP showed an increase in nuclear pleomorphism, hyperchromasia, mitotic rate and the number of bizarre tumor giant cells. We did not observe these in our case.

The histogenesis of PCTP still remains unsettled because of discrepancies in the immunohistochemical and electron-microscopic findings.

More specific neuroendocrine markers, such as synaptophysin and chromogranin, have been consistently negative in PCTP (5). Acinar cell differentiation has been suggested by the reactivity of all PCTPs

examined for alpha-1-antitrypsin (AAT) (12,18,20) and the presence of intracoplasmic electron-dense granules resembling zymogen granules (5,18). However, immunoreactivity for AAT is not restricted to exocrine pancreatic cells and should not be considered evidence of acinar cell differentiation. Morohoshi and associates reported (20) that AAT represents a characteristic, but not specific, immunocytochemical marker for PCTP; furthermore, it may represent AAT of plasma origin deposited in areas of tumor degeneration.

A recent report about a papillary cystic tumor of the liver that was strikingly similar to PCTP in clinical,

gross, histologic, and ultrastructural features suggest that it may have originated from metaplastic pancreatic tissue. This, in turn, reflects a common developmental origin of biliary and pancreatic epithelium from the caudal foregut.

We conclude that PCTP is a distinctive clinicopathologic entity with a benign clinical course or a very low malignant potential and a high cure rate by surgical resection alone. So that we performed total resection of the mass with the tail of the pancreas. In our case No recurrence or metastases were found in the follow-up period of one year.

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