Papillary-cystic neoplasm of the pancreas

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Abstract. Papillary-cystic neoplasm of the pancreas is a rare, nonfunctioning low-grade malignant tumor seen in young patients, most often female. Ultrasound and CT show a circumscribed, solid nonhomogeneous mass with cystic areas, with peripheral but not central enhancement and occasional calcification. Prognosis after excision is usually excellent. We describe a case of the papillary-cystic neoplasm of the pancreas in a 13-year-old girl to illustrate the radiological findings.

Cancer of the pancreas is very rare in the pediatric age group. Most of the pancreatic neoplasms among children are endocrine (beta-cell tumors) [1]. At least 10 types of primary nonendocrine neoplasms have also been described [2], among which papillary-cystic neoplasm is notable because of its rarity and low-grade malignant potential. Its generally good prognosis, if completely resected, makes knowledge of this rare entity very important [3].

Case report

A 13-year-old white girl was admitted with a 5-day history of vomiting and a 2-day history of abdominal pain. She was afebrile. Physical examination was unremarkable except for tenderness on palpation in the epigastric region. There were no masses or hepatosplenomegaly. Laboratory data were normal with the exception of LDH of 223 U/l (normal range 94-172 U/l) and a slightly elevated ESR of 49 mm/h (normal range 0-20 mm/h). No leukocytosis was noted. Serum amylase was normal. The following tumor markers were measured: serum level of cancer antigen 125 was 46 U/ml (normal range 0-35 U/ml), serum level of carbohydrate antigen 15-3 was 23 U/ml (normal range 0-31 U/ml), and plasma level of carcinoembryonic antigen was 0.9 ng/ml (normal range less than 4.0 ng/ml).
Fig. 2a Non-enhanced CT scan shows a large mass difficult to separate from the tail of pancreas. The mass is well delineated, circumscribed and of homogeneously low density. Calcification was seen in other slices (not shown). b Enhanced CT scan shows that only the wall of the mass enhances.

On pathologic examination the mass was soft and cystic. The cut surface showed a 5-cm cyst filled with necrotic tissue (Fig. 3a). Microscopic examination showed that the mass had the distinctive features of papillary-cystic neoplasm of the pancreas. In some areas the tumor was composed of sheets of uniform polygonal cells (Fig. 3b), and extensive foci of necrosis were present. In other areas the tumor had a papillary configuration (Fig. 3c).

Discussion

Preoperative recognition of papillary-cystic neoplasm of the pancreas is important, since complete resection results in a cure in most cases [4]. It occurs mostly among female patients, especially of African descent [5]. Most patients are younger than 40 years of age and the mean age is around 20 years [6].

Papillary-cystic neoplasm of the pancreas is also known as papillary solid or papillary epithelial neoplasm, or papillary carcinoma of the pancreas [7]. The tumor demonstrates two cellular patterns. The first consists of sheets of cells with interspersed cystic spaces of varying size. The second consists of long thin papillary structures with a central fibrovascular core. These cells have a homogeneous appearance with regular, round, indented or folded nuclei of slightly variable size, minimal mitotic activity and glassy bright eosinophilic cytoplasm. The cytological features of the tumor suggest that this neoplasm originates from the epithelium of the small pancreatic ducts [8].

Ultrasonography demonstrates a solid mass, well-demarcated and containing hypoechoic cystic areas of variable number and size. On CT the mass is well delineated, nonhomogeneous, of uneven soft-tissue density with central necrosis [9]. There is no central enhancement after intravenous injection of contrast, but peripheral enhancement has been noted. Areas of attenuation higher than water in the cystic space are consistent with intratumoral hemorrhage and cystic degeneration. Calcification is rare. A few cases of hepatic metastases have been reported [10, 11]. On MRI the tumor is well demarcated with areas of high signal intensity on T1-weighted images corresponding to hemorrhagic necrosis or debris. A low-intensity rim is consistent with the fibrous capsule or compressed residual pancreatic tissue. On T2-weighted images signal intensity ranges from very low to high [10]. Sonographic and CT findings of our case were typical according to the few descriptions of this mass in the radiologic literature [9, 11, 12]. MRI was not performed in our case.

Differential diagnosis of papillary-cystic neoplasm of the pancreas includes: serous cystadenoma, mucinous cystic neoplasms, nonhyperfunctioning islet cell tumor, and pancreatoblastoma. Pancreatic duct adenocarcinoma is the most common pancreatic malignancy in adults, although this does not apply to children. In the literature there are descriptions of imaging features useful for differential diagnoses of the nonendocrine neoplasms of the pancreas [10, 12], but in the majority of patients it is not possible to differentiate these tumors by their radiologic appearance alone.

Treatment of papillary-cystic neoplasm of the pancreas consists of complete surgical excision. Unlike other malignant pancreatic tumors surgical treatment is usually associated with an excellent prognosis [12]. The purpose of this short report is to alert radiologists to the possibility of this form of neoplasm in a child presenting with a pancreatic mass fulfilling the imaging criteria described.