CASE REPORT

Pancreatic PEComa (Sugar Tumor): MDCT and EUS Features

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ABSTRACT

Context PEComas (tumors showing perivascular epithelioid cell differentiation) of the pancreas are exceedingly rare. **Case report** We herein report on a 60-year-old female who noticed a bulge in her right upper quadrant while exercising. Subsequent multidetector-row CT scan showed a 3.5 cm well-defined, encapsulated, hypovascular, solid tumor in the body of the pancreas. Endoscopic ultrasound demonstrated a mixed hypo- and hyper-echoic, well-defined, heterogeneous tumor. **Conclusions** Although three pancreatic PEComas (sugar tumors) have been described previously, to the best of our knowledge, this is the first report of a pancreatic PEComa with illustration of its multidetector-row CT and endoscopic ultrasound features in the radiological literature.

INTRODUCTION

PEComas (perivascular epithelioid cell tumors) encompass a group of rare mesenchymal neoplasms with shared cellular characteristics that have been found in numerous anatomic sites throughout the body [1]. They consist of distinctive perivascular epithelioid cells and also demonstrate characteristic immuno-phenotypic features [2, 3]. The development of PEComas has been genetically linked to mutations within the tuberous sclerosis complex [4]. Malignant variants of PEComas have previously been described demonstrating the importance of early diagnosis and management [5].

Pancreatic PEComas are exceedingly rare neoplasms. To the best of our knowledge, we report only the 4th case in the English language literature [6, 7, 8]. Two additional cases described primary pancreatic angiomyolipomas [9, 10]. Moreover, all prior reports have been published in the pathologic literature and have not illustrated the radiologic findings. We, herein, report on a case that demonstrates the multidetector-row computed tomography (MDCT) and endoscopic ultrasound (EUS) characteristics of a histo-pathologically proven case of pancreatic PEComa.

Received April 20th, 2009 - Accepted September 15th, 2009

Key words Endosonography; Pancreatic Neoplasms; Perivascular Epithelioid Cell Neoplasms; Tomography, X-Ray Computed

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Document URL http://www.joplink.net/prev/200911/11.html

CASE REPORT

A 60-year-old woman was referred to our institution for the evaluation of a bulge in her right upper quadrant that she first noticed while exercising. Biochemical evaluation revealed no significant abnormalities. The CA 19-9 level was 20.9 U/mL (reference range: 0-35 U/mL). A multiphasic contrast-enhanced MDCT scan with multiplanar reconstruction was performed before and after intravenous administration of 100 mL of Ultravist® 300 (Bayer Healthcare, Seattle, WA, USA) at 3 mL/sec with 40 and 70 second delays. The images revealed the presence of a 3.5 cm, round, solid, well-defined, homogeneously enhancing mass in the body of the pancreas. The lesion appeared hypovascular to the surrounding pancreatic parenchyma with the exception of a hypervascular capsule (Figure 1). Because of a high suspicion for a pancreatic neoplasm, the patient was referred for endoscopic ultrasound-guided fine needle aspiration biopsy of the lesion. EUS confirmed the presence of a round mass in the pancreatic body. The mass was mixed hypo- and hyper-echoic, heterogeneous, and solid. It measured 3.3 cm by 2.6 cm on maximal cross-sectional diameter (Figure 2). The endosonographic borders were well-defined and the lesion appeared to be partially encapsulated. The pancreatic tail showed mild dilation of the pancreatic duct and hyperechoic ductal walls. There was neither evidence for vascular invasion nor presence of

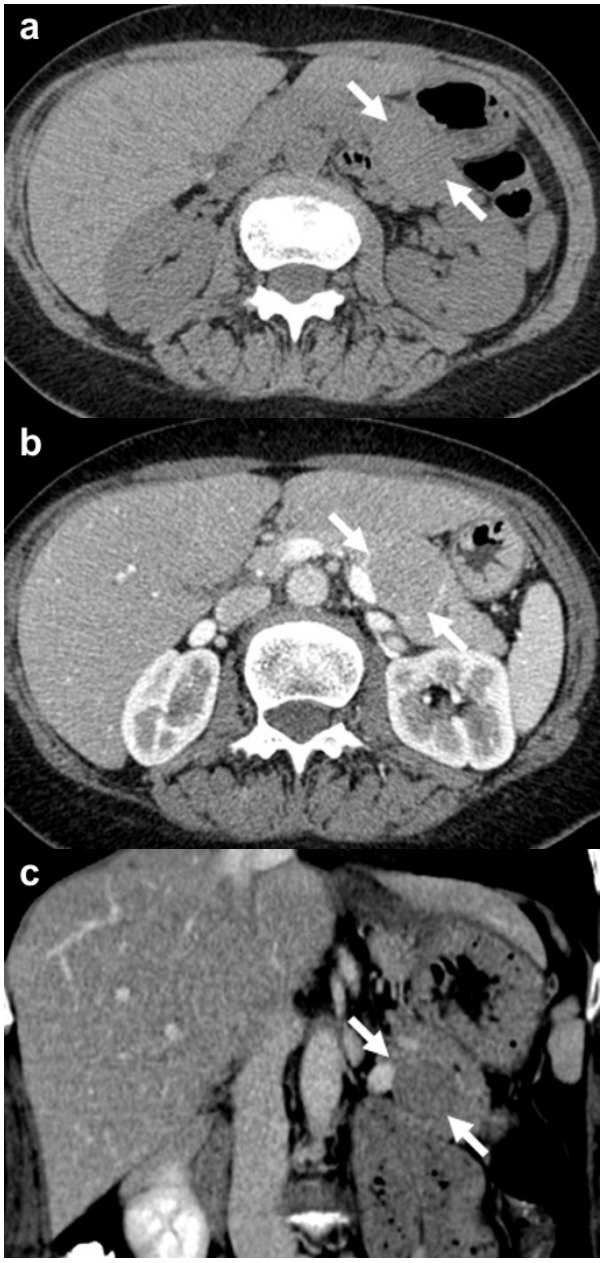

Figure 1. a. Non-contrast enhanced axial CT image of the abdomen reveals a homogeneous pancreatic mass (arrows) in the body isodense to the surrounding parenchyma. **b.** Arterial phase contrast enhanced axial abdominal CT image demonstrates a round, well defined, predominantly hypovascular mass with a hypervascular capsule (arrows). **c.** Contrast-enhanced, coronally reconstructed image of a portal venous phase contrast enhanced abdominal CT demonstrates a well defined, partially encapsulated hypovascular mass in the body of the pancreas (arrows).

lymphadenopathy. Color Doppler imaging was utilized prior to needle puncture to confirm a lack of significant vascular structures within the needle path. Two passes were made with a 22-gauge needle using a transgastric approach. On cytology, malignant cells consistent with poorly differentiated carcinoma were identified. There was insufficient material present in the cellblock preparation to allow for further characterization of the lesional cells.

Because of the presumed resectability of the pancreatic mass on imaging, the patient underwent an elective distal pancreatectomy and splenectomy. Histo-pathological analysis of the resection specimen revealed a 3.2 cm solid, encapsulated tumor located in the body of the pancreas. No necrosis was identified. There was no evidence of malignancy. Pancreatic parenchymal and posterior pancreatic surface resection margins were negative for tumor. Microscopically, the tumor was composed of sheets of epithelioid to spindled cells with abundant granular eosinophilic to clear cytoplasm and mild to moderate nuclear pleomorphism. Rare mitotic figures were seen. Immunophenotypically, the tumor cells were positive for both melanocytic markers (HMB-45, melan-A, and MiTF) and smooth muscle markers (SMA and desmin), and negative for all cytokeratins (Figure 3). These distinctive features were characteristic for a PEComa [7]. The patient underwent surgical resection of the PEComa without adjuvant therapy and had an uneventful post-operative course. An abdominal CT scan 3 weeks later showed no evidence of tumor. She has remained asymptomatic and shows no signs of recurrence in the subsequent 7 months of follow-up. To the best of our knowledge, the patient does not carry any mutations within the tuberous sclerosis gene complex predisposing her to additional tumors.

DISCUSSION

PEComa is a term that describes “a mesenchymal tumor composed of histologically and immunohisto-chemically distinctive perivascular epithelioid cells” [11]. The term has been used to describe a group of related pathologic processes including epithelioid renal angiomyolipoma, lymphangiomeyomatosis, clear cell sugar tumor of the lung, clear-cell myomelanocytic tumor of the falciform ligament/ ligamentum teres, as well as clear cell tumors in other anatomic locations [5]. PEComas have been described in the bladder, prostate, uterus, ovary, vulva, vagina, liver, bone, orbit, retroperitoneum, soft tissue, nasal cavity, pelvis, GI

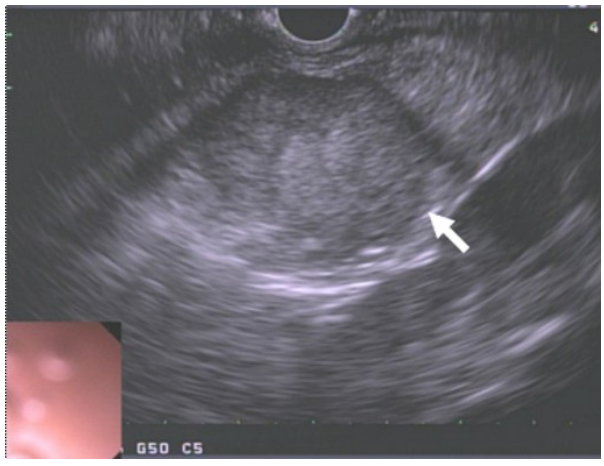

Figure 2. Endoscopic ultrasound image demonstrates a well defined, heterogeneous mass in the pancreas.

tract, and pancreas [5]. The epidemiology of these neoplasms can prove helpful in diagnosis as PEComas have a strong female gender predilection [1]. Histopathological examination demonstrates nests and sheets of cells with clear to granular eosinophilic cytoplasm often found in close proximity to blood vessel walls [1]. The tumors are well-circumscribed and demarcated from surrounding parenchymal tissue by a thin capsule [1, 5, 7, 8]. PEComas are often referred to as "sugar" tumors because the cytoplasm of the perivascular epithelioid cells is glycogen rich [12]. Immunophenotypically, the tumors demonstrate reactivity against melanocytic (HMB-45 and melan-A) and smooth muscle (actin and desmin) markers [1].

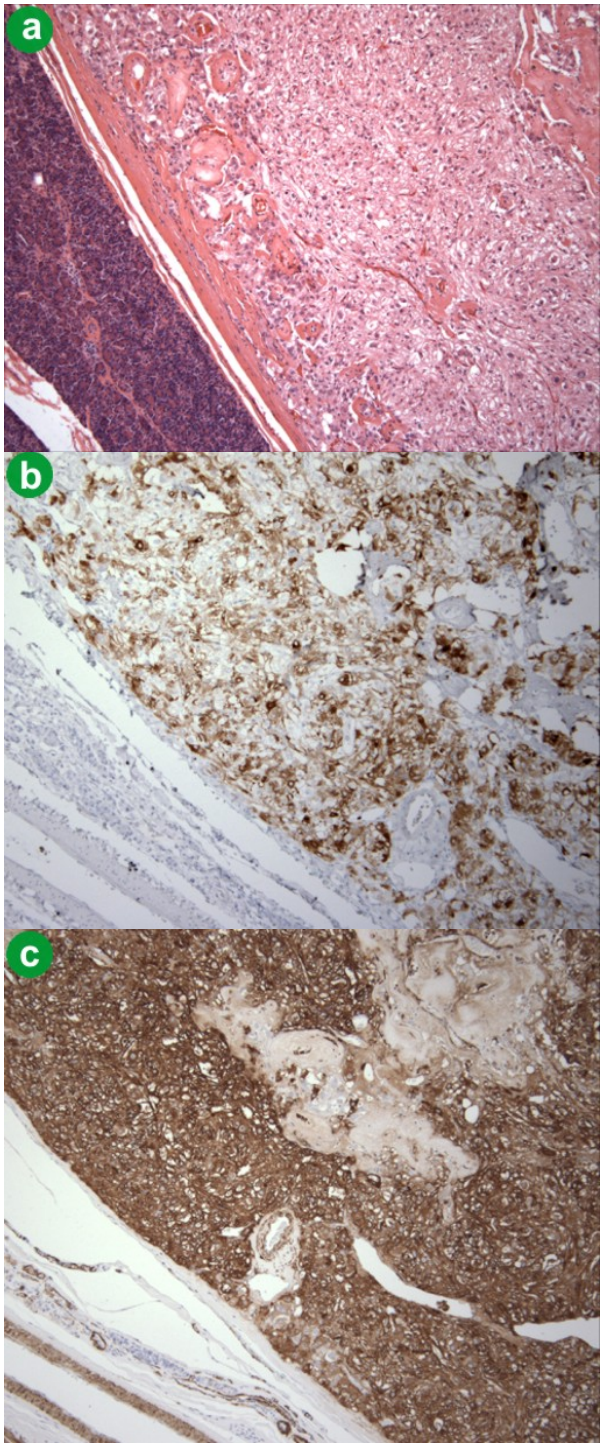

Figure 3 a. Histological sections of pancreas PEComa. H&E stained section showing a well-demarcated tumor adjacent to normal pancreatic parenchyma (left-lower corner). The tumor is composed of sheets of epithelioid to spindled cells with abundant clear to granular cytoplasm, and mild to moderate nuclear pleomorphism. **b.** The tumor cells are immunophenotypically positive for the melanocytic marker Melan-A. **c.** The tumor cells are immunophenotypically positive for the smooth muscle marker SMA.

The pathologic recognition of a PEComa, as illustrated in our case, is difficult and requires cytological and immunohistochemical analysis. PEComas exhibit a wide spectrum of imaging findings, although shared characteristics should alert the radiologist to this possible diagnosis. Non-pancreatic gastrointestinal PEComas have been described as large heterogeneous or homogeneous masses that are usually well demarcated [13, 14]. Pancreatic PEComas are exceedingly rare tumors with, to the best of our knowledge, only 3 previously reported cases in the literature [6, 7, 8]. A single pancreatic PEComa has been described as a well-circumscribed, homogenous soft tissue mass on CT in a previous report [7].

Given the rarity of PEComas, one must first rule out common pancreatic masses. Primary pancreatic tumors either arise from epithelial or mesenchymal tissue. Those of epithelial origin are further subcategorized as ductal, acinar, or endocrine neoplasms [15]. The most common, pancreatic ductal adenocarcinomas, account for 85% of all pancreatic neoplasms, appear as poorly defined hypodense masses on CT that are often metastatic at the time of discovery, and are associated with a poor prognosis [15, 16]. Epithelial acinar tumors include rare entities such as pancreatoblastoma, acinar cell carcinoma, giant cell tumors, oncocytic tumors, choriocarcinoma, colloid carcinoma, medullary tumors, and adenosquamous tumors [15]. These tumors have unique histopathological and serological characteristics that help differentiate them from other neoplasms including well-defined margins and elevated serum markers, such as alpha-fetoprotein in pancreatoblastomas and lipase in acinar cell carcinomas [17, 18, 19]. Epithelial endocrine tumors include insulinomas, glucagonomas, gastrinomas, VIPomas, and somatostatinomas, as well as carcinoid [20]. They exhibit increased enhancement compared to adjacent pancreatic parenchyma secondary to their hypervascularity [15].

Mesenchymal pancreatic tumors, such as the PEComa in our case, constitute the rarest type of pancreatic solid neoplasms [15]. Besides PEComas, other tumors in this category include schwannomas, fibromas, lipomas, fibrosarcomas, liposarcomas, hemangiomas, malignant hemangiopericytomas, lymphomas, and leiomyosarcomas [15]. Though rare, these tumors should be considered if a pancreatic mass does not readily fit into one of the more common categories of pancreatic lesions.

In our opinion, the presumptive diagnosis of a mesenchymal tumor in our patient was suggested by

the CT and ultrasound imaging. The well-defined, encapsulated appearance of the mass without signs of local invasion was not consistent with a primary pancreatic ductal adenocarcinoma [16]. Acinar tumors are not encapsulated, decreasing the likelihood of this diagnosis [15]. Endocrine tumors can occasionally present with pseudo-capsules; however, they are hypervascular as compared to the surrounding parenchymal pancreas in contrast to the tumor in our patient making the diagnosis of mesenchymal tumor a more likely diagnostic possibility [15].

Because of the rarity of pancreatic PEComas, much uncertainty remains regarding the prognosis for patients with these tumors. However, a recent study proposed classifying individual PEComas as benign, uncertain malignant potential, and malignant [20]. Malignant PEComas can present as high grade sarcomas with local invasion and distant metastasis mandating aggressive therapy [5]. Current management has relied on surgery because irradiation and chemotherapy have failed to yield significant benefit [5]. Further study is warranted to determine the optimal management of these rare tumors. Early recognition of pancreatic PEComas on imaging could dramatically impact both patient therapy and prognosis compelling us to report this case.

In conclusion, we believe that radiologists should consider the diagnosis of pancreatic PEComa if they encounter a well-defined, encapsulated, and hypo-vascular pancreatic mass.

Conflict of interest The authors have no potential conflicts of interest

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