

The Differential Broadens. EUS FNA Appearance and Cytological Findings of Pancreatic Angiomyolipoma

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ABSTRACT

Context Angiomyolipoma is a rare tumor characterized histologically by a mixture of spindle cells, adipose tissue, epithelioid cells, and vascular tissue. It usually involves the kidney followed by the liver whereby the majority of affected patients are female, and many cases arise in the setting of tuberous sclerosis.

Case report We report a case of a 33-year-old female with an asymptomatic incidental right renal mass suggestive of an angiomyolipoma in conjunction with numerous pancreatic masses.

Conclusions The utility of EUS in the differential diagnosis of pancreatic tumors is well established. This is the first known

reported EUS detection and FNA confirmation of angiomyolipoma metastatic to the pancreas and should now be added to the already broad differential of metastatic pancreatic tumors.

INTRODUCTION

Renal angiomyolipomas, sometimes referred to as clear-cell “sugar” tumors, belong to the perivascular epithelioid cell family of tumors. Although slow growing, their malignant potential has been demonstrated. The majority of cases have been reported in women. However, the rarity of this tumor prohibits clear determination of both the gender predominance and malignant potential [1, 2]. They originate from the renal parenchyma and contain smooth muscle, vascular elements and fatty tissue [1]. To date, angiomyolipomas have been described in numerous extra-renal sites including the skin, shoulder, forearm, soft palate, larynx, lung, liver, pancreas, colon, rectum, uterus, and ovary [3, 4, 5, 6, 7, 8, 9, 10, 11].

CASE REPORT

An asymptomatic 33-year-old female underwent a transabdominal abdominal ultrasound as a volunteer participant in a clinical study which revealed a 26 mm right renal mass suggestive of an angiomyolipoma. In addition, while three hyperechoic pancreatic masses (head, 10 mm; body, 10



Figure 1. Computed tomography demonstrates a 2.6 cm angiomyolipoma in the upper lateral aspect of the right kidney.

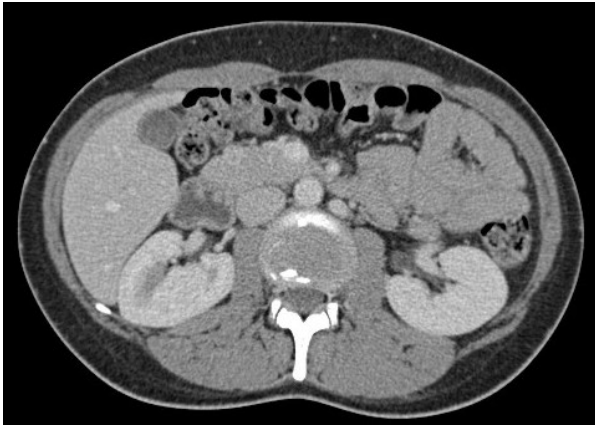


Figure 2. Computed tomography (venous phase) demonstrates an enhancing 10 mm lesion within the pancreatic head/neck.

mm; tail, 7 mm) were identified, they differed in appearance from the renal mass. These same findings were then demonstrated on abdominal computed tomography (CT) (Figures 1, 2 and 3). Upon retrospective review of an abdominal CT performed 4 years prior, to evaluate for abdominal pain, a 0.7 cm renal mass was noted with a normal appearing pancreas. She has no evidence of tuberous sclerosis, which has been associated with angiomyolipoma. However, she uses a birth control patch that has been associated with growth of this tumor type.

Subsequent EUS revealed 5 pancreatic masses. Two were located in the pancreatic tail and appeared iso/hypoechoic and measured 12x8 mm (with internal cysts) and 16x9 mm (with internal calcification) (Figure 4). One lesion in the body measured 10x7 mm and was hypoechoic with a subtle hyper-



Figure 3. Computed tomography reveals a 9 mm enhancing mass in the pancreatic tail lying in close proximity to a small focus of calcification.



Figure 4. Endoscopic ultrasound appearance of the 16x9 mm lesion located in the pancreatic tail that contains a focus of calcification.

echoic rim (Figure 5). One lesion in the pancreatic neck was hypoechoic, measured 6x6 mm, and contained cystic spaces. A lesion in the uncinate measured 13x10 mm and contained internal cysts. EUS also demonstrated the right renal mass that was hyperechoic and measured 25x20 mm. There was no evidence of malignant lymphadenopathy or liver metastasis.

Four FNAs were taken each from a pancreatic tail and body lesion. Cytology revealed mature adipocytes mixed with clusters of spindle or oval-shaped cells with indistinct cytoplasmic borders, fine nuclear chromatin and minimal atypia. No thick-walled blood vessels were noted in the cytological smears. The cytological features were suggestive of angiomyolipoma (Figure 6).

DISCUSSION

Ramuz *et al.* reported the EUS appearance of a pancreatic “sugar” tumor, as a hypoechoic nodule suggestive of a benign endocrine

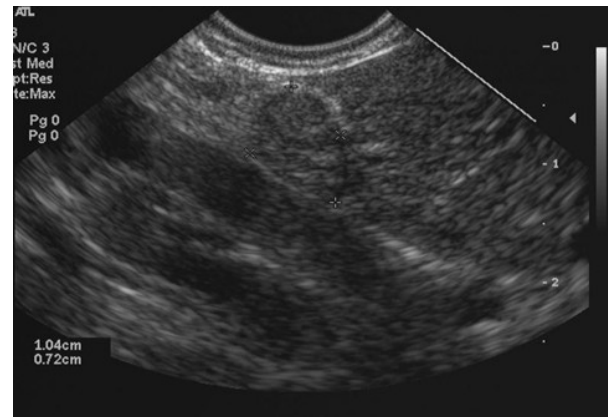


Figure 5. Endoscopic ultrasound reveals a 10x7 mm isoechoic lesion with a subtle hyperechoic rim located in the pancreatic body.

tumor. Although EUS FNA in their patient was negative, the diagnosis was established following laparoscopic resection [12]. This is the first known reported EUS detection and FNA confirmation of angiomyolipoma metastatic to the pancreas and should now be added to the already broad differential of metastatic pancreatic tumors. The utility of EUS in the differential diagnosis of pancreatic tumors is well established. Imaging alone, however, may not reliably establish the tumor type or distinguish primary versus metastatic lesions. The appearance has been shown to vary considerable and often differs from the primary tumor site [13]. In our patient, while the appearance varied between that of the primary and metastatic tumors, the presence of multiple pancreatic lesions offered a clue to the diagnosis. Clear distinction between primary and metastatic pancreatic tumors is important given their varied prognosis and management. For this reason and because of the unreliability of image morphology, we considered FNA essential for this patient. In addition to confirming the diagnosis, the EUS exam more fully defined the extent of disease, which is likely to impact the extent of resection, should surgery become necessary. Following a multidisciplinary meeting to include the general surgery and urology services, the overall consensus was that this lady had multi focal small pancreatic lesions representing angiomyolipoma in the absence

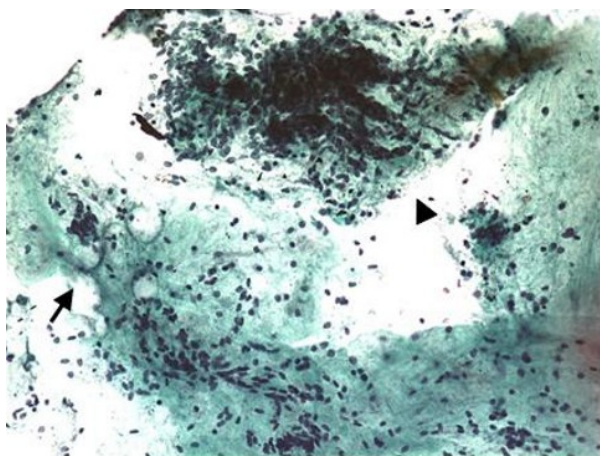


Figure 6. FNA smears of angiomyolipoma showed sheets or bundles of spindle and oval cells (arrow head) admixed with mature fat cells (arrow) (Papanicolaou stain, x200).

of tuberous sclerosis. However, a literature review revealed limited information pertaining to the need and timing of resection as the pathological behavior of angiomyolipoma has not been clearly elucidated. It was elected to perform a partial right nephrectomy and defer surgical intervention of the pancreas. There are data suggesting that hormone stimulation, via estradiol or tamoxifen administration or during pregnancy, can stimulate angiomyolipoma cell growth [14, 15, 16]. Use of a birth control patch may account for the interval changes in our patient and we have encouraged her to seek alternate means of birth control. While there are no established guidelines pertaining to pancreatic angiomyolipoma surveillance, we will perform a CT and EUS one year following the last exam to evaluate for interval growth and/or new lesions.

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