INTRODUCTION

Mesenchymal tumors of the pancreas are rare and they account for only 1–2% of all primary pancreatic tumors (1). In order of decreasing frequency, the most commonly reported primary benign or intermediate (borderline) mesenchymal tumors of the pancreas are as follows: schwannoma, inflammatory myofibroblastic tumor, solid and cystic hamartoma, and solitary fibrous tumor (2). Owing to very few case reports describing primary mesenchymal tumors of the pancreas, no definitive protocol for the treatment of these lesions has been established (3).

Primary pancreatic angiomyolipoma (AML), a part of a family of mesenchymal tumors, is a common fat-containing solid neoplasm. Kidney and liver are the main sites of AML; rarely, primary pancreatic AML has also been reported. Here, we present a case of pathologically proven primary pancreatic AML in a middle-aged female patient, based on multidetector computed tomography scan, endoscopic ultrasound, positron emission tomography, and magnetic resonance imaging findings.

CASE REPORT

A 59-year-old female patient was referred to our hospital for treatment of a pancreatic mass. The mass was incidentally noted on a MDCT scan for abdominal trauma work-up. Her physical examination was unremarkable, and laboratory findings including carcinoembryonic antigen and cancer antigen 19-9 levels were within normal limits. In addition to the abdominal MDCT scan, we performed EUS, PET, and MRI for further evaluation of the pancreatic tumor.

On a CT scan (Somatom Sensation-16; Siemens Medical Solutions, Erlangen, Germany and Brilliance 64; Philips Medical Systems, Cleveland, OH, USA), non-enhanced images demonstrated a 2-cm isodense mass with a mean attenuation value of 48 Hounsfield units (HU) and no evidence of fat components or calcifications. Enhanced CT scan images demonstrated peripheral enhancement during the arterial, parenchymal, and portal venous phases (Fig. 1A). The mass displayed a well-defined margin, no invasion of adjacent organs, and no signs of internal hemor-
rhage, necrosis, or duct dilatation. EUS revealed a 2-cm hyper-
echogenic mass in the body of the pancreas (Fig. 1B) without 
hypervascularity noted on Doppler ultrasound. On a PET scan, 
the mass exhibited a maximum standardized uptake value of 2.2, 
which suggested a low-grade malignant or benign tumor. On 
MRI (Avanto; Siemens Medical Solutions), the mass displayed a 
heterogeneous peripheral high signal intensity (SI) on half-
Fourier acquisition single-shot turbo spin-echo T2-weighted imag-
ing [repetition time (TR)/echo time (TE), 1000/154; flip angle, 
160°; section thickness, 6 mm], and a homogenous low SI on T1-
weighted images (TR/TE, 160/4.92; flip angle, 70°; section thick-
ness, 6 mm). Chemical shift gradient-echo MR imaging showed 
no definite loss of SI. On diffusion weighted imaging (DWI; b-
value of 1000), the mass revealed peripheral high SI (Fig. 1C). 
But on the apparent diffusion coefficient map, the periphery of 
the tumor did not show low SI. We suggested that high SI on DWI 
was due to the T2 shine-through artifact, not true diffusion re-
striction. Dynamic MRI also revealed strong peripheral en-
hancement and poor central enhancement during the arterial 
phase. As the arterial phase transitioned to the parenchymal and 
portal venous phases, the mass showed heterogeneous enhance-
ment with peripheral isoenhancement and central subtle poor 
enhancement. Considering all the results, the final radiographic 
diagnosis indicated a neuroendocrine tumor in the body of the 
pancreas. Subsequently, a medial pancreatectomy was performed.

On gross pathological examination, the mass appeared as a 
well-defined, gray-white nodular tumor in the body of the pan-
creas measuring 2 × 2 cm. Hematoxylin and eosin staining re-
vealed that the mass was composed of blood vessels, adipose 
tissue, and spindle cells. Additionally, the mass had a hypercel-

tular central area and a hypocellular peripheral area with loose 
connective tissue (Fig. 1D). This histologic finding of the tumor 
was correlated with the enhancement pattern on the MDCT 
and MRI images. The immunohistochemistry analysis revealed 
that the mass had positive immunoreactivity to human melano-
ma black-45 (HMB-45) and smooth muscle actin (SMA) (Fig. 
1E). Accordingly, the final pathological diagnosis of a primary 
pancreatic AML was determined. No signs of recurrence or me-
A 59-year-old woman with primary angiomyolipoma of the pancreas.

**D.** Gross pathologic examination (left photograph) shows a 2 × 2 cm, well-defined, gray-white nodular tumor (white arrow) in the body of the pancreas. Hematoxylin and eosin staining (right photograph, ×100) reveals that the mass is composed of thick-walled blood vessels (open arrow), adipose tissue (black arrows), and spindle cells. Compared with the central area of the tumor, the peripheral area shows low cellularity with fibri-noid-like connective tissue.

**E.** HMB-45 (left photograph) and SMA (right photograph) staining (immunohistochemistry, ×200) show diffusely positive immunoreactivity. Considering these results (D, E), the final pathologic diagnosis of the pancreatic tumor was determined as primary pancreatic AML.

AML = angiomyolipoma, HMB-45 = human melanoma black-45, SMA = smooth muscle actin

tastasis have been identified on follow-up CT scans performed over 30 months.

**DISCUSSION**

The incidence and general radiographic features of primary pancreatic AML are not known; this is because this disease has previously been reported only twice in the literature (2, 3). Pathologically, AML is composed of a mixture of smooth muscle, adipose tissue, and blood vessels. AMLs react with HMB-45 and SMA on immunohistochemical analysis (5). The mass in our case displayed the same pathological and immunohistochemical characteristics.

The imaging findings associated with the mass in our case differed from those of the first reported primary pancreatic AML, renal AML, and hepatic AML. In 2004, Heywood et al. (3) described the first known primary pancreatic AML. This mass was located in the uncinate process and was accompanied by hemorrhage, measuring 4.5 × 3 × 2.5 cm. Most of the pathological and immunohistochemical results in that report were simi-
lar to those in our case; however, other results such as imaging findings were quite different, which may be due to hemorrhage. The mass in that case appeared heterogeneous on US and as an irregular, thick-walled, cystic mass on a CT scan (3).

Renal AML is located in the renal cortex and exhibits diffusely high echogenicity on US (6). Most renal AMLs demonstrate fat density (less than -20 HU) on a non-enhanced CT scan, although lipid-poor AMLs show high attenuation. On an enhanced CT scan, they show homogeneous enhancement with a prolonged enhancement pattern (6-8). The MRI signals vary depending on the amount of intratumoral fat. However, with the exception of lipid-poor AMLs, most renal AMLs are easily detected using a fat suppression or chemical shift technique (7, 8).

Hepatic AMLs appear as well-circumscribed, hyperechogenic lesions and may show relative hypervascularity on US (8, 9). Unlike renal AMLs, 50% of hepatic AMLs lack an appreciable fat content. As a result, hepatic AMLs show two different imaging types on a CT scan. One type is a lipid-poor hepatic AML with a peripheral angiomyomatous component and soft tissue attenuation. The other type is a lipid-rich hepatic AML with an attenuation value less than -20 HU (9). In the early phase, hepatic AMLs demonstrate marked enhancement with or without visible large central vessels (10). On MR, hepatic AMLs have the same imaging characteristics as renal AMLs (7-10). In conclusion, CT and MRI imaging findings of a primary pancreatic AML in our patient differ from those of the first reported primary pancreatic AML as well as renal and hepatic AMLs.

To date, this is the first known report describing CT, EUS, PET and MRI imaging findings of a primary pancreatic AML without hemorrhage. This report has limitations owing to the scarcity of documented cases of primary pancreatic AML. Further evaluation is recommended to establish the general imaging features, standard imaging modalities, and prognostic factors.

REFERENCES

췌장의 원발성 혈관근육지방종의 영상소견 1예: 증례 보고

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혈관근육지방종(angiomyolipoma)은 위장관간질성종양(mesenchymal tumor)에 속하며, 지방을 포함하는 고형종괴로서 흔히 볼 수 있다. 신장과 간은 혈관근육지방종이 호발하는 부위지만, 췌장의 원발성 혈관근육지방종은 보고된 바가 드물다. 저자들은 조직병리학적으로 진단된 증상과 반복적으로 병발된 췌장의 혈관근육지방종 1예를 경험하였기에 다중결절검출기 CT, 내시경 초음파, 양전자 방출 단층촬영, 자기공명영상 소견을 중심으로 보고하고자 한다.

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