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Positron emission tomography/computed tomography manifestations of primary hepatic myxoid liposarcoma
A case report

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Abstract

Rationale: Computed tomography (CT) and magnetic resonance imaging (MRI) are conventional used to evaluate liver tumors. Detection of fat component is considered an important clue to the imaging diagnosis of hepatic myxoid liposarcoma. However, the positron emission tomography (PET)/CT report of hepatic myxoid liposarcoma is scarce, and the metabolic characteristics of primary hepatic myxoid liposarcoma are still unknown.

Patient Concerns: In this report, we report the PET/CT manifestations of a rare case of primary hepatic myxoid liposarcoma that was confirmed by pathologic examination. A 29-year-old male patient presented with a nodule adjacent to the umbilicus that had been present for 2 weeks. PET/CT showed a hypoattenuating mass with moderate uptake (maximum standardized uptake value [SUVmax] 1.9), except for a slightly hyperattenuating focal area with intense uptake (SUVmax 3.1).

Diagnoses: The final pathologic diagnosis was confirmed, by means of laparotomy, to be a primary hepatic myxoid liposarcoma with multiple extrahepatic metastases.

Interventions: The lesions in the left liver and periumbilical abdominal wall were punctured. During the laparotomy, hepatic malignant tumors were confirmed and metastasized widely in the abdominal cavity.

Outcomes: An abdominal CT performed 4 months after initial presentation showed extensive metastasis.

Lessons: Primary hepatic myxoid liposarcoma may manifest as moderate metabolism with less fat on PET/CT. PET/CT is not only valuable in reflecting the round cell component of hepatic myxoid liposarcoma, but also in estimating its origin.

Abbreviations: FDG = fluorodeoxyglucose, MRI = magnetic resonance imaging, PET/CT = positron emission tomography/computed tomography, SUVmax = maximum standardized uptake value, T1WI = T1-weighted images, T2WI = T2-weighted images.

Keywords: computed tomography, hepatic liposarcoma, imaging, magnetic resonance imaging, positron emission tomography/computed tomography

1. Introduction

Myxoid liposarcoma is the second most common type of liposarcoma, accounting for 30% to 40% of all liposarcomas.[1] Myxoid liposarcoma most commonly occurs in the lower extremities, followed by the buttocks, retroperitoneum, and trunk.[1] Primary hepatic liposarcoma is very rare; <12 cases have been identified to date.[2,3] Detection of fatty tissue in the tumor helps to narrow the diagnosis. Primary hepatic myxoid liposarcoma should be further differentiated from other fat-containing masses such as angiomyolipoma, hepatocellular cancer with steatosis, and teratoma.[4] Recently, positron emission tomography (PET)/computed tomography (CT) was confirmed to be capable of differentiating hibernoma from liposarcoma in the thigh.[5] However, literature regarding PET/CT evaluation of hepatic liposarcoma is still scarce. Furthermore, there is no report about the PET/CT findings of hepatic myxoid liposarcoma. In this case study, we discuss the PET/CT images of a pathology-confirmed rare hepatic myxoid liposarcoma.

2. Case presentation

A 29-year-old male presented with a small nodule above the umbilicus that was first noticed 2 weeks prior to presentation. Abdominal ultrasound examination of the lesion, performed at a local hospital, revealed a mass in the left lobe of the liver that was most likely a liver tumor with abdominal metastasis. The patient had no obvious symptoms during the course of his evaluation and treatment. He was referred to our institution for further diagnosis and treatment. Physical examination revealed a firm mobile nodule above the umbilicus. Laboratory testing revealed a slightly elevated direct bilirubin (9 μmol/L, reference range 1–7 μ
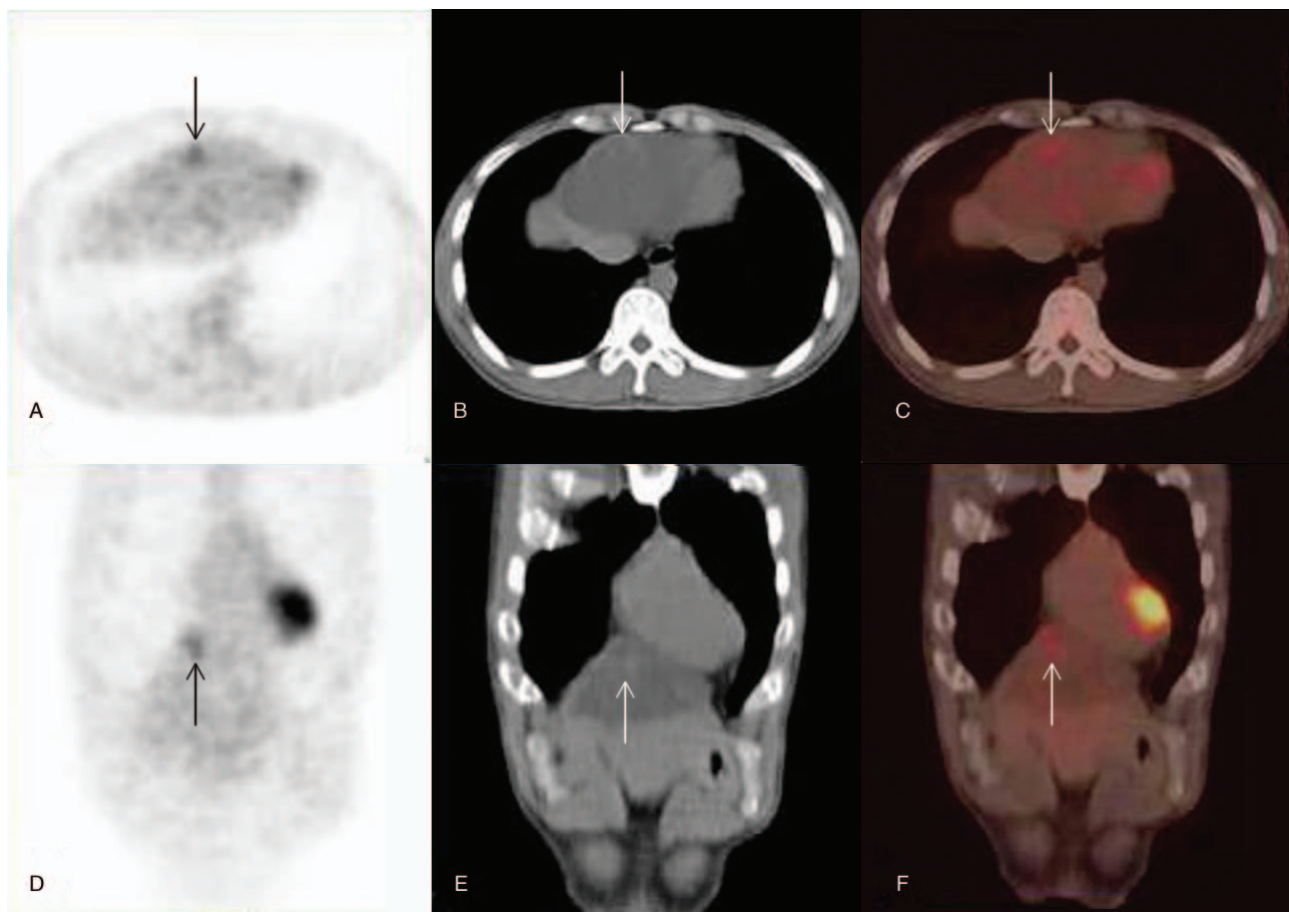
Figure 1. A large hepatic mass was detected by ^{18}fluorodeoxyglucose (^{18}F-FDG) positron emission tomography/computed tomography (PET/CT) imaging (A, D: PET images; B, E: CT images; C, F: fusion images). Near the diaphragmatic surface of the left liver lobe, a large hypoattenuating mass was identified with a small ring-shaped mildly hyperattenuating enhancement (arrow, B, E) on CT. Intense focal uptake (arrow, A, D) was found in the mass with a maximum standardized uptake value (SUVmax) of 3.1.

mol/L); other liver function indices were all within normal limits. Serum tumor markers were also within normal limits, including alpha-fetoprotein, carcinoembryonic antigen, carbohydrate antigen (CA) 199, CA 125, ferritin, and total prostate-specific antigen.

Abdominal noncontrast CT revealed a well-circumscribed hypoattenuating mass measuring approximately $9.1 \times 9.6\,cm^2$ in the left lobe of the liver with slight heterogeneity. Contrast-enhanced CT imaging showed nodularity and ring-shaped enhancement of the hepatic mass. An additional nodule with attenuation and an enhancement pattern similar to the hepatic lesion was also observed in the middle of the anterior abdominal wall. Abdominal magnetic resonance imaging (MRI) revealed a well-circumscribed mass in the left liver lobe that was heterogeneously hypointense on T1-weighted images (T1WIs) and hyperintense on T2-weighted images (T2WIs). There was signal loss of the tumor on out-of-phase MRI. Furthermore, the mass showed slightly increased signaling in diffusion-weighted MR images ($b = 800$). Contrast-enhanced MRI also showed nodularity and ring-shaped reinforcement. The signal intensity and enhancement pattern of the nodule in the middle of the anterior abdominal wall were similar to the imaging characteristics of the hepatic lesion.

To further evaluate the lesions, the patient was examined with a Siemens Biograph 16 PET/CT scanner (Siemens Medical Solutions USA, Inc, Malvern, PA) with intravenous injection of fluorodeoxyglucose (FDG) at 5.5 MBq/kg. PET/CT revealed a hypoattenuating mass approximately $8.3 \times 11.0\,cm^2$ (11.2 HU) in the left lobe of the liver. Radioactive uptake of the mass was moderate, that is, uptake of the mass was similar to hepatic parenchyma uptake, with a maximum standardized uptake value (SUVmax) of 1.9. A small, ring-shaped, mildly hyperattenuating area of the mass showed intense uptake, that is, lesion uptake was higher than hepatic parenchyma uptake, with an SUVmax of 3.1 (Fig. 1). The SUVmax was 3.6 on the delayed scan. Multiple small hypoattenuating nodules were revealed below the umbilicus in the abdominal wall and peritoneum, which showed slight uptake with an SUVmax of 1.8.

To clarify the diagnosis, the patient underwent laparotomy. The tumor could not be excised via surgery due to extensive intraperitoneal metastasis. The diagnosis of hepatic myxoid liposarcoma was confirmed by pathology (Fig. 2). The patient then returned to the local hospital for chemotherapy. Four months later, abdominal CT showed extensive metastasis in the abdominal and pelvic cavities. The patient died a few weeks later due to extensive recurrence of the tumor.

3. Discussion

The age of onset of primary liver liposarcomas ranges from 2 to 86 years old, with a mean age of 47 years old, without obvious
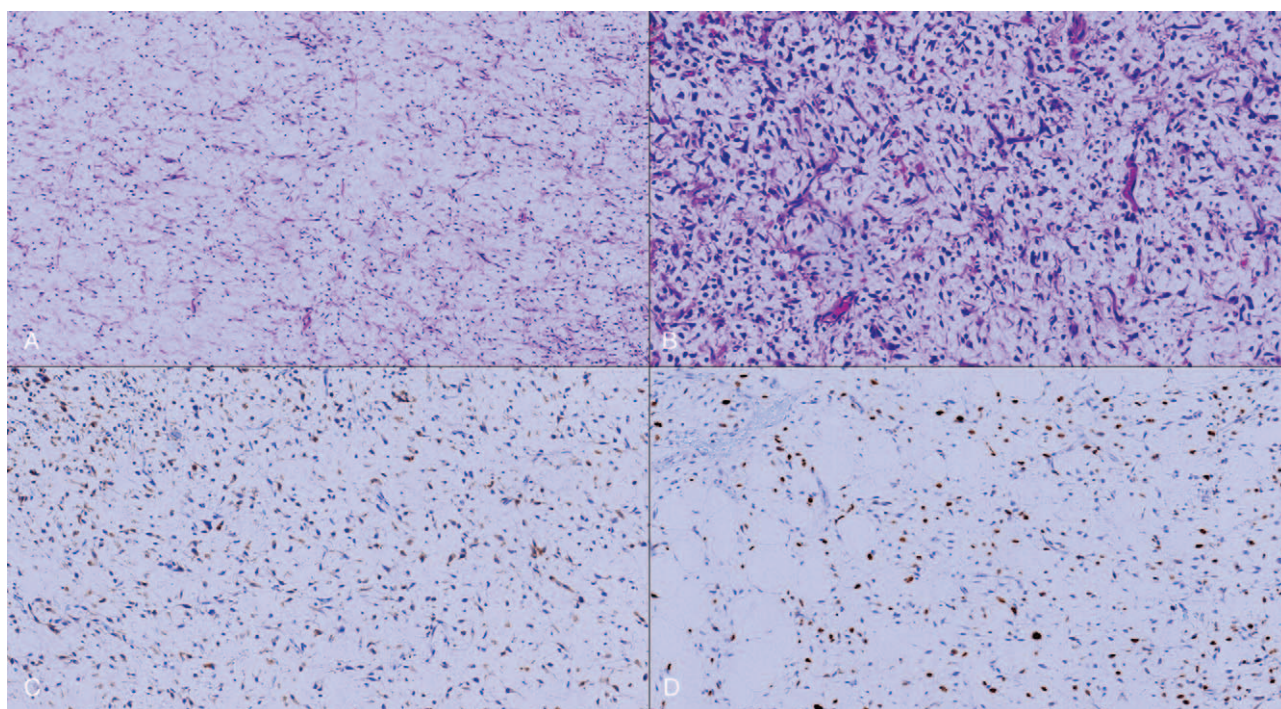
Figure 2. Photomicrograph shows a large area of loose myxedematous matrix (A) and focal short shuttle cells with heterogeneous nuclei (B). Immunohistochemistry (IHC) demonstrates S-100 as positive (C) and Ki-67 as 20% (D) (×100 A, B; ×200 C, D).

gender preference.[3–13] Primary symptoms of liver liposarcomas include abdominal pain, nausea, vomiting, jaundice, and weight loss.[3–13] These symptoms typically result from the compression of peripheral nerves, vessels, and the biliary tract or other nearby organs by the abdominal mass.[6] Laboratory tests may reveal abnormality of liver function indices, including alanine aminotransferase, aspartate aminotransferase, gamma-glutamyl-transpeptidase, and total bilirubin.[3,5,6,10] Tumor markers have not shown any diagnostic value in previous reports, nor did they in this study, except for one reported case that had an increase of the CA 199 level.[6] The patient in this case study presented with an abdominal wall nodule, which was found to be an extrahepatic metastatic liposarcoma. The clinical progression of this case is significantly different from that of previous reports.

Liposarcomas can be divided into 4 subtypes: well-differentiated liposarcoma, de-differentiated liposarcoma, myxoid-round cell liposarcoma, and pleomorphic liposarcoma.[14] Hepatic myxoid liposarcomas account for approximately half of liver liposarcomas.[3–13] The characteristic histopathologic finding of myxoid-round cell liposarcomas is the presence of hypocellular mild spindle cells in a rich myxoid background.[14] The genetic abnormality of myxoid liposarcoma is a reciprocal chromosomal translocation of t(12;16)(q13;p11), and the affected genes include DDIT3 (CHOP) and FUS (TLS).[15,16] The resultant abnormality is the FUS-DDIT3 chimeric gene.

The CT findings of hepatic liposarcomas included a well-defined hypoattenuating mass with cystic degeneration, occasional bleeding, and margin enhancement.[3,6,7,9,11,12] The MRI findings of hepatic liposarcomas included T1WI and T2WI hyperintensity of fat-containing components, T1WI hypointensity and T2WI hyperintensity of other components, and signal loss with fat suppression.[5,9] Similarly, hepatic myxoid liposarcomas containing myxoid components and nonfat nonmyxoid components showed heterogeneous enhancement on contrast-enhanced MRI.[17] In this case, CT did not show any detectable fat component but did reveal heterogeneous reinforcement. In-phase and out-of-phase MRI confirmed the presence of a fat component in this case. The identification of a fat component of the tumor provided the basis for an image-based diagnosis of hepatic myxoid liposarcoma.

The PET/CT revealed moderate uptake of the hepatic myxoid liposarcoma, which was consistent with the hypocellularity of the mass.[17] The area of intense focal uptake on PET/CT was consistent with the MRI-identified nonfat nonmyxoid component, which reflects the nature of round cell clusters.[18] Thus, PET/CT alone could not fully reflect the molecular biologic behaviors of myxoid liposarcoma, but when combined with MRI allowed for a comprehensive evaluation of hepatic myxoid liposarcoma. The unique advantage of PET/CT was the confirmation that the tumor in this case originated from the liver, thereby excluding the diagnosis of metastatic liver liposarcoma.

Fine-needle aspiration biopsy has not been shown to be a valuable diagnostic tool when evaluating primary hepatic liposarcoma. Only 1 case was confirmed as liposarcoma with this method.[3,5,7,9] Surgical resection is regarded as the treatment of choice for primary liposarcomas, and surgical therapy can also be used to treat recrudescent resectable foci.[6] Radiotherapy is modestly effective, but can only be applied to specific areas.[3] The 5-year survival rate of hepatic liposarcomas after surgery or radiotherapy is about 50%.[7] It has also been reported that older age (>45 years), necrosis, and high histologic grading are associated with a poorer prognosis in myxoid liposarcoma patients.[19]

In general, PET/CT evaluation revealed that primary hepatic myxoid liposarcomas could be represented as a moderately metabolic tumor with little fat. Primary hepatic myxoid liposarcoma should be included in the differential diagnosis of liver disease with moderate uptake on PET imaging.

Author contributions

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