Nested Stromal-epithelial Tumor of Liver with Recurrent Extrahepatic Metastasis: Role of Fluorodeoxyglucose Positron Emission Tomography/Computed Tomography

Abstract

Nested stromal-epithelial tumor (NSET) is a very rare nonhepatocytic and nonbiliary primary tumor of the liver. An 8-year-old boy was incidentally detected with hepatic lesions, involving both lobes of the liver for which he later underwent orthotopic liver transplant. The hepatic lesions were confirmed to be NSET following histopathological examination of explant liver specimen. He later developed recurrence with multiple metastatic lesions, including multi-station nodal and right talar bone involvement. We here present the case highlighting the importance of ^{18}F-fludeoxyglucose positron emission tomography/computed tomography in the management of this rare tumor, in particular for monitoring disease progression and/or recurrence.

Keywords: *^{18}F-fludeoxyglucose positron emission tomography/computed tomography, hepatic tumor, liver transplantation, nested stromal-epithelial tumor*

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An 8-year-old boy presented with high-grade fever. Laboratory investigation revealed pancytopenia with deranged coagulation profile. The computed tomography (CT) and magnetic resonance imaging (MRI) scan of the abdomen demonstrated diffuse masses throughout the liver [Figure 1]. The hepatic masses were found to be fludeoxyglucose (FDG) avid on positron emission tomography (PET)/CT [Figure 1]. Tumor markers, including alpha-fetoprotein and carcinoembryonic antigen, were within normal range. The patient underwent biopsy of one of the mass, and on histopathological examination, it was found to be consistent with undifferentiated malignant tumor, suggesting diagnosis of sarcoma. Given the diffuse hepatic involvement and no extrahepatic metastases, the patient underwent orthotopic liver transplant. The tumor in explant liver specimen was re-examined and was found to be a nested stromal-epithelial tumor (NSET). Postoperative period was unremarkable. Postliver transplant, the patient was started on immunosuppressant therapy (tacrolimus).

Two months posttransplant, on a follow-up PET/CT examination, multiple FDG avid mediastinal and aortocaval lymph nodes (LNs) were detected. For mediastinal LNs, the patient underwent thoracic LN resection. However, during laparoscopic abdominal exploration, the aortocaval LNs were found to be intimately attached to the pancreas, superior mesenteric artery, and inferior vena cava. Therefore, the patient was treated with chemotherapy for aortocaval LNs. Following which, on subsequent PET/CT the aortocaval LNs became FDG negative. The patient later presented with pain in the right lower limb and MRI of the right lower extremity revealed new metastases in talus of the right foot [Figure 2]. The PET/CT confirmed right talar metastases and in addition showed multiple lymph nodal metastases [Figure 2]. Patient was started on proton beam radiation therapy to abdomen and right foot for abdominal LNs and right talus metastases, respectively. Since then patient has been tolerating the therapy well, with pain in the foot reasonably controlled with acetaminophen.

NSET is a nonhepatocytic and nonbiliary primary tumor of the liver.[1] It is characterized by cellular nest composed of spindled or epithelioid cells surrounded by desmoplastic stroma with variable calcification and ossification.[2]

NSET is a very rare tumor with few reported cases in the literature.[3] Most of

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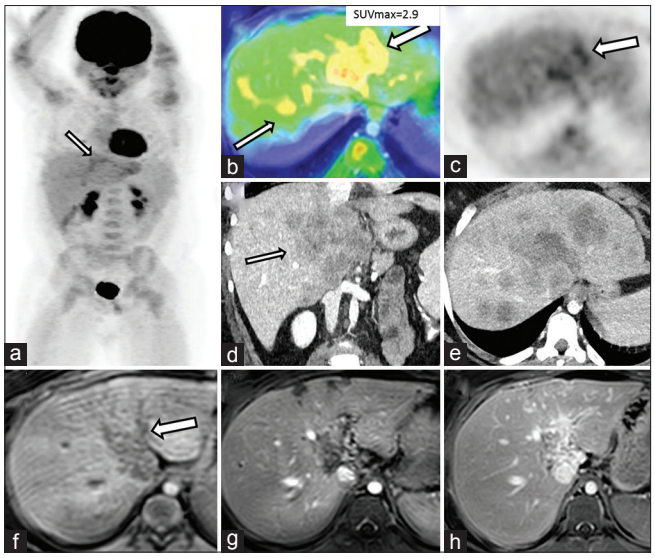

Figure 1: ^{18}F-fludeoxyglucose positron emission tomography, ^{18}F-fludeoxyglucose positron emission tomography/computed tomography, computed tomography and magnetic resonance imaging of nested stromal epithelial tumor of the liver (white block arrow). Maximum intensity projection (MIP) (a), axial positron emission tomography/computed tomography (b) and positron emission tomography images (c) showed multiple fludeoxyglucose-avid lesions throughout the liver. The lesions were hypodense on coronal (d) and axial (e) contrast enhanced computed tomography. The lesions showed heterogeneous signal intensity on T1-weighted image (f) with no enhancement on arterial phase (g) and heterogeneous enhancement on delayed phase images (h)

these cases have been reported in young females, in the right lobe of the liver.[4] Its pathogenesis remains unclear, however, based on some immunohistochemical studies these neoplasms are suspected to originate from hepatic mesenchymal precursor cell with primitive differentiation along the bile duct lineage.[1] These tumors are often detected incidentally on imaging.[1] Few cases have been reported in association with Cushing syndrome and Beckwith-Wiedemann syndrome. Diagnosis is made by histopathological examination. Immunohistochemical examination can help confirm epithelioid (CD56, CD117, AE1/AE3, keratin), nested spindled cells (vimentin), and stromal myofibroblastic cells (smooth muscle actin) in the tumor.

Surgical resection of the primary tumor mass is the first line of treatment, providing the best prognosis.[5,6] Liver transplant can be useful in patients who have untreatable liver mass and no extrahepatic disease. Since these tumors have a known metastatic potential,[1] a postoperative follow-up with PET can be helpful.[1,5,7,8]

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Conflicts of interest

There are no conflicts of interest.

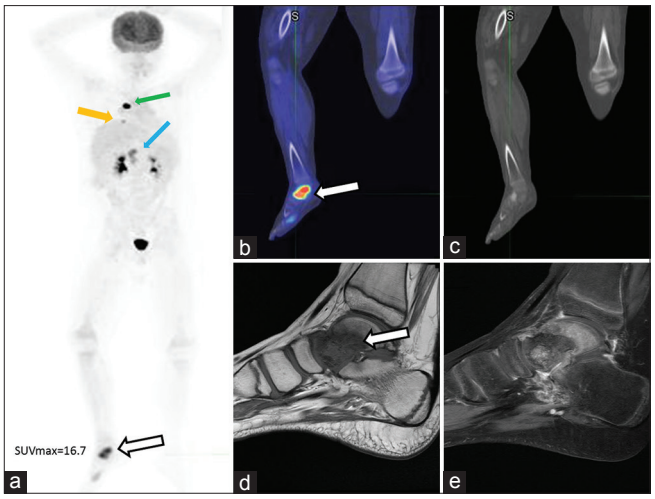

Figure 2: ^{18}F-fludeoxyglucose positron emission tomography, ^{18}F-fludeoxyglucose positron emission tomography/computed tomography, computed tomography and magnetic resonance imaging of right talar metastasis (white block arrow). MIP (a), positron emission tomography/computed tomography (b) and computed tomography images (c) showed fludeoxyglucose-avid lesion in the right talus. The lesion showed heterogeneous signal intensity on sagittal T1-weighted (d) and T1-weighted fat saturated (e) images. Additional fludeoxyglucose avid pericaval (blue arrow), mediastinal lymph nodes (yellow arrow), and an implanted vascular access device (green arrow) were seen on MIP image (a)

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