

¹⁸F-fluorodeoxyglucose positron emission tomography/computed tomography in childhood metastatic epithelioid hemangioendothelioma of the liver: Altering the management plan

ABSTRACT

Hepatic epithelioid hemangioendothelioma (EHE) is an uncommon vascular tumor, primarily seen in 20–40 years of age. We report the case of a 14-year-old boy with EHE and distant metastases identified on 18F-fluorodeoxyglucose positron emission tomography/computed tomography (¹⁸F-FDG PET/CT), leading to a change in the management plan. ¹⁸F-FDG PET/CT proved to be useful in identifying the rare sites of metastasis and planning further treatment.

Keywords: 18F-fluorodeoxyglucose positron emission tomography/computed tomography, epithelioid hemangioendothelioma, hepatic hemangioendothelioma

INTRODUCTION

Hepatic hemangioendothelioma is a rare vascular tumor, primarily diagnosed on computed tomography (CT) imaging due to its characteristic features. However, 18F-fluorodeoxyglucose positron emission tomography/CT (¹⁸F-FDG PET/CT) has an added advantage of whole-body imaging and the ability to capture the metabolic status of the disease process. Here, we report a 14-year-old boy who underwent PET/CT for metastatic workup of hepatic epithelioid hemangioendothelioma (EHE). 18F-FDG PET/CT proved useful in an accurate detection of metastatic disease, thus altering the management plan for the patient.

CASE REPORT

A 14-year-old boy presented with pain in the right hypochondrium for 5 months, associated with hepatomegaly. Ultrasonography of the abdomen revealed multiple, predominantly hypoechoic liver lesions. Contrast-enhanced magnetic resonance imaging (CEMRI) of the abdomen showed moderate ascites and hepatomegaly with multiple mass

lesions in both lobes of the liver with restricted diffusion and peripheral enhancement on arterial and venous phases. In view of the US and CEMRI findings, suggesting hepatic EHE, the patient was planned for liver transplantation. Subsequently, ¹⁸F-FDG PET/CT was performed for lesion characterization and evaluation of disease extent [Figure 1]. An ultrasound-guided fine-needle aspiration cytology, performed on the liver lesion, revealed hemangioendothelioma on histopathological examination. With the diagnosis of metastatic hepatic EHE, the transplant was deferred, and the patient was advised for chemotherapy.

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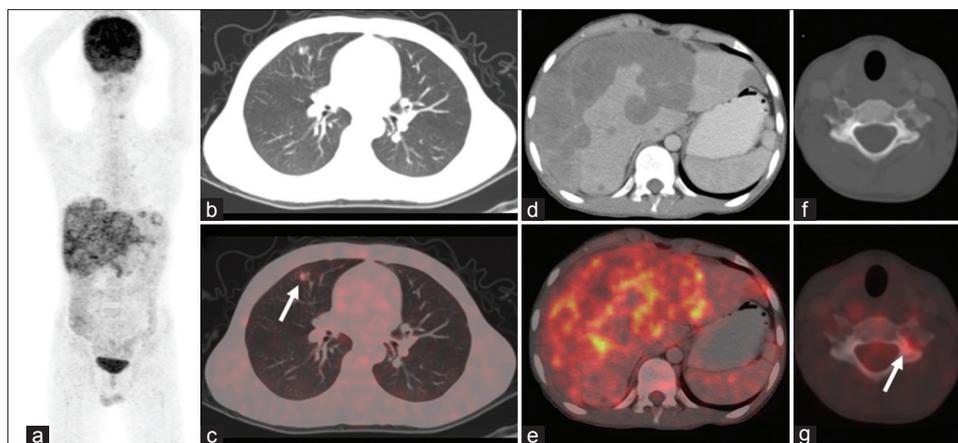


Figure 1: 18F-fluorodeoxyglucose positron emission tomography/computed tomography maximum intensity projection (a), transaxial computed tomography (b), and fused positron emission tomography/computed tomography (c) images showing low-grade fluorodeoxyglucose avid multiple parenchymal nodules in the lung fields (arrow; c), peripheral fluorodeoxyglucose avid multiple hypo-enhancing lesions in both lobes of the enlarged liver (d and e), moderate ascites and mild fluorodeoxyglucose avidity in the pedicle of C7 vertebra (f and g; arrow) suggesting marrow involvement

DISCUSSION

EHE is a rare vascular tumor, originating from endothelial cells, with an estimated incidence of $<0.1/100,000$ and an unpredictable malignant potential.^[1,2] The peak incidence of the disease is seen in 20–40 years of age; however, the age distribution extends on either side of this limit, as in our case. Common presentations of EHE include involvement of the liver only (21%), liver and lung (18%), lung only (12%), and bone only (14%).^[3] Commonly seen in the extremities, liver, lungs, and bone are the relatively rare sites^[4] all three of which were involved in the present case. Cases of EHE have been reported in older patients with liver, lungs, mediastinum, and skeletal involvement.^[5–9] The primary hepatic EHE is often treated by resection/liver transplant, with overall survival at 1, 3, and 5 years reported as 100%, 87.5%, and 71.3%, respectively, posttransplant.^[10] However, patients with metastatic disease are not potential candidates for transplant and need alternate management options such as chemotherapy.^[11]

Imaging techniques such as MRI are helpful for soft-tissue delineation of the primary lesion but are unable to comment on the disease extent as a whole. ¹⁸F-FDG PET/CT is useful in the evaluation of these patients, providing information about the metabolic status of the tumor as well as the identification of distant site involvement. In the present case, the detection of metastatic disease on ¹⁸F-FDG PET/CT changed the management plan from liver transplantation to chemotherapy.

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Conflicts of interest

There are no conflicts of interest.

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