^{18}F-FDG PET/CT imaging of multiple intrahepatic Epstein-Barr virus-associated smooth muscle tumors in a pediatric patient after heart transplantation

Abstract

Epstein-Barr virus-associated smooth muscle tumor (EBV-SMT) is an exceedingly rare neoplastic disease with a predisposition in immune-compromised individuals, especially in patients with prior transplantation, human immunodeficiency virus infection, or congenital immunodeficiency. Here, we present imaging findings of EBV-SMT in multiphasic contrast-enhanced computed tomography (CT) and fluorine-18-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography (PET)/CT in a two-and-a-half-year-old boy with prior heart transplantation.

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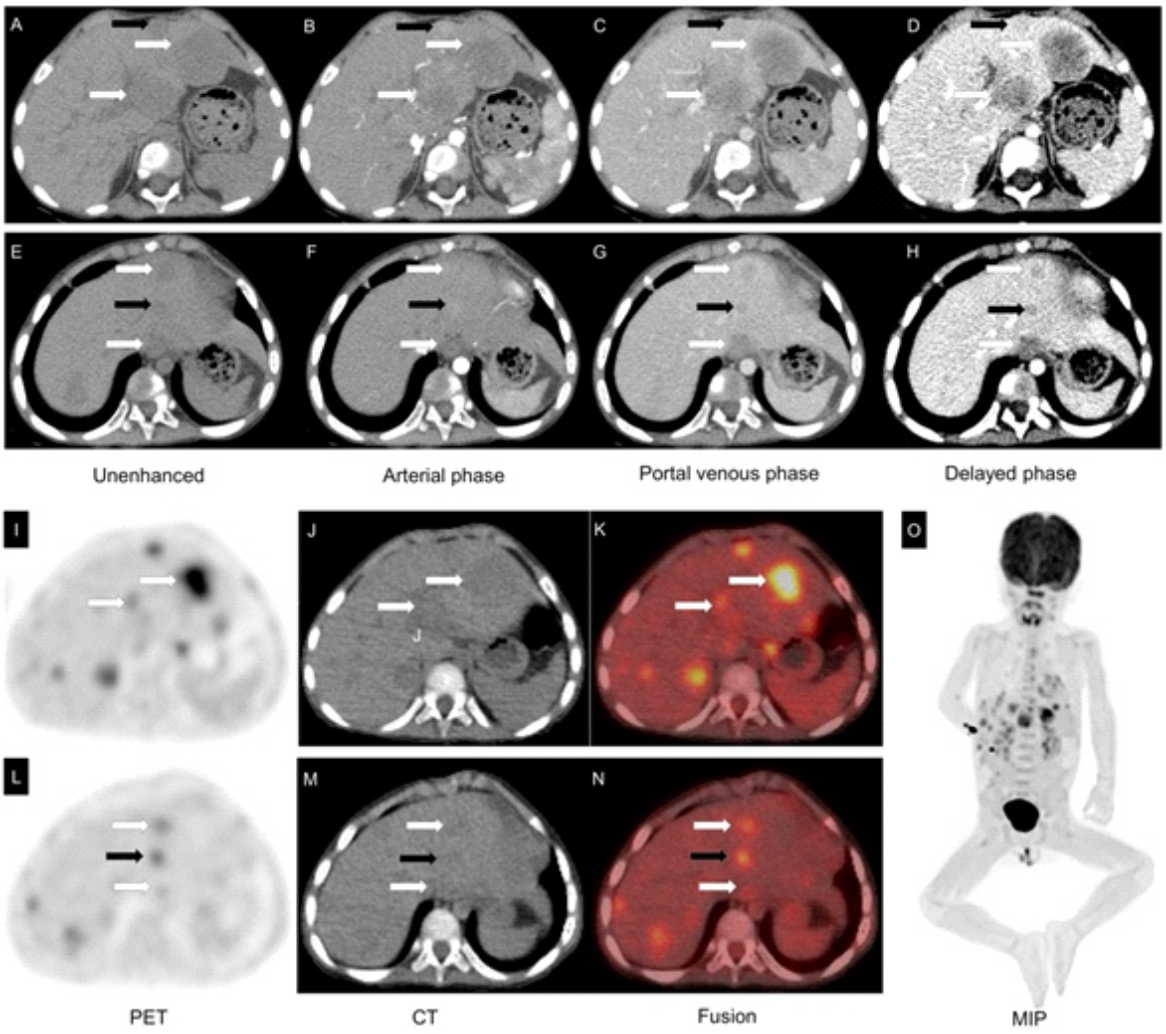

Figure 1. A two-and-a-half-year-old boy was admitted to the hospital for persisting cough and sputum over 10 days. He had a prior heart transplantation due to dilated cardiomyopathy at one year old. The laboratory test at his administration revealed a high amplification of Epstein-Barr virus (EBV) DNA (2100 copies/mL, reference<500 copies/mL), elevated serum lactate dehydrogenase level (340U/L, reference <250U/L), and otherwise unremarkable findings. Physical examination identified hepatic enlargement with several nodules. Subsequent multiphase contrast-enhanced computed tomography (CT) scan demonstrated numerous intrahepatic hypodense lesions with faint-to-mild homogenous (black arrows) and rim-like (white arrows) arterial enhancement and wash-out in portal-venous and delayed phases (A-H). Whole-body fluorine-18-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography (PET)/CT was performed to evaluate the involved extent of the disease, which showed innumerable hypermetabolic foci within the liver with maximum standardized uptake value (SUVmax) up to 8.2 (I-O), and bilateral lung inflammation with mild ^{18}F-FDG uptake (images not showed). Differential diagnoses of infection, primary and metastatic liver malignancies, and post-transplant lymphoproliferative disorder (PTLD) were made.

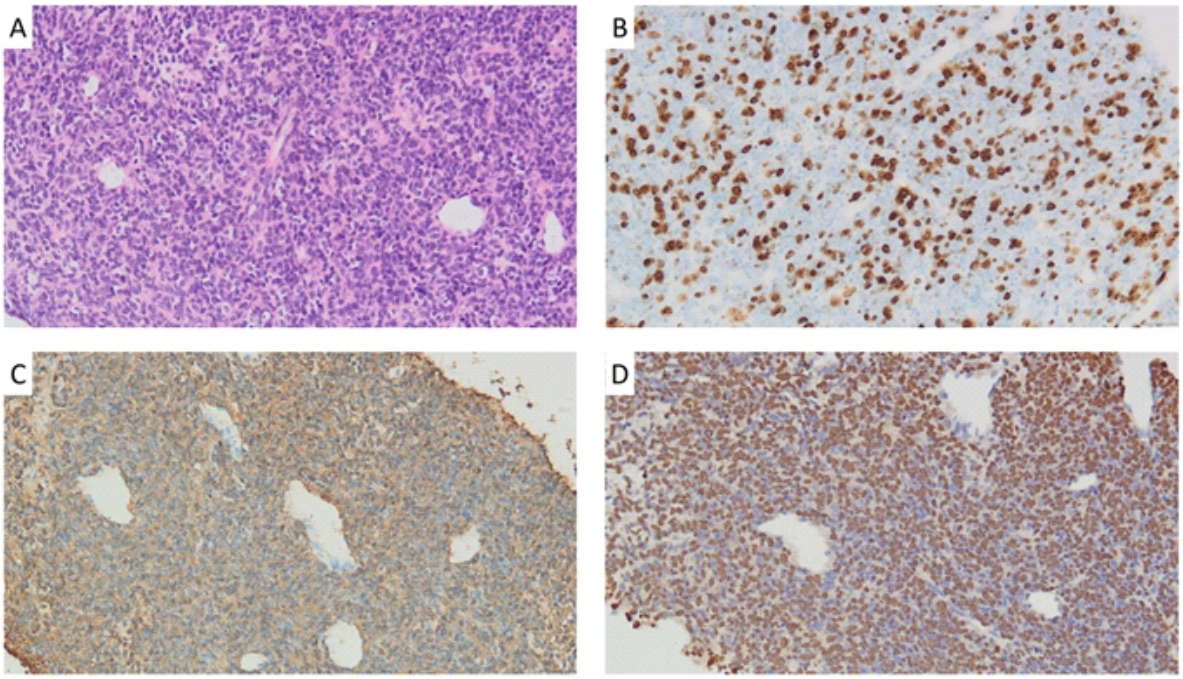

Figure 2. The patient underwent a CT-guided fine-needle biopsy at the hepatic S2 mass. The histological study revealed a mixture of spindle-shaped to ovoid cells on hematoxylin-eosin (HE) staining (A), which were positive for Ki-67 (B) and smooth muscle actin (SMA) (C) on immunohistochemical staining, and EBV in situ hybridization (D), confirming the diagnosis of EBV-associated smooth muscle tumor (EBV-SMT).

Epstein-Barr virus has been identified as a transforming virus linked to many diseases, including lymphoma, nasopharyngeal carcinoma, gastric carcinoma, and PTLD, but rarely EBV-SMT [1-4]. Epstein-Barr virus-SMT is mainly diagnosed in patients with prior transplantation or infection with human immunodeficiency virus and is usually multifocal, involving sites that are uncommon for conventional SMT [5-8]. It lacks characteristic clinical and imaging manifestations and thus requires histopathology for confirmation [8]. Histologically, it appears as a spectrum from well-differentiated spindle-shaped smooth muscle cells to ovoid cells with incomplete smooth muscle cell differentiation and can be diagnosed using the following criteria: 1) positive for at least one smooth muscle marker including smooth muscle actin (SMA), caldesmon and desmin, and 2) positive for EBV by in situ hybridization [5, 9, 10].

Due to the extensive disease spread within the liver, the patient was a non-surgical candidate. He received only conservative management, including a reduction in immunosuppression, antiviral therapy using Acyclovir, and oral sirolimus as a mammalian target of rapamycin (mTOR) inhibitor. Unfortunately, he developed disease progression 3 months later, as shown by the follow-up abdominal MRI scan, and thus is currently under palliative care.

The authors declare that they have no conflicts of interest.

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