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^{18}F-FDG PET/CT Imaging of a Grade 3 Lymphomatoid Granulomatosis in an Immunocompromised Pediatric Patient

Primer İmmün Yetmezliği Olan Evre 3 Lenfomatoid Granülomatozis Tanılı Pediatrik Bir Hastanın ^{18}F-FDG PET/BT Görüntülemesi

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Abstract

Lymphomatoid granulomatosis is a rare extranodal Epstein-Barr virus-driven B-cell lymphoproliferative disease, involving predominantly lung, less often skin, kidney, and central nervous system. Here, we present a pediatric case with primary immunodeficiency, diagnosed with pathologically proven pulmonary grade-III lymphomatoid granulomatosis. ^{18}F-fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) imaging demonstrated ^{18}F-FDG avid pulmonary masses with central air-bronchograms and cavitations. Although the definitive diagnosis depends on biopsy, ^{18}F-FDG PET/CT serves as a complementary imaging tool to evaluate the extent of the disease and response to treatment.

Keywords: Lymphomatoid granulomatosis, pulmonary involvement, ^{18}F-FDG PET/CT

Öz

Lenfomatoid granülomatozis, çoğunlukla akciğer, daha az sıklıkla deri, böbrek ve merkezi sinir sistemi tutulumu ile giden, nadir görülen ekstranodal Epstein-Barr virüsü ilişkili B-hücreli lenfoproliferatif bir hastalıktır. Burada, primer immün yetmezlikli, biyopsi ile pulmoner grade-3 lenfomatoid granülomatozis tanısı almış pediatrik bir olgu sunulmaktadır. ^{18}F-florodeoksiglukoz (FDG) PET/BT görüntülemesi, santral kesimde hava bronkogramları ve kavitasyonlar içeren ve ^{18}F-FDG tutulumu gösteren pulmoner kitleler göstermiştir. Kesin tanı biyopsiye bağlı olsa da ^{18}F-FDG PET/BT tetkiki hastalık yaygınlığının ve tedaviye yanıtın değerlendirilmesinde yardımcı bir görüntüleme yöntemidir.

Anahtar kelimeler: Lenfomatoid granülomatozis, akciğer tutulumu, ^{18}F-FDG PET/BT

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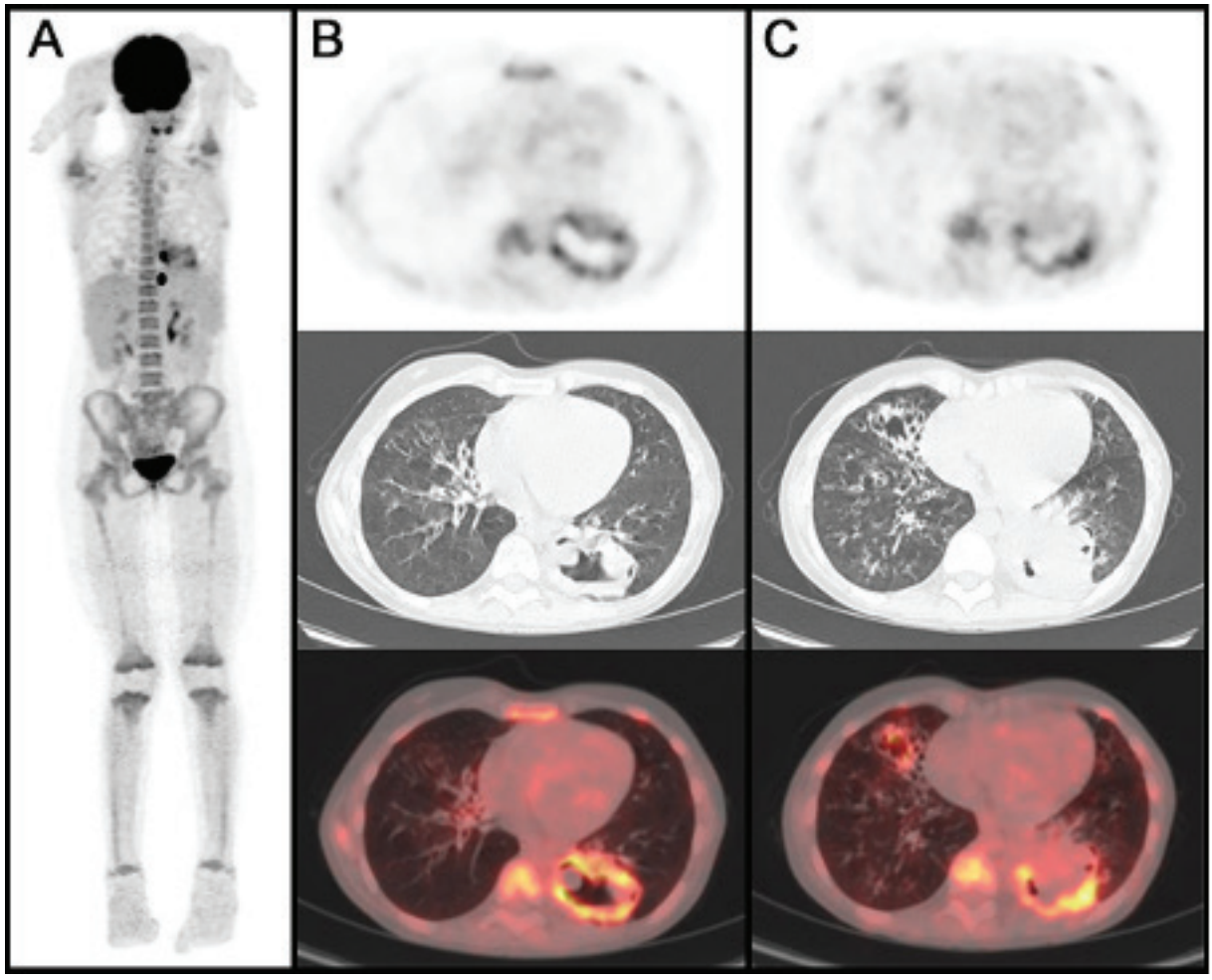

Figure 1. A 13-year-old girl, who has major histocompatibility complex class I deficiency, was presented with persistent cough, and computed tomography (CT) imaging revealed nodular lesions in both lungs. Biopsy was directed to the lesion in the left lower lobe of the lung, and histopathologic examination was consistent with grade 3 lymphomatoid granulomatosis (LG). A combination of chemoimmunotherapy was initiated and she was referred to ^{18}F-fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) to evaluate the extent of the disease. ^{18}F-FDG PET/CT maximum intensity projection image showed ^{18}F-FDG avid pulmonary masses (A). Axial PET, CT, and fused PET/CT images demonstrated peripheral intense ^{18}F-FDG uptake with a maximum standardized uptake value of 19.2 in the pulmonary nodules/mass that have central cavitations, internal air-bronchograms, and subpleural distribution, in the left lower (B) and the mid lobe of the lung (C). No extrapulmonary lesions were identified.

LG is a rare Epstein-Barr virus (EBV)-associated B-cell lymphoproliferative disorder, that involves extranodal sites, such as the skin, central nervous system, liver, and kidneys, with the lung almost always being affected (1). LG is classified into three histological grades depending on the number and density of atypical large EBV-positive B-cells, angio-invasive/angio-destructive reactive T-cell infiltration, and necrosis, which is essential in selecting appropriate treatment strategies (2). Corticosteroids and/or single-agent chemotherapy are used in low-grade (grade 1 and grade 2) LG, whereas high-grade (grade 3) LG, has an inferior outcome with a higher incidence of multisystem involvement and is best managed by chemoimmunotherapy (3). Most of the patients do not have underlying immunodeficiency, while immunosuppressed individuals have increased risk, as in our patient. LG typically presents in middle-aged adults, and reports of this disease in children are limited in the literature (4).

Pulmonary LG, which predominantly affects the mid to lower lung, presents with nodules and masses mostly in the peribronchovascular and subpleural regions, which corresponding to lymphatic dissemination, and central low attenuation, cavitation, and peripheral enhancement of the nodules/masses are related to the angio-invasive and angio-destructive nature of the disease (5). CT manifestations may mimic infection, bronchioloalveolar lung cancer, pulmonary lymphoma, sarcoidosis, or Wegener's granulomatosis, and LG should be considered a differential diagnosis while evaluating ^{18}F-FDG PET/CT images of pediatric patients with respiratory and/or B symptoms.

^{18}F-FDG PET/CT findings in patients with LG, involving the lung or central nervous system, were limited to case reports in the literature (6,7,8,9,10). Like other ^{18}F-FDG-avid high-grade lymphomas, ^{18}F-FDG PET/CT is reported to be an effective imaging tool for staging and monitoring response to treatment in patients with LG (9). With the ability to present both morphologic and functional information, whole-body ^{18}F-FDG PET/CT scan can assist in identifying the involved sites, guide high-yield biopsy, and help the management of the disease.

Ethics

Informed Consent: We have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: S.K., F.Ş., S.Ö., T.Ö., Concept: S.K., F.Ş., Design: S.K., F.Ş., Data Collection or Processing: S.K., F.Ş., Analysis or Interpretation: S.K., F.Ş., Literature Search: S.K., F.Ş., Writing: S.K., F.Ş.

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