Nucl Med Mol Imaging
Vol. 43, No. 3, Jun 2009

Case Report

피하 지방층염양 T-세포 림프종의 F-18 FDG PET/CT 소견

영남대학교 의과대학 핵의학교실[1], 병리학교실[2], 내과학교실[3]
공은정[1] · 조인호[1] · 천경아[1] · 배영경[2] · 최준혁[2] · 현명수[3]

F-18 FDG PET/CT Findings of Subcutaneous Panniculitis - Like T- Cell Lymphoma : A Case Report

Eun-Jung Kong, M.D.[1], Ihn-Ho Cho, M.D.[1], Kyung-Ah Chun, M.D.[1], Yeung-Kyung Bae, M.D.[2], Joon-Hyuk Choi, M.D.[2], and Myung-Soo Hyun, M.D.[3]
Departments of [1]Nuclear Medicine, [2]Pathology and [3]Internal Medicine, Yeungnam University College of Medicine, Daegu, Korea

F-18 FDG PET is a metabolic imaging modality that is efficacious in staging and assessment of treatment response for variety of lymphomas. We report usefulness of F-18 FDG PET/CT in evaluating severity of the disease and response to therapy in a patient with subcutaneous panniculitis- like T-cell lymphoma (SPTCL). Here we describe a case of SPTCL in 24-year-old man who had wide spread firm and tender nodular lesions with increased F-18 FDG uptake. After chemotherapy follow up F-18 FDG PET/CT image shows disseminated malignancy and then the patient died with hemophagocytic syndrome. This report suggests that F-18 FDG PET/CT may be useful in determining disease activity at the time of initial diagnosis, after treatment, and evaluating a suspected outcome of SPTCL. (Nucl Med Mol Imaging 2009;43(3):240-244)

Key Words: PET/CT, subcutaneous panniculitis-like T-cell lymphoma, hemophagocytic syndrome

Introduction

Subcutaneous panniculitis- like T-cell lymphoma (SPTCL) is a rare T-cell neoplasm characterized by a lipotrophic lymphohistiocytic infiltration commonly presenting as subcutaneous nodules resembling panniculitis.[1)] Two clinical courses are reportedly observed: a prolonged course of recurrent panniculitis or a rapid clinical deterioration secondary to the hemophagocytic syndrome (HPS). Histiocytic proliferation and activation within the reticuloendothelial system are associated with phagocytosis of blood elements, fever, hepatosplenomegaly, and coagulopathy characterize the HPS. Therefore, detection and accurate staging at the time of diagnosis is of critical importance for planning treatment and determining prognosis.[2-4)]

The diagnosis of primary cutaneous lymphoma is currently based on clinical and histological findings and/or relatively invasive procedures such as bone marrow and lymph node biopsies.[1)] Although CT is a noninvasive imaging modality that is widely used for staging in patients with lymphoma, but provide a little information on cutaneous tumor burden.[2)] We present the usefulness of F-18 FDG PET/CT scanning for staging patients with SPTCL, choosing treatment strategies, objectively evaluating therapeutic response and predicting of prognosis.

• **Received**: 2009. 6. 20. • **Revised**: 2009. 6. 24.
• **Accepted**: 2009. 6. 25.
• Address for reprints: Ihn-Ho Cho, M.D., Department of Nuclear Medicine, Yeungnam University Hospital, 317-1 Daemyung 5-dong, Nam-gu, Daegu, 705-717, Korea
Tel: 82-53-620-3076, Fax: 82-53-651-7415
E-mail: nuclear126@ynu.ac.kr

Case Report

A 24-year-old man presented with 2 weeks history of multiple nodules on his trunk and extremities along with malaise for 4 weeks. He also had 3 kilograms weight loss for 4 weeks. On physical examination, there were multiple

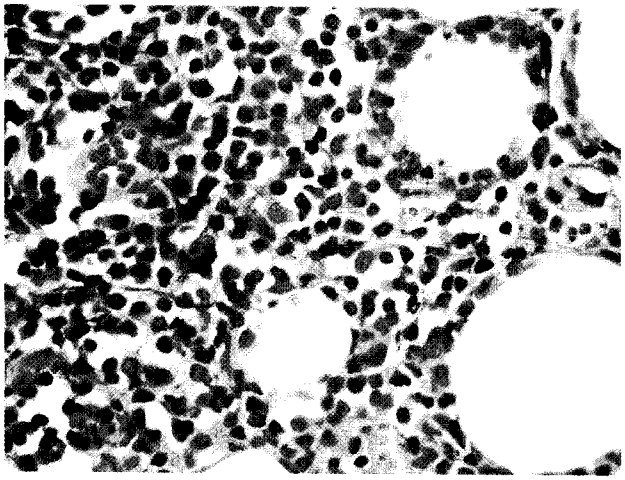

Figure 1. The lymphoma infiltrates the subcutaneous adipose tissue and consists of small to medium-sized pleomorphic lymphocytes. Hematoxylin and eosin stain; original magnification ×400.

firm and tender nodules that spread from abdomen to both lower extremities and his shoulder.

The laboratory evaluation showed a WBC of 2,950 /μL, with 17% band, 11.5 g/dL Hgb, 149 K/μL platelet, and 1580 U/L LDH. Biopsy of abdominal wall mass showed a lymphomatous infiltrate involving predominantly the subcutaneous lobules in a lobular panniculitis-like pattern. The tumor cell surrounded individual fat cell with focal fat necrosis and karyorrhexis (Fig. 1). Immunohistochemical evaluation showed CD3 and CD8 positivity and CD20, CD4 and CD56 negativity. Neither a T cell receptor (TCR) gene rearrangements study nor bone marrow examination was done in this case. Although definite proof was lacking, our case most probably presented an SPTCL α/β T-cell phenotype given CD4-, CD8+ and CD56- immunophenotyping. A diagnosis of SPTCL was made.

For systemic survey, whole body PET/CT was performed. Images were obtained 1 hour after intravenous injection of 470 MBq of F-18 FDG using a scanner (Discovery, ST, GE). The patient fasted for 6 hours; the serum glucose level, measured before examination, was found to be less than 140 mg/dl. The F-18 FDG PET/CT image revealed multiple foci of intense FDG uptake superficially within the neck, both arm, chest, back, abdomen and both lower extremity, with extracutaneous

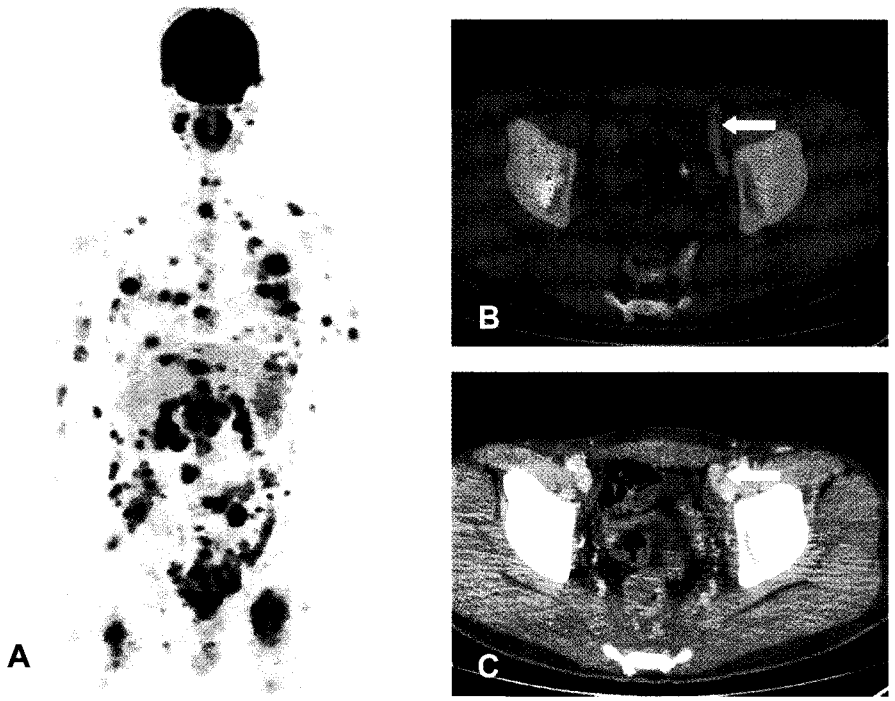

Figure 2. F-18 FDG PET/CT images of 24-year-old man diagnosed as an subcutaneous panniculitis-like T-cell lymphoma. (A) MIP image shows multifocal increased uptake in subcutaneous tissue and extracutaneous lesions. Selected transverse images of (B) PET/CT and (C) CT images show high F-18 FDG uptake lesions of intrapelvic lymphadenopathy (arrows) and extracutaneous lesions.

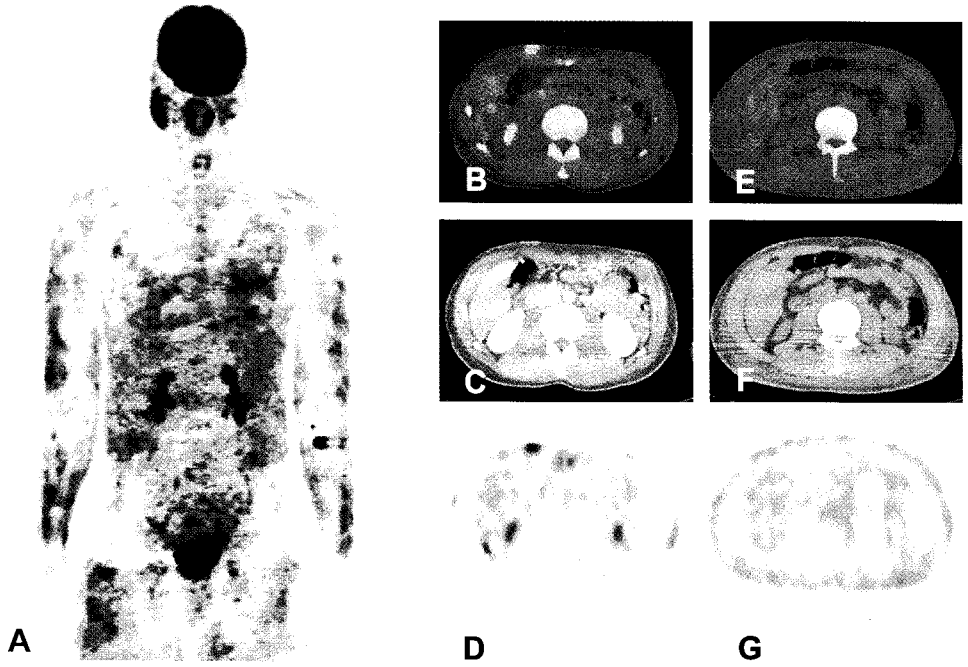

Figure 3. The patient underwent PET/CT again after the completion of the 3 cycle chemotherapy. (B) MIP image demonstrates more increased F-18 FDG uptake with expansion of the affected lesion. Chemotherapeutic response was assessed by a follow up F-18 FDG PET/CT. (B,C,D): at the time of diagnosis and (E,F,G) after the 3 cycle chemotherapy.

disease (i.e., lymphadenopathy in pelvic cavity and abdominal cavity) (Fig. 2). The maximum standardized uptake value (maxSUV) was 17.3.

The patient was treated on CEOP-E chemotherapy consisting of cyclophosphamide 750 mg/m^2, epirubicin 20 mg/m^2, vincristine 0.5 mg, predinisone 100 mg and etoposide 80 mg/m^2 for a total of three courses. After chemotherapy, repeated F-18 FDG PET/CT scan revealed more advanced disease. The maxSUV was 19.7 (Fig. 3). The chemotherapy regimen was changed to DHAP (dexamethasone, cisplatin, cytarabine) but, the patient was expired with HPS after 1 month from follow up F-18 FDG PET/CT.

DISCUSSION

SPTCL is a rare cytotoxic T-cell lymphoma of the skin and accounts for less than 1% of all non-Hodgkin's lymphoma cases.[1] Clinically, SPTCL is predominantly disease that affects young adults. Lymphoma lesions consist of solitary or multiple subcutaneous nodules and/or erythematous, infiltrated plaques or ulcerated skin tumors, mostly located on the extremities, trunk, and face. Clinical symptoms include malaise, fatigue, myalgia and weight loss; manifestations of systemic involvement are fever, hepatosplenomegaly, mucosal ulcers and sometimes serosal effusions. Some patients may develop HPS, a complication often precipitating the clinical course and it carries an 81% mortality.[4-6]

Recent studies have shown clinical, histological and immunophenotypical differences between cases of SPTCL with an α/β T-cell phenotype and those with a γ/δ T-cell phenotype. Whereas SPTCL with an α/β T-cell phenotype are indolent clinical behavior in many patients, SPTCL with an γ/δ T-cell phenotype overlap with other type of γ/δ- positive T/NK -cell lymphoma and generally run a very aggressive clinical course.[3,4] In the WHO-EORTC classification, the term SPTCL is only used for cases with an α/β T-cell phenotype, whereas case with a γ/δ T-cell phenotype are included in a category of cutaneous γ/δ T-cell lymphoma.[7-8] In our case, the patient most probably presents an SPTCL α/β T-cell phenotype given CD4-, CD8+ and CD56- immunophenotyping, but he presented with multiple focal

areas of intense FDG uptake with hypermetabolic lymphadenopathy in pelvis and abdomen on F-18 FDG PET/CT which suggest of aggressive disease.[8] Our patient showed no response to chemotherapy and died with systemic HPS.

F-18 FDG PET reveals enhanced rates of glucose metabolism by malignant cells, allowing differentiation between neoplastic and benign processes. F-18 FDG PET has been shown to be highly sensitive and specific for imaging nodal and extranodal lesions in patients with lymphomas when compared with other conventional imaging modalities such as CT and gallium scintigraphy. In addition, PET can also be used for selecting a biopsy site when the location is considered difficult to find.[9-11] In our patients, this technique confirmed the presence of subcutaneous disease at sites of clinical suspicion and also show hypermetabolic lymph nodes in pelvic cavity and hypermetabolic lesion in abdomen. The presence of extracutaneous disease is important in planning the treatment and predicting prognosis in both initial staging and restaging following therapy of cutaneous lymphoma. F-18 FDG PET is known more accurate than CT in detecting distant metastasis. F-18 FDG PET showed an overall accuracy of 97% for evaluating extracutaneous disease while CT had an overall accuracy of 82%.[12,13] CT has a high sensitivity and specificity in pretreatment staging; however, it has poor specificity in detecting residual or recurrent disease. As the detection of disease with F-18 FDG PET is based on metabolism rather than on physical size, PET is able to detect post-therapy changes earlier than conventional anatomical imaging with CT.[12] Also, more recently, the MR imaging findings were nonspecific for soft tissue tumors and soft tissue inflammation.[14]

Only a few studies have been published in the literature on F-18 FDG PET in the evaluation of cutaneous lymphoma. Valencak et al.[15] reported that none of the nine patients in stage Ia had a positive PET scan, whereas all four patients with stage IV disease had a positive F-18 FDG PET scan. They concluded that F-18 FDG PET has a limitation for initial early staging, but might add valuable information for patients with advanced disease, and facilitate planning of therapy and follow-up. Shapiro et al.[16] reported two cases of cutaneous lymphoma, one cutaneous T cell lymphoma and one B cell lymphoma. F-18 FDG PET was useful in determining disease activity both at the time of initial diagnosis and after treatment, and in evaluating a suspected recurrence. Moreover, recently published two papers reported that F-18 FDG PET study was valuable in monitoring treatment response and detecting extrasubcutaneous lesion in SPTCL.[17,18]

The SUV calculation provided by F-18 FDG PET/CT imaging may aid clinicians in quantifying FDG uptake. Our patient showed high FDG uptake (maxSUV=17.3) in the involved lesion with more increased FDG uptake (maxSUV=19.7) after chemotherapy. This, in addition to the patient's significant systemic symptoms and disease refractory to multitherapy, may suggest an aggressive course.

Although there is need for further study, the findings in our case also suggest that F-18 FDG PET/CT images provide valuable information in detecting lesions, tumor aggressiveness, and consequently influence treatment strategy in SPTCL. F-18 FDG PET/CT scanning may also assist clinicians in evaluating therapeutic response and detecting early recurrence in patients with cutaneous lymphoma.

References

1. Gonzalez CL, Medeiros LJ, Braziel RM, Jaffe ES. T-cell lymphoma involving subcutaneous tissue: a clinicopathologic entity commonly associated with hemophagocytic syndrome. ***Am J Surg Pathol*** 1991;15:17-27.
2. Salhany KE, Macon WR, Choi JK, Elenitsas R, Lessin SR, Felgar RE, et al. Subcutaneous panniculitis-like T-cell lymphoma: clinicopathologic, immunophenotypic, and genotypic analysis of alpha/beta and gamma/delta subtypes. ***Am J Surg Pathol*** 1998;22:881-93.
3. Siegel RS, Pandolfino T, Guitart J, Rosen S, Kuzel TM. Primary cutaneous T-cell lymphoma: review and current concepts. ***J Clin Oncol*** 2000;18:2908-25.
4. Go RS, Wester SM. Immunophenotypic and molecular feature, clinical outcomes, treatments, and prognostic factors associated with subcutaneous panniculitis-like T-cell lymphoma. A systematic analysis of 156 patients reported in the literature. ***Cancer*** 2004;101:1404-13.
5. Willemze R, Meijer CJ. Classificatioin of cutaneous T-cell lymphoma: from Alibert to WHO-EORTC. ***J Cutatan Pathol*** 2006;33(suppl.1):18-26.
6. Marzano AV, Berti E, Paulli M, Caputo R. Cytophagocytic

histiocytic panniculitis and subcutaneous panniculitis-like T-cell lymphoma. ***Arch Dermatol*** 2000;136:889-96.
7. Willemze R, Jaffe ES, Burg G, Cerroni L, Berti E, Swerdlow SH, et al. WHO-EORTC classficaion for cutaneous lymphomas. ***Blood*** 2005;105:3768-85.
8. Willemze R, Jansen PM, Cerroni L, Berti E, Santucci M, Assaf C, et al. Subcutaneous panniculitis-like T-cell lymphoma: definition, classification, and prognostic factors: an EORTC cutaneous lymphoma group study of 83 cases. ***Blood*** 2008;111: 838-45.
9. Moog F, Bangerter M, Diederichs CG, Guhlmann A, Kotzerke J, Merkle E, et al. Lymphoma: role of whole-body 2-deoxy-2-[F-18]fluoro-D-glucose (FDG) PET in nodal staging. ***Radiology*** 1997;203:795-800.
10. Moog F, Bangerter M, Diederichs CG, Guhlmann A, Merkle E, Frickhofen N, et al. Extranodal malignant lymphoma: detection with FDG PET vs. CT. ***Radiology*** 1998;206:475-81.
11. Kostakoglu L, Goldsmith SJ. Positron emission tomography in lymphoma: comparison with computed tomography and gallium-67 single photon emission computed tomography. ***Clin Lymphoma*** 2000;1:67-74.
12. Guay C, Lepine M, Verreault J, Benard F. Prognostic value of PET using ^{18}F-FDG in Hodgkin's disease for post-treatment evaluation. ***J Nucl Med*** 2003;44:1225-31.
13. Kumar R, Xiu Y, Zhuang HM, Alavi A. ^{18}F-fluorodeoxyglucose-positron emission tomography in evaluation of primary cutaneous lymphoma. ***Br J Dermatol*** 2006;155:357-63.
14. Kim YH, Kim HS, Kim SY, Hwang YJ, Seo JW, Lee JY, et al. MR Findings of subcutaneous panniculitis-like T-cell lymphoma: a case report. ***J Korean Radiol Soc*** 2007;57:479-82.
15. Valencak J, Becherer A, Der-Petrossian M, Trautinger F, Raderer M, Hoffmann M. Positron emission tomography with [^{18}F] 2-fluoro-D-2-deoxyglucose in primary cutaneous T-cell lymphomas. ***Haematologica*** 2004;89:115-6.
16. Shapiro M, Yun M, Junkins-Hopkins JM, Vittorio CC, Schulman N, Saidman BH, et al. Assessment of tumor burden and treatment response by ^{18}F-fluorodeoxyglucose injection and positron emission tomography in patients with cutaneous T- and B-cell lymphomas. ***J Am Acad Dermatol*** 2002;47:623-8.
17. Rodriguez VR, Joshi A, Peng F, Rabah RM, Stockmann PT, Savaşan S. Positron emission tomography in subcutaneous panniculitis-like T-cell lymphoma. ***Pediatr Blood Cancer*** 2009;52: 406-8.
18. Ravizzini G, Meirelles GS, Horwitz SM, Grewal RK. F-18 FDG uptake in subcutaneous panniculitis-like T-cell lymphoma. ***Clin Nucl Med*** 2008;33:903-5.