Clinical note

FDG PET/CT appearance of multicentric Castleman’s disease mimicking lymphoma

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INTRODUCTION

We report a case of a multicentric form of Castleman’s disease with multiple lymph nodes showing intense FDG uptake on whole body scan mimicking non-Hodgkin’s lymphoma. In this report, the patient had multiple cervical, mediastinal, hilar, retroperitoneal and abnormal lymph nodes in the groin. 18F-fluorodeoxyglucose positron emission tomography/computed tomography was performed before tissue sampling. 18F-FDG/PET demonstrated multiple areas of increased uptake in cervical, mediastinal, hilar, retroperitoneal and groin lymph nodes, suggesting a generalized disease of the lymphatic system including non-Hodgkin’s lymphoma. The final diagnosis is based on the histopathological findings of the material obtained from the cervical lymphadenectomy. The histological diagnosis was multicentric plasma cell variant of Castleman’s disease. 18F fluorodeoxyglucose positron emission tomography/computed tomography scan helped to identify the lymph nodes involved throughout the whole body, but did not help to differentiate non-Hodgkin’s lymphoma. The clinical conclusions and PET/CT findings are described in this report.

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Distribución de la FDG PET/TAC en la enfermedad de Castleman simulando Linfoma

RESUMEN

Se presenta un caso de enfermedad de Castleman multicéntrica con múltiples ganglios linfáticos que muestran una intensa captación de FDG, imitando el linfoma no-Hodgkin. El paciente presentó múltiples nódulos linfáticos cervicales, mediastínicos, hiliares, retroperitoneales e inguinales, así como nódulos linfáticos anormales en la ingle.

La tomografía por emisión de positrones 18F-fluorodeoxyglucosa/tomografía computarizada se realizó antes de obtener las muestras de tejido. La 18F FDG-PET mostró múltiples áreas de aumento de captación en ganglios linfáticos cervicales, mediastínicos, hiliares, retroperitoneales e inguinales, sugiriendo una enfermedad generalizada del sistema linfático, como el linfoma no-Hodgkin. El diagnóstico final se basó en los hallazgos histopatológicos obtenidos de la linfadenectomía cervical. El diagnóstico histológico fue el de enfermedad de Castleman multicéntrica variante de células plasmáticas. La 18F fluorodeoxyglucosa tomografía por emisión de positrones/tomografía computada ayudó a identificar la extensión corporal de la afectación ganglionar, pero no permitió un diagnóstico diferencial conel linfoma no Hodgkin. Se describen las conclusiones clínicas y los hallazgos de la PET/TAC.

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Introduction

Castleman’s disease (CD) or angiofollicular lymph node hyperplasia is a rare benign lymphoproliferative disorder, which was first described in 1954.1 The etiology of this disorder is unknown yet. Histologically, CD is classified into three subtypes: the hyaline vascular type, the plasma cell type and the intermediate (mixed) cell type.2

One of the critical issues in diagnosis is to differentiate between CD and Hodgkin’s and non-Hodgkin’s lymphoma. This is most challenging in multicentric plasma cell form of CD as involvement of multiple lymph nodes mimics lymphomas more than any other variants and forms of CD.

It is therefore important to document the distribution of involved lymph nodes throughout the body. Whole body 18F-FDG PET/CT imaging is thus the right modality to investigate the distribution of the diseased lymph nodes. We report a patient with multicentric form of plasma cell variant CD showing a similar uptake pattern of non-Hodgkin’s lymphoma on 18F-FDG PET/CT images, which caused a challenge in differential diagnosis.

Case report

A 73-year-old woman with anorexia, weight loss (7 kg in 3 months), fever and night sweats was admitted to the Hematology Department. Physical examination revealed slightly enlarged bilateral cervical lymph nodes. The initial laboratory findings showed hypochromic anemia with a hemoglobin level of 9.8 g/dL and 24.7% hematocrit, elevated erythrocyte sedimentation rate (69 mm/h), and serum C-reactive protein level of 18.32 mg/L.
Chest X-ray showed multiple bilateral enlarged hilar and mediastinal lymph nodes, atelectasis in right lobe and pleural effusion in the right hemithorax. HIV antibody results were negative. Peripheral blood smear indicated leukocytosis with atypical lymphocytes.

The differential diagnosis included lymphoma, nonspecific reactive lymphoid hyperplasia, toxoplasmosis, tuberculosis, Epstein-Barr virus infection and Castleman’s disease. Bone marrow biopsy was planned. The patient was referred to Nuclear Medicine Department for a PET/CT scan to document the distribution of the enlarged lymph nodes, and if the enlarged nodes were metabolically active. After 6 h fasting with a serum glucose 95 mg/dl, 447 MBq of $^{18}$F-FDG was injected intravenously. After 1 h of waiting for distribution and uptake of FDG in a semireclined relaxed chair in a quite booth, the patient was imaged in an integrated full-ring PET/CT (Discovery 600, General Electric Medical Systems, Waukesha, WI).

The patient was not given any contrast agent for CT scan which was performed for attenuation correction and defining anatomical landmarks on PET images. Increased $^{18}$F-FDG accumulation was observed in bilateral cervical, axillary, multiple bilateral mediastinal, retroperitoneal, groin lymph nodes and crescent-shaped image in the right thorax related to pleural effusion on both PET images (Fig. 1). The areas of increased FDG uptake were well correlated with the enlarged lymph nodes detected on CT images. The maximum standardized uptake values (SUVmax) for the cervical, axillary and mediastinal, hilar, retroperitoneal, groin lymph nodes were 9.8, 12.3 and 14.7, 10.1, 12.2, respectively.

Bone marrow biopsy was performed, which revealed lymphoid aggregates, plasmacytosis but no malignant cells. Histopathological examination of an excised left cervical lymph node disclosed plasma cell variant of Castleman’s disease. With the multiple nodes detected on $^{18}$F-FDG PET/CT images, the diagnosis was therefore multicentric plasma cell variant of Castleman’s disease. The patient was informed about the expected benefits and complications of treatment options available and she preferred to be followed up without any treatment.

Fig. 1. Increased $^{18}$F-FDG accumulation was observed in bilateral cervical, axillary, multiple bilateral mediastinal, retroperitoneal, groin lymph nodes and crescent-shaped image in the right thorax related to pleural effusion on whole body $^{18}$F-FDG PET image (A), increased $^{18}$F-FDG accumulation was observed in bilateral cervical lymph nodes on transaxial $^{18}$F-FDG PET and PET/CT images (B).
Discussion

Castleman's disease was first described by Benjamin Castleman in 1956, when he reported a series of patients with solitary hyperplastic mediastinal lymph nodes with small germinal centers. CD has many synonyms, including angiofollicular mediastinal lymph node hyperplasia, angiomatous lymphoid hamartoma, lymph nodal hamartoma, follicular lymphphoreticulum and benign giant lymphoma. The etiology of CD is unknown, but there are several hypothetic mechanisms that have been proposed, such as chronic low-grade inflammation, a hamartomatous process, an immunodeficient state, and autoimmune disorders. CD has been linked to interleukin-6 (IL-6) overproduction. IL-6 is a major contributor to the systemic manifestations of CD.

Despite its high sensitivity $^{18}$F-FDG PET cannot differentiate between CD and other benign or malignant lymphoproliferative diseases. Active granulomatous and inflammatory disorders also show high accumulation of $^{18}$F-FDG mimicking malignant diseases including lymphoma. Similar to our case, it was previously shown that slight to moderate $^{18}$F-FDG uptake was reported in enlarged lymph nodes in patients with CD. Furthermore, low and intermediate grade lymphomas also show low grade $^{18}$F-FDG uptake on PET scans, while intense $^{18}$F-FDG uptake was seen in high grade lymphomas.

Although the enlarged lymph nodes in CD were benign, the high uptake suggests high metabolism in these nodes as seen in the lymphoma. The accumulation of $^{18}$F-FDG in our patient was high, but the appearance of diseased lymph nodes on $^{18}$F-FDG PET/CT images were not helpful to exclude Hodgkin's and non-Hodgkins lymphoma. It is therefore fair to conclude that whole body FDG PET/CT is effective in disclosing the involved lymph nodes and establishing new sites throughout the body, but it was not useful in differentiating CD from lymphoma.

The $^{18}$F-FDG PET/CT appearance of Castleman's disease is similar to those of malignant diseases and benign lymphoproliferative disorders, and thus can offer little help in differential diagnosis of enlarged lymph nodes of multicentric distribution. It may be useful in mapping the involved lymph nodes throughout the body in patients with Castleman's disease and in disclosing new sites of enlarged lymph nodes which are otherwise difficult to document.

References