CASE REPORT

Mediastinal Lymphadenopathy without Cervical Lymphadenopathy in a Case of Kikuchi-Fujimoto Disease

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Abstract

A 44-year-old man was referred to our hospital because of persistent high fever. Both CT and PET-CT demonstrated lymph node lesions limited to the mediastinal region without cervical lymphadenopathy. Histology of a mediastinal lymph node obtained by video-assisted thoroscopic excision confirmed the diagnosis of histiocytic necrotizing lymphadenitis. To our knowledge, this is the first report of Kikuchi-Fujimoto disease with isolated mediastinal lymphadenopathy. Although Kikuchi-Fujimoto disease is rare, we should consider this disease in patients with a high fever and no other symptoms.

Key words: Kikuchi-Fujimoto disease, mediastinal lymph node, malignant lymphoma PET-CT

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Introduction

Histiocytic necrotizing lymphadenitis, the so-called Kikuchi-Fujimoto disease, is a rare disease affecting mainly young women. The presenting symptoms are high fever and cervical adenopathy, with pathological findings of histiocytic necrotizing lymphadenitis (1, 2). Several authors have reported cases with lymphadenopathy in an atypical location, and such cases are difficult to differentiate from malignant lymphoma (3-6). A biopsy is necessary to arrive at a final histological diagnosis.

Case Report

A 44-year-old male was referred to the Department of Internal Medicine, Respiratory & Allergy Diseases Divisions, Toho University Sakura Medical Center with an over one week history of fever, malaise and no other symptoms. He had been treated in a local clinic with medications including antibiotics but failed to respond. He had no remarkable medical history and had not travelled abroad. His laboratory data showed a normal white cell count (4,050/μL) and slight elevation of lactase dehydrogenase (264; normal 120-240 IU/L) and CRP (0.55; normal under 0.30 mg/dL). Physical examination showed no abnormalities even in cervical and supraclavicular lymph nodes. Anti-nuclear antibody was negative. A chest radiograph as well as abdominal and cardiac ultrasonographic studies also revealed no abnormalities. Subsequently, enhanced CT demonstrated some enlarged mediastinal lymph nodes (Fig. 1). Whole body ¹⁸F-2-deoxy-D-glucose (FDG) positron emission tomography (PET) was performed, which showed high maximum standardized uptake values (maxSUV 13.5 to 21.1) in the mediastinal region (Fig. 2). Late-phase augmentation of SUV in PET suggested malignant lymphoma, although serum soluble IL-2 receptor concentration was almost within the normal range (509; normal 124-466 U/mL). Therefore bronchial fibroscopy was performed. Transcarinal aspiration cytology revealed solitary small atypical cells with irregularly shaped
nucleus and significant endoblast, which strongly suggested malignant lymphoma. However a transcarnal lymph node biopsy was not able to establish a definitive diagnosis of malignant lymphoma due to an inadequate specimen. After discussing with the patient the importance of an accurate diagnosis of his disease, we conducted video-assisted thoroscopic excision of #3 mediastinal lymph node. The histological findings of the specimen were compatible with histiocytic necrotizing lymphadenitis, better known as Kikuchi-Fujimoto disease (Fig. 3).

By the time he received the above-mentioned procedure, his fever had subsided. Four weeks later, soluble IL-2 receptor concentration had declined to 254 U/mL. A subsequent chest CT showed that the mediastinal lymphadenopathies had resolved spontaneously, even though the patient had not received medications such as NSAIDs, steroids, antiviral drugs and antibiotics (Fig. 4). This clinical course is consistent with the distinctive self-limiting nature of Kikuchi-Fujimoto disease, which is generally known as a benign disease.

Discussion

Almost four decades ago, two Japanese clinicians Kikuchi and Fujimoto simultaneously reported a clinicopathological entity seen mostly in young women with clinical symptoms of cervical adenopathy, fever and sore throat, and pathological finding of histiocytic necrotizing lymphadenitis, which
runs a benign course (1, 2). Laboratory investigations show no specific findings except mild leukocytopenia (7-9). In some recent cases manifesting lymphadenopathies in atypical sites, PET tended to be performed to differentiate benign from malignant lymphoma but most cases required surgical biopsy for a final diagnosis (3). In the present case, a middle-aged man presented with persistent fever but no sign of cervical lymphadenopathy, and PET detected enhanced uptake confined to the mediastinal region. All these findings are atypical of histiocytic necrotizing lymphadenitis.

Although many articles reported CT findings of systemic lymph node swelling in histiocytic necrotizing lymphadenitis, the existence of cervical adenopathy was a unique common feature. In the present case, physical investigation showed no superficial lymphadenopathies. Furthermore, both enhanced chest CT and PET confirmed no lesion in the cervical region. To the best of our knowledge, this is the first case of Kikuchi-Fujimoto disease with isolated mediastinal lymphadenopathy reported in the literature. Some cases of systemic lymphadenopathies have been described previously. Lee et al (4) reported increased uptake on PET in the right cervical parotid space, bilateral upper to lower cervical, supraclavicular, axillary, intra-abdominal, and retroperitoneal regions (4). Another case with no evidence of cervical or supraclavicular adenopathy was reported recently by Fernández et al (5), in which CT showed not only enlarged mediastinal and subcarinal lymph nodes but also small peripherally distributed subpleural nodules.

FDG-PET is a convenient tool widely used now to investigate the location of malignancy foci (10). This method utilizes the feature of amplified glucose activity and glycolysis in malignant cells (11). However, benign lesions, some inflammation (12) and sarcoidosis (13) also demonstrate increased FDG uptake as in malignant lesions, and it is not possible to distinguish malignant from benign disease by SUV at a single time point. New advanced PET technology examines early- and late-phase SUV, and increased late-phase compared to early-phase SUV is expected to suggest malignant (14). In the present case, PET also showed late-phase SUV augmentation. This finding suggested malignant lymphoma affecting mediastinal lymph nodes (15), and surgical excision of the lymph node was necessary to make a definitive diagnosis. In several reports of Kikuchi-Fujimoto disease showing high late SUV on PET, surgical biopsies of lymph node were necessary for differentiation from malignant lymphoma.

In the present case, physical examination, routine laboratory data and radiographs showed no specific abnormalities, and the patient had no chest-related symptoms. In the clinical setting, when a patient with the above conditions is encountered, not all doctors will proceed to perform enhanced chest CT. Therefore, it is possible that some cases of Kikuchi-Fujimoto disease could have been missed or misdiagnosed as some type of infection and prescribed antibiotics.

The authors state that they have no Conflict of Interest (COI).

References

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