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Waldenström Macroglobulinemia Mimicking A Primary Lung Carcinoma

Primer Akciğer Karsinomunu Taklit Eden Waldenström Makroglobulinemisi

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ABSTRACT

Waldenström macroglobulinemia (WM) is a rare hematological disease that accounts for 1-2% of non-Hodgkin lymphomas. WM is a variant of lymphoplasmocytic lymphoma and is characterized by bone marrow involvement as well as immunoglobulin M monoclonal gammopathy. It occurs as 3 to 4 cases per million each year. Extramedullary involvement of this entity is rare, and lung involvement may present radiologically as a mass, nodular lesion, diffuse infiltration, or pleural effusion. The diagnosis can be made by performing a biopsy of the pulmonary parenchyma, bronchoalveolar lavage, or cytological examination of pleural fluid. Here, we aimed to present a case of WM mimicking primary lung carcinoma radiologically.

Keywords: Waldenström macroglobulinemia, lung cancer, VATS, PET-CT

ÖZ

Waldenström makroglobulinemisi (WM), Hodgkin dışı lenfomaların %1-2'sini oluşturan nadir bir hematolojik hastalıktır. WM, lenfoplazmositik lenfomanın bir varyantıdır ve kemik iliği tutulumu ve immünoglobulin M monoklonal gammopati ile karakterizedir. Yılda milyonda 3-4 vakada görülür. Bu antitenin ekstramedüller tutulumu nadirdir ve akciğer tutulumu radyolojik olarak kitle, nodüler lezyon, yaygın infiltrasyon veya plevral efüzyon olarak ortaya çıkabilmektedir. Tanı, pulmoner parankim biyopsisi, bronkoalveolar lavaj veya plevral sıvının sitolojik incelemesi ile konulabilmektedir. Burada, radyolojik olarak primer akciğer karsinomunu taklit eden bir WM olgusunu sunmayı amaçladık.

Anahtar Sözcükler: Waldenström makroglobulinemisi, pulmoner karsinom, VATS, PET-CT

INTRODUCTION

Waldenström macroglobulinemia (WM) is a rare hematological disease that accounts for 1-2% of non-Hodgkin lymphomas (NHL) (1). It is characterized by B lymphocytes, lymphoplasmacytoid cells, and plasma cell infiltration in the bone marrow, as well as serum immunoglobulin M (IgM) monoclonal gammopathy (2). It occurs as 3-4 cases per million each year. Extramedullary involvement is rarely seen in patients with WM and is usually seen found in the lungs, soft tissue, central nervous system, kidneys, and bone (3). Lung involvement of WM is rare and may present as a mass, nodular lesion, diffuse infiltration, or pleural effusion (2). When a lung lesion is detected in patients with WM, extramedullary involvement of WM, pneumonia, primary, and metastatic lung carcinoma should be considered in the differential diagnosis (4). Here, we aimed to present a case of lung involvement of WM mimicking primary lung cancer.

CASE REPORT

A 58-year-old male patient, an active smoker, was admitted with left hip pain, and a mass lesion was detected in the left iliac wing. A

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biopsy was taken from the bone, and histopathological examination revealed that bone material contained B-cell lymphoid neoplasia with prominent plasmacytic differentiation. In further examinations, the bone marrow was observed to be normocellular. In laboratory findings, serum protein electrophoresis showed: beta globulin-9.73%; gamma globulin-44.28%; and an M-spike was observed. The final diagnosis was WM, and the patient received 2 cycles of rituximab and bendamustine. In the interim evaluation, a suspicious lesion was observed in the patient's chest X-ray. Thorax computed tomography (CT) was performed, and a nodular lesion with a 2 cm diameter was detected in the upper lobe of the right lung (Figure 1A). Positron emission tomography-CT showed pathologically increased uptake maximum standardized uptake (SUV_{max}: 5.9) of 18F- florodeoksiglukoz on the pulmonary nodule, and there was neither mediastinal lymph node involvement nor extrapulmonary uptake (Figure 1B). Videothoracoscopic wedge resection/frozen section procedure was planned because transthoracic fine needle aspiration biopsy was not diagnostic. Frozen section examination was reported to be a haematological malignancy. The postoperative period of the patient was uneventful, and he was discharged on the 4^{th} day of the operation. Histopathological examination revealed low-grade B-cell lymphoma, with marked plasmacytic differentiation. Tumor consisted of lymphoid cells with small hyperchromatic nuclei, indistinct nucleoli, and layers of plasma cells containing prominent Russell bodies. The immunohistochemical study showed that lymphoid infiltration was stained with CD20, while plasma cell sheets were positive with CD138, MUM-1, and VS38C. Although plasma cells were Kappa negative, they were monotypic with lambda (Figures 2, 3). The third cycle of the rituximab and bendamustine protocol was administered to the patient, and the follow-up of the patient continued uneventfully. The informed consent form was obtained from the patient for publication.

DISCUSSION

Here we present a case of pulmonary WM mimicking primary lung malignancy. It has been emphasized in the literature that the incidence of WM is higher at age 70 and in males (2). Although our case was male he was younger than those in the literature. Most of the WM patients with lung involvement are asymptomatic at the time of diagnosis,some may present with cough, shortness of breath and chest pain (5). Also, our case was asymptomatic in agreement with the literature. In radiological imaging, lung lesions can be observed as a mass, nodular lesion, diffuse infiltration, or pleural effusion (2). In our case, the lesion presented as a solitary pulmonary nodule, and it was not possible to rule out primary lung malignancies with radiological imaging alone. There are cases of coexistence of NHL with non-small cell carcinoma in the literature (6). Banwait et al. (2) reported the rate of extramedullary involvement of WM was 4.3% in a large series. Lung involvements occurred as mass, nodules, or pleural effusion. Patients with IgM monoclonal gammopathy of uncertain significance (MGUS) have an increased risk of developing WM. Therefore, IgM MGUS is thought to be the precursor of WM, and myeloid differentiation primary response 88 (MYD88) mutations in most cases of WM indicate the potential role of this mutation in pathogenesis. The definitive diagnosis of pulmonary WM is made by histopathological examination and immunohistochemical study. B cell markers; immunoglobulin light chain and Ig-M positivity; cyclin D1, CD5, and CD10 negativity were observed in immunohistochemical studies (3). Histopathological examination and immunohistochemical studies of our case were consistent with the literature. However, no

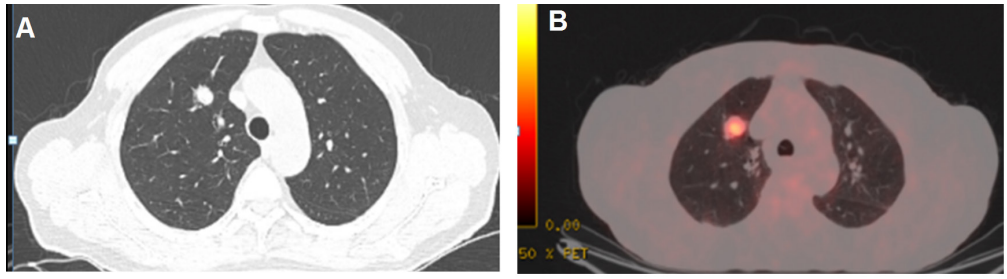

Figure 1. (A) Thorax computed tomography shows a solitary pulmonary nodule with a diameter of 2 cm in the right upper lobe. B) Pathologic increased uptake of 18f-FDG on positron emission tomography/ computed tomography is seen (SUV-max: 5.9).

FDG: Florodeoksiglukoz, Max: Maximum

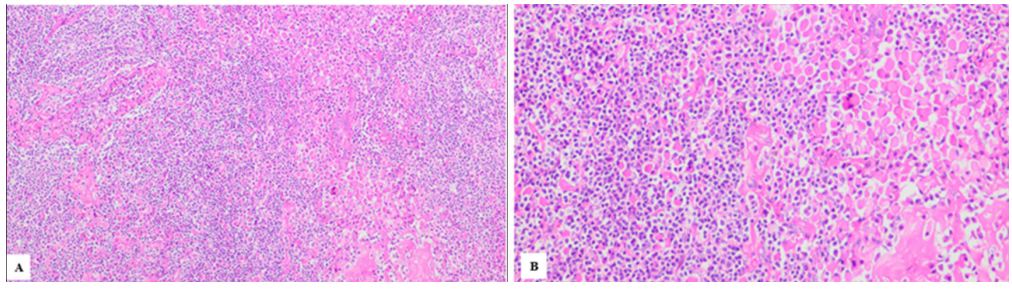

Figure 2. Hematoxylin and Eosin stain shows, low-grade B-cell lymphoma with marked plasmacytic differentiation and tumor cells consist of lymphoid cells with small hyperchromatic nuclei, indistinct nucleoli and layers of plasma cells containing prominent Russel bodies (A: 100 magnification, B: 200 magnification).

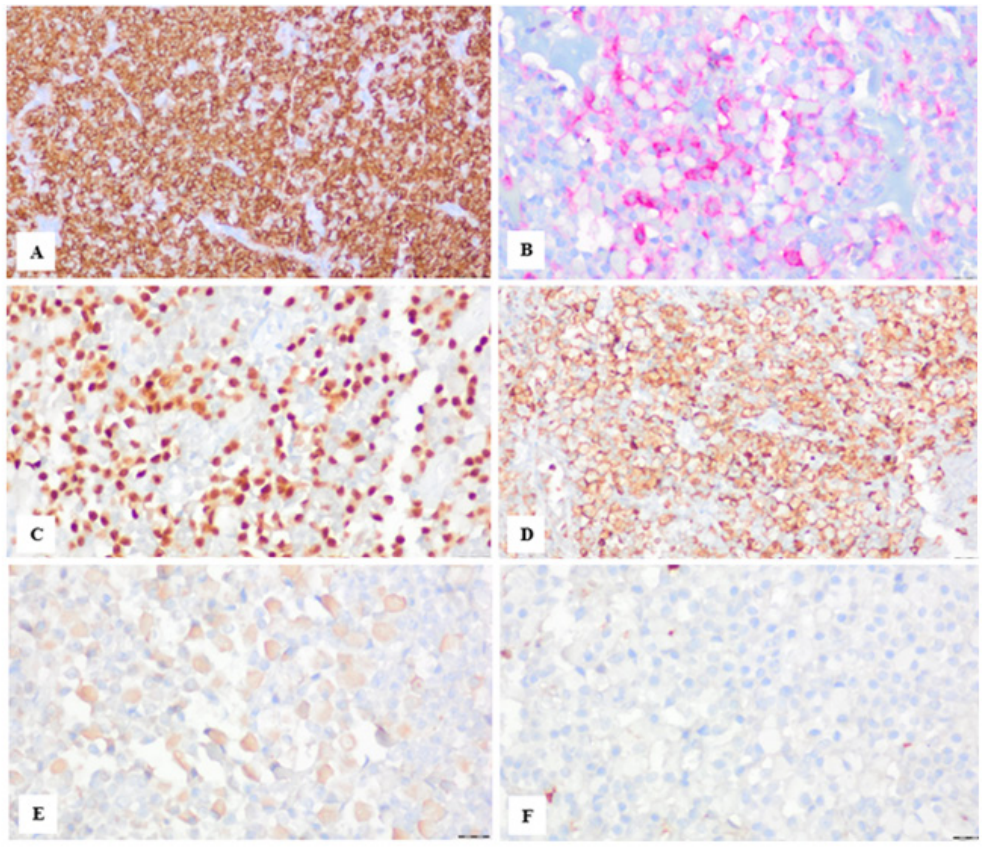

Figure 3. Immohistochemical study of lesion; (A) lymphoid infiltration shows CD20 positivity, B) plasma cell sheets are positive with CD138, C) plasma cell sheets are positive with MUM-1, D) plasma cell sheets are positive with VS38C, E) plasma cells are monotypic with lambda, F) kappa is negative.

MYD88 mutation was observed. WM is usually a slow-progressing disease, and the median survival is relatively good. Advanced age, peripheral blood cytopenia, high beta-2 microglobulin levels, and high serum IgM (>7 g/dL) levels were reported as poor prognostic factors (3). None of the poor prognostic factors was present in our case, and his follow-up continues uneventfully.

CONCLUSION

Although the association of NHL and pulmonary malignancies is rarely reported, pulmonary malignancies should be kept in mind in the differential diagnosis and histopathological confirmation should be performed. Informed consent form was obtained from the patient for publication

Ethics

Informed Consent: Informed consent form was obtained from the patient for publication.

Footnotes

Authorship Contributions

Surgical and Medical Practices: I.A., E.V., M.T., M.S, A.C., T.K., N.A., Concept: I.A., N.A., M.S., A.C, Design: I.A., N.A., M.S. A.C, Data Collection or Processing: I.A., E.V., M.T., M.S., T.K., N.A., Analysis or Interpretation: I.A., N.A, M.S., A.C., Literature Search: I.A., E.V., M.T., N.A., M.S., A.C., Writing: I.A., T.K., N.A., M.S., A.C.

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