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Case Report
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High Efficacy Achieved in the Case of Lymphomatoid Granulomatosis with Targeted Therapies

Buyuksimsek M[1], Ogul A[1], Yetisir AE[1], Koseci T[1], Duman BB[1], Tohumcuoglu M[1], Kesen O[1], Cil T[1], Arbag HC[2] and Sumbul HE[2]*

[1]*Department of Medical Oncology, University of Health Sciences, Adana Health Practice and Research Center, Turkey*

[2]*Department of Internal Medicine, University of Health Sciences, Adana Health Practice and Research Center, Turkey*

Abstract

Objective and Importance: In this article we present a case of the lymphomatoid granulomatosis which good response achieved with targeted therapies.

Clinical Case: A 71-year-old female patient presented with complaints of upper respiratory tract and weight loss. A mass lesion which filled the right hemithorax almost completely was detected and there were multiple lymph nodes in the mediastinum in the tomography imaging. Trucut biopsy was performed and pathology report was compatible with CD20+, CD30+ lymphoproliferative disease, Lymphomatoid Granulomatosis (LG). The patient was planned to be treated with CD20 and CD30 targeting agents, rituximab, brentuximab vedotin, and the antineoplastic agent bendamustine. After 3 cures of treatment, almost complete regression was detected in the lung mass lesion and mediastinal lymph nodes.

Discussion: High-grade LG cases are considered to be aggressive high-grade lymphomas and require aggressive treatments. We wanted to present this case to emphasize the importance of using targeted agents in LG patients for whom treatment modalities are not clear.

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Correspondence:
Mahmut Buyuksimsek, Department of Medical Oncology, University of Health Sciences, Adana Health Practice and Research Center, Dr. Mithat Özsan Bulvarı Kışla Mah, 4522 Sok. No: 1 Yüreğir, Adana, Turkey, Tel: +90-0322455 (Ext: 9000414); E-mail: mahmutbuyuksimsek@gmail.com

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Introduction

Lymphomatoid Granulomatosis (LG) is a rare lymphoproliferative disease associated with Epstein-Barr virus, characterized by multiple pulmonary nodular lesions with lymphocytic invasion of the vascular walls on biopsy [1,2]. The most common presenting symptoms are cough, fever, malaise, weight loss, neurological abnormalities, and shortness of breath [3]. Lesions in the lung are usually well circumscribed and consist of small lymphocytes, plasma cells, and a variable number of large atypical mononuclear cells. Most small lymphocytes are T cells, while large atypical cells are usually B cells [4]. B cells typically express CD20, but rarely CD30. They are negative for CD15 [5]. Rarely, patients are asymptomatic and have histopathologically low-grade (grade 1 and 2) disease limited to the lungs. Sometimes spontaneous remission can occur in this patient group so these patients can be followed up with clinical and radiological evaluations. Active treatment is usually required if symptoms develop or if the disease progresses radiologically [6]. In general, the recommended treatment options for high-grade (grade 3) LG consist of the treatment modalities used for Diffuse Large B-Cell Lymphoma (DLBCL), but there is not much data on the effectiveness of these treatments. Other recommended treatments include multi-agent chemotherapy and targeted agents such as anti-CD20 and anti-CD30 monoclonal antibodies (rituximab, brentuximab vedotin) [7-9].

Case Presentation

A 71-year-old female patient presented with complaints of cough, shortness of breath, and weight loss. In the whole body tomography imaging, a mass lesion with central hypodense soft tissue density, which filled the right hemithorax almost completely, was detected and there were multiple lymph nodes in the mediastinum (Figure 1). Trucut biopsy of the right lung was performed on the patient; pathology report was reported as compatible with CD20+, CD30+ Lymphoproliferative disease, Lymphomatoid Granulomatosis. The patient was planned to be treated with CD20 and CD30 targeting agents, rituximab and brentuximab vedotin, and the antineoplastic agent bendamustine. Rituximab 375 mg/m^2 1st day, brentuximab vedotin 1.8 mg/kg 1st day, bendamustine 90 mg/m^2 1+2 Day; treatment was started every 28 days. After the first course of treatment, the

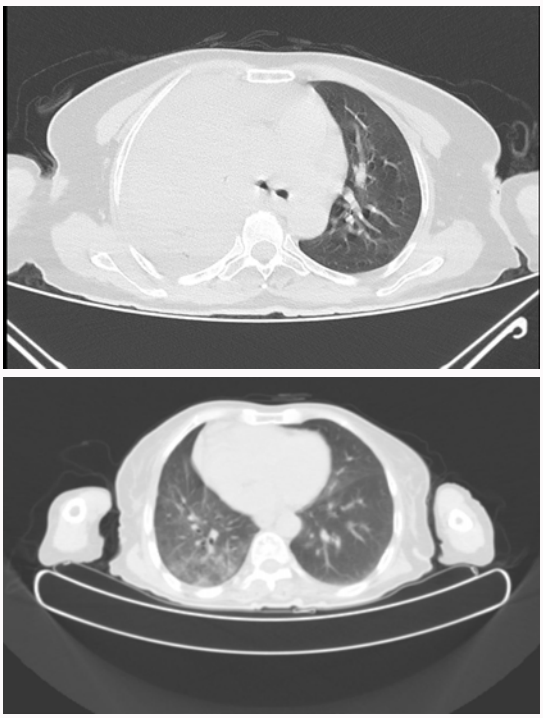

Figure 1, 2: Thorax tomography images of the patient.

patient's clinical complaints regressed significantly. After 3 cures of treatment, the patient was evaluated by taking a tomography again. Almost complete regression was detected in the lung mass lesion and mediastinal lymph nodes (Figure 2). After the patient's treatment was completed for 6 cycles, it was planned to discontinue bendamustine and continue with rituximab and brentuximab vedotin targeting agents.

Discussion

LG was first described in 1972 by Liebow et al. [10]. It is a very rare type of extranodal lymphoma and it exhibits a very characteristic angiodestructive behavior. High-grade cases are considered to be aggressive high-grade lymphomas and patients require prompt and intensive treatment with immunochemotherapy agents including rituximab [11]. In our case, besides CD 20 positivity, CD 30 was also detected. Thereupon, we added brentuximab vedotin treatment together with rituximab to our patient who had a mass lesion covering the right lung. We also preferred bendamustine as a chemotherapeutic agent and we obtained a very satisfactory result in our patient. We wanted to present this case to emphasize the importance of using targeted agents in LG patients for whom treatment modalities are not clear.

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