Case Report

Primary Pulmonary Diffuse Large B-Cell Lymphoma Presenting as an Endobronchial Lesion: The Youngest Adult Patient in the Literature

ONUR TURAN[1], PAKIZE AYŞE TURAN[2], MEHMET ALI UYAROĞLU[3], FATOŞ POLAT[1], AYLIN ÖRGEN ÇALLI[3]

[1]*Chest diseases Department, Izmir Katip Celebi University Atatürk Training and Research Hospital, Izmir, Turkey*

[2]*Chest diseases Department, Menemen State Hospital, Izmir, Turkey*

[3]*Pathology Department, Izmir Katip Celebi University Atatürk Training and Research Hospital, Izmir, Turkey*

ABSTRACT: A 20-year-old female patient was admitted to hospital with complaints of chest and back pain in September 2018. There was a cavitary lesion in the upper zone of the left lung in the chest X-ray. Thorax CT revealed an irregular contoured and shaped mass with 87x67x79 mm sizes, in the upper lobe of the left lung lying to paramediastinal area. Since there was a doubt about malignancy, positron emission tomography (PET) was performed; there was a cavitary lesion in the left upper lobe with high FDG uptake (SUVmax: 23.2). Bronchoscopic examination revealed an endobronchial lesion with nearly complete occlusion in the apicoposterior segment of the left upper lobe. Bronchoalveolar lavage (BAL) performed in this session for acid-fast bacilli (AFB) was negative. The patient was diagnosed as primary pulmonary diffuse large B-cell lymphoma (DLBCL) by histopathological and immunohistochemical evaluation of endobronchial biopsy specimens. Following the final diagnosis of Bronchus-Associated Lymphoid Tissue Lymphoma (BALTOMA), the patient was referred to the department of haematology, and chemotherapy was planned for therapy. Since DLBCL is extremely rare, and uncommonly presenting with an endobronchial lesion, we want to present this patient as the youngest adult case of primary endobronchial BALT lymphoma in the literature.

KEYWORDS: *B-cell lymphoma, endobronchial lesion, primary pulmonary lymphoma*

Introduction

Primary pulmonary lymphoma (PPL), an uncommon type of non-Hodgkin's lymphoma (NHL), involves pulmonary parenchyma and/or mediastinal lymph nodes in the or hilar areas [1].

It represents approximately 0.5-1% of primary pulmonary malignancies [2].

Primary pulmonary diffuse large B-cell lymphoma (DLBCL) is also a rare type of PPL [3]. Since DLBCL has non-specific clinical symptoms and signs, it is not easily recognized and diagnosed. Open thoracotomy or chest computed tomography (CT)-guided percutaneous biopsy are the most preferred methods to put the diagnosis of DLBLC [4].

Bronchus Associated Lymphoid Tissue Lymphoma (BALTOMA) is a rare subgroup of pulmonary non-Hodgkin's lymphomas (NHLs) which forms less than 1% of all cases [5]. The most common histological subtype of PPL is known to be mucosa-associated lymphoid tissue (MALT) type in the literature [6]. BALTOMA is considered to arise and originate from this MALT type [7].

Endobronchial lesion infrequently occurs in lymphoma, which is more often in Hodgkin's disease (HD) than in NHL [8]. There are two major ways of endobronchial invasion for lymphoma; firstly, submucosal infiltrations or nodules by lymphatic or haematogenic metastasis, and secondly, solitary endobronchial lesion by direct invasion of nearby lymph nodes [9].

An extremely rare case of DLBCL presenting as an endobronchial involvement, which has been diagnosed by bronchoscopic biopsy is reported.

Case Report

A 20-year-old female patient was admitted to hospital with complaints of chest and back pain in September 2018. The patient had no prior history of lung disease. She was a student in university. Besides, she was a nonsmoker without any important disease as a family history. She had no exposure to any pollutants in her anamnesis.

The vital signs of the patient were in normal ranges. There was no positive finding in physical examination, including respiratory system. In addition, no palpable lymph nodes observed.

A complete blood test including a white cell count, hepatic and renal functions of the patient was unremarkable. She had a raised erythrocyte sedimentation rate (ESR) as 39mm/h, and C-reactive protein (CRP) as 3mg/dl.

There was a cavitary lesion in the upper zone of the left lung in the chest X-ray (Figure 1).

Thorax CT revealed an irregular contoured and shaped mass with 87x67x79mm sizes, in the upper lobe of the left lung lying to paramediastinal area (Figure 2).

The serum levels of carcinoembryonic antigen, neuron-specific enolase and CYFRA21-1 were unremarkable.

Since there was a doubt about malignancy, positron emission tomography (PET) was performed afterwards. There was a cavitary lesion in the left upper lobe with high FDG uptake (SUVmax: 23.2) (Figure 3).

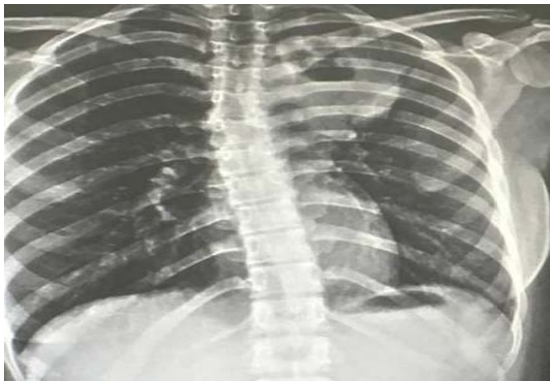

Figure 1. A cavitary lesion in the upper zone of the left lung (chest X-ray).

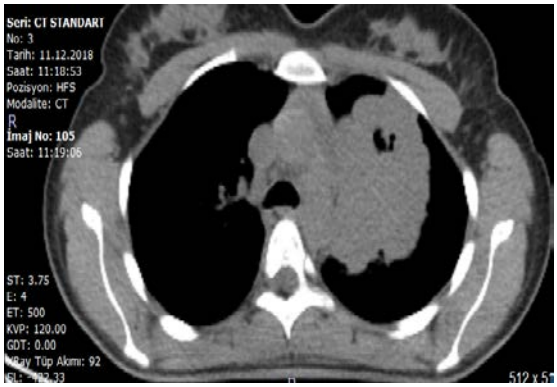

Figure 2. A lesion in the upper lobe of the left lung lying to paramediastinal area (thorax CT).

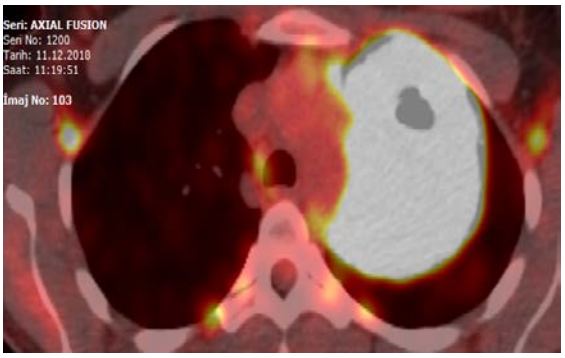

Figure 3. A lesion in the left upper lobe with high FDG uptake (positron emission tomography).

In the same week, bronchoscopy (bronchial biopsy by forceps and BAL) was performed, which revealed an endobronchial lesion with nearly complete occlusion in the apicoposterior segment of the left upper lobe (Figure 4).

Bronchoalveolar lavage (BAL) examination for acid-fast bacilli (AFB) was negative. A routine immunohistochemistry antibody panel from endobronchial biopsy specimens revealed that the tumor cells were positive for CD20, CD10, LCA, Pax-5 and MUM-1, which confirmed the diagnosis of primary pulmonary diffuse large B-cell lymphoma (DLBCL) (Figures 5-6).

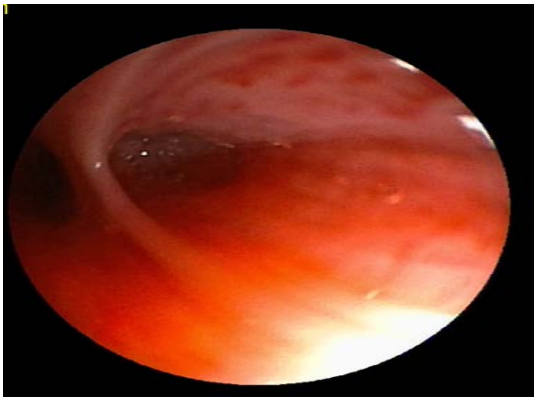

Figure 4. An endobronchial lesion in the apicoposterior segment of the left upper lobe.

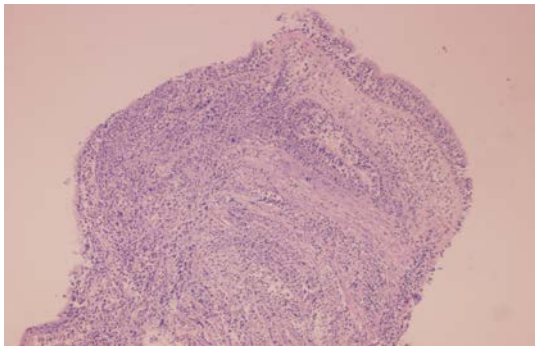

Figure 5. Diffuse infiltrate of tumor cells with scant cytoplasms (HE staining), 5x.

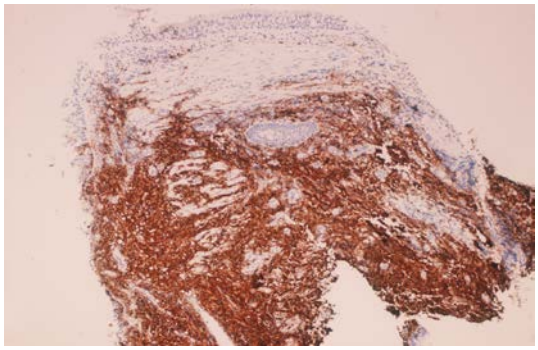

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Figure 6. CD20 immunostaining in tumor cells; which confirm the diagnosis of primary pulmonary diffuse large B-cell lymphoma, 5x.

Following the final diagnosis, the patient was admitted in the department of haematology, and R-CHOP chemotherapy (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) was planned for therapy.

Discussion

PPL is an uncommon type of both lymphomas and primary pulmonary malignancies. It comprises less than 1% of all NHL cases [8]. Primary pulmonary diffuse large B-cell lymphoma (DLBCL) occurs only in 10% cases of primary pulmonary NHLs [10].

Since pathogenesis of primary pulmonary DLBCL has not yet to be clearly identified, there are a few theories about its etiology. One of them specifies that chronic inflammatory diseases could provoke clonal lymphoid proliferation affecting lung tissue [11]. Another review indicates that it may be associated with an immunosuppression observed in allograft recipients of lung (as a solid organ) [12]. It can be explained as the presence of chronic inflammation of the lung and various autoimmune diseases together.

Lung is one of the most frequent sites where mucosa-associated lymphoid tissue (MALT) lymphoma, named as BALTOMA, originates [13]. There are some scenarios about existence of DLBCL; some of them may show up with an isolated pulmonary parenchymal disease, and some can only arise with hilar or mediastinal involvement in the absence of extrathoracic disease [8]. There are a few reports on endobronchial BALT lymphoma existing as a primary tumour [14]. An endobronchial lesion, such as in our patient, occurs rarely as the primary presentation of DLBCL.

Endobronchial involvement with lymphoma is an infrequent condition, which usually occurs in the setting of widely disseminated disease [15]. Endobronchial lymphoma may present in two different types, according to pattern of involvement [16]. First type, which seems more usual, presents with prominent hilar or mediastinal adenopathy with direct extension to bronchi as the main cause of endobronchial involvement. The other type is an endobronchial growth as the only manifestation of the lymphoma, similar to our patient. As there was no additional lymphomatous involvement, endobronchial lesion seems to be the origin of lymphoma.

Most of the PPL cases in the literature present with systemic symptoms related to lymphoma or with localized pulmonary symptoms due to pulmonary involvement. Our patient had no systemic symptom about lymphoma. However, she had chest pain because of the nearby localization of the lesion with chest wall.

There was a cavitary lesion in the chest X-ray of the patient, and her thorax CT revealed a mass in the upper lobe of the left lung. Radiological findings are various in primary pulmonary DLBCL, which may generally include pulmonary nodules, masses, consolidation or hilar/mediastinal lymph nodes [17]. Chest X-rays and CT scans usually revealed endobronchial lesions with varying degrees of lobar collapse [18]. As seen, thoracic radiological findings are not specific, and they may only lead for further consideration of a pulmonary lesion.

The use of bronchoscopic evaluation has been reported in previous studies, but since the rate of diagnosis by bronchoscopic biopsy is rare, open thoracotomy or chest CT-guided percutaneous biopsy are the preferred methods [19]. The diagnostic contribution of bronchial biopsy was present in approximately 10% of patients in the previous studies [19,20].

In the present case report, fiberoptic bronchoscopy was performed, an endobronchial lesion was observed, and diagnosis was established by this bronchoscopic biopsy. Although open thoracotomic biopsy has a high positive diagnostic value, but it is also associated with some complications. Bronchoscopic examination with biopsy is essential to differentiate the endobronchial lesions from primary bronchongenic cancer. Our case demonstrates the importance and priority of bronchoscopic evaluation in these types of patients.

BALT lymphoma usually show good prognosis and a favorable long-term survival [21]. Besides, primary endobronchial lymphomas also have a better prognosis, favorable response to treatment with complete remission [8]. Our patient has undergone chemotherapy, and the period of treatment has been processing.

In conclusion, primary pulmonary DLBCL is a rare type of NHL with nonspecific signs and symptoms. It may present as an isolated endobronchial lesion without extrathoracic

manifestation. The clinicians may consider BALTOMA in the differential diagnosis of endobronchial lesions, which shows the importance of bronchoscopic examination in this type of cases. We report our case with an unexpected diagnostic type of lymphoma, primary pulmonary NHL presenting with an endobronchial lesion, which seems to be the youngest case of primary endobronchial BALT lymphoma in the literature.

Conflict of interests

None to declare.

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Corresponding Author: Onur Turan, Chest diseases Department,
Izmir Katip Celebi University Atatürk Training and Research Hospital, Izmir, Turkey,
e-mail: onurtura@yahoo.com