PRIMARY PULMONARY LYMPHOMA; A RARE CASE

PRİMER PULMONER LENFOMA; NADİR BİR OLGU

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Key words: Primary pulmonary lymphoma, primary pulmonary peripheral T-cell lymphoma (PTCL) lymphoepiteloid types, benign cystic disease, surgery.
Anahtar sözcükler: Primer Pulmoner Lenfoma, Primer Pulmoner periferik T-Hücreli Lenfoma lenfoepiteloid tip, Bening Kistik hastalık, Cerrahi

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SUMMARY

Lymphoproliferative diseases that affect the lung emerges in a broad clinical and pathologic spectrum. Although malignant lymphomas are usually seen primarily nodal origin, they can be seen in the extranodal localization. Lymphomas, starting in lungs primarily, are rare. Fifty three year-old female patient with ten-month persistent cough, sputum, complaints of shortness of breath was admitted to the clinic. Cystic lung diseases were thought in clinical and radiographic evaluations. Thoracotomy was performed. A cystic lesion was seen in the middle lobe. The lesion was sent for frozen section examination. Middle lobectomy was performed since frozen section examination was reported as malignancy for suspected material. Histopathological examination revealed the diagnoses of lymphoma. Clinical, radiological and histopathological diagnosis are difficult in Primary pulmonary lymphoma (PPL). Treatment is primarily surgical. In our study, it is highlighted that PPL is seen rarely and is similar to benign cystic lung disease and difficulties in diagnosis and treatment of PPL.

ÖZET

Akciğeri etkileyen lenfoproliferatif hastalıklar geniş bir klinik ve patolojik spektrumda ortaya çıkmaktadır. Malign lenfomalar genellikle primer lenf nodu kökenli olarak görülmesine rağmen, ekstranodal lokalizasyonda da görülebilir. Primer olarak akciğerden başlayan lenfomalar nadirdir. On aylık inatçı öksürük, balgamı olan 53 yaşındaki kadın hasta, nefes darlığı şikayeti ile polikliniğimize başvurdu. Kllinik ve radyolojik değerlendirmelerde kistik akciğer hastalığı olarak düşünüldü. Torakotomi yapıldı. Kistik lezyon orta lobda görüldü. Lezyon frozen inceleme için gönderildi. Frozen inceleme malignite kuşkulu olarak rapor edildi. Frozen incelemesi malignite kuşkulu olarak rapor edildiği için orta lobektomi yapıldı. Histopatolojik incelemede lenfoma tanısı rapor edildi. Klinik, radyolojik ve histopatolojik tanı Primer pulmoner lenfoma (PPL) 'da zordur. Tedavi öncelikle cerrahidir. Çalışmamızda, benign kistik akciğer hastalığı ile karışabilen ve nadir görülen PPL'nın tanı ve tedavisindeki zorlukları vurgulamak istedik.

INTRODUCTION

When Lymphoproliferative diseases affecting the lung occur, they come across a wide clinical and pathological spectrum (1). Althought malignant lymphomas are usually seen in nodal, they can be seen extranodal localization. PPL; are seen in 0.5-1% of all nonhodgkin lymphomas (NHL) (1). Clinical, radiological and histopathological diagnosis is difficult and is still controversial. In this study, a very rare case of PPL similar to benign cystic lung disease is presented and difficulties in diagnosis and treatment phases are highlighted.

CASE REPORT

Fifty three-year-old female patient with ten months persistent cough, sputum, shortness of breath was admitted to our clinic. In the history, she had ischemic heart disease and hypertension, crepitan crackles were present in middle and lower zones of the right hemithorax. Laboratory tests were normal except HB: 11.8 g / dl, WBC: 11300 / mm3, platelets: 123 000 / mm3, LDH: 383, albumin: 3.39. There were 59,7% neutrophils, 34,6% lymphocytes, 5,25% monocytes in peripheral blood smear. In the chest radiograph, oval shaped diaphragm-based nodular opacity was diagnosed at the lower zone of right lung medially (Figure 1). Gastroscopy and echocardiography were normal. In thorax CT in January 2013; dimensions of 70x60 mm in the medial segment of the right middle lobe, causing compression of the right atrium, well-defined, sharply circumscribed, homogeneous internal structure, low in soft tissue density (45-50 HU) mass lesion was seen. No significant invasion was seen on neighbor soft tissues. Pathologic size lymphadenopathy was not seen in mediastinum. In thorax CT in December 2013; no significant changes were diagnosed except minimal compression atelectasis in the lateral segment of the middle lobe (Figure 2). Abdominal and brain CT scan were normal. Malignancy was not diagnosed in bronchoscopy, postbronchoscopic sputum and bronchoalveolar lavage (BAL). Patient was operated with the pre-diagnosis of the cystic lesion of the lung. In thoracotomy, soft with palpation, well-circumscribed, round, 12x10x5 cm cystic mass was seen in the medial of the middle lobe. Cystotomy was performed. The interior of the cyst was filled with dark mucoid fluid and necrotic tissue fragments. Biopsy was taken and sent for frozen section. Frozen section was reported as germinal centers not containing significant follicle structure in terms of creating the diffuse lymphoid infiltration and suspected malignancy because of lymphoid infiltration. Other lung fields were normal. Middle lobectomy was performed and mass was removed (Figure 3).

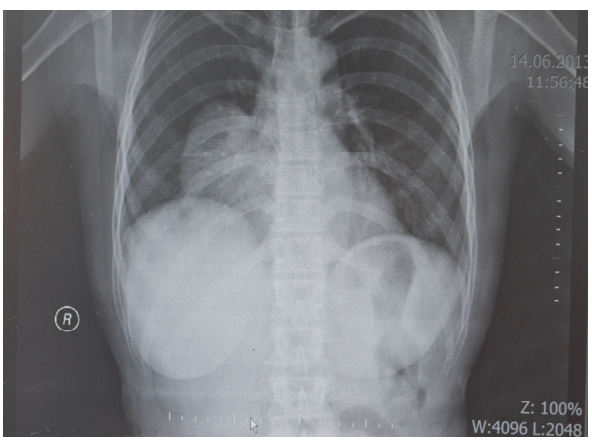

Figure 1.

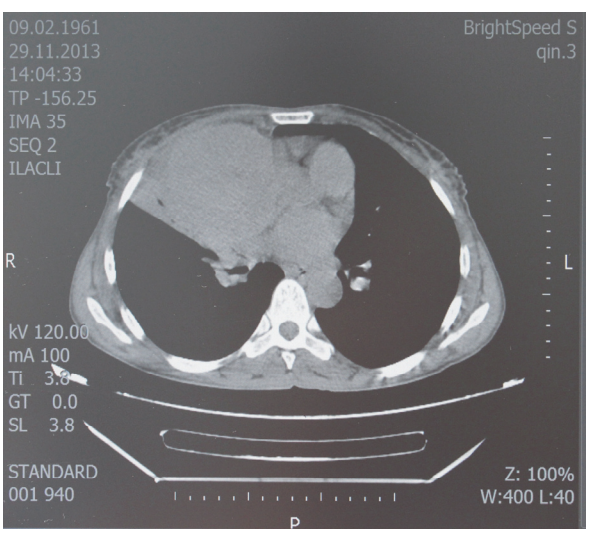

Figure 2.

Figure 3.

In histopathological examination; tumor, seperated from the surrounding lung parenchyma by sharp boundaries, was consist of slim, vascularized and rich for histiocytes, diffuse, with small hyperchromatic nuclei, in some places significant nucleoli, pleomorphism including lymphocytic cells and between them settled in large cytoplasm, large vesicular nuclei, prominent nucleoli, with single distributed variant with Hodgkin, similar to Reed Sternberg (RS) cells (Figure 4). In Immunohistochemical examination of the material; tumor cells were stained with strong positive CD 3, CD8 and CD 5. Focally CD 20 and CD 79 was seen weak stained positively Reed Sternberg-like cells variant with Hodgkin. Defined in large vesicle nuclei cells, focal weak positive staining with CD15 was seen. CD 30 was negative, histiocytes stained strong positively with CD 163 were seen in the ground. Peripheral T-cell lymphoma (PTCL), lymphoepiteloid (Lennert) type were considered with these findings but Hodgkin's disease (HD) and TCELL \ rich histiocytes B-cell lymphoma (THBL) could not be fully ruled out. Hilar, paratracheal, intraparenchymal paraesophageal lymphadenopathies were reported as anthracotic, reactive lymph nodes.

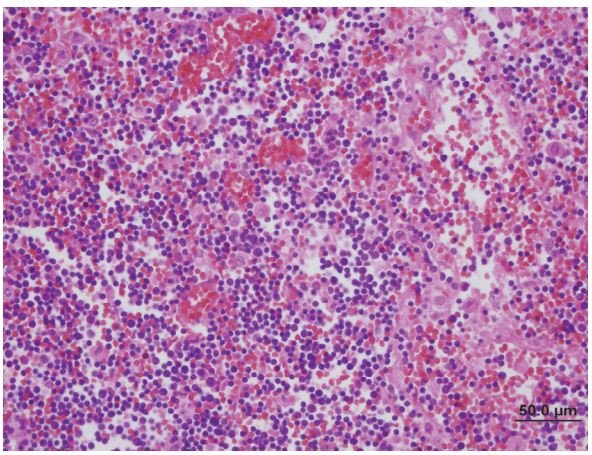

Figure 4.

Metastases and lymph node involvement were not observed in 3-month postoperative chest-abdominal CT scan. However, it was learned that the patient hampered the controls in the directed oncology clinic, thus the treatment was delayed. At postoperatively 6-months, brain MR was taken since she was with TA: 190/100 mmHg, right hemiplegia, motor aphasia. Acute ischemia and extensive edema were seen in frontal part of her brain. She died in neurology clinic where she was treated. The time between first diagnosed radiologically and death was eighteen months.

DISCUSSION

Pulmonary lymphomas are usually localized secondarily, rarely encountered in the primary location. When PPL was defined, it was only used for lymphomas arising from the lung parenchyma without extrathoracic and mediastinal involvement. It was defined as lymphoproliferative disease of single or both lung and/or bronchus without extrapulmonary involvement during diagnosis or following 3 months (2). Koss et al. accept the lymphoma of lung or lung with regional lymph nodes without extrathoracic focus as PPL (3).

PTCL is seen in PPL rarely and starting in the lung is a very rare case. PTCL does not correspond to any classification of nodal and extranodal mature T-cell lymphoma. They are usually seen in adults. Specific T-cell lymphoma entity should be excluded completely to diagnose PTCL (5).

It was reported that the time between the first clinical application or radiological findings and diagnosis in PPL changes five months to eight years. It is usually quiet and indolent, about half of the patients have no complaints at diagnosis. Patients may present with cough, dyspnea and chest pain. Physical examination findings are usually normal. It is stated that the clinical diagnosis is difficult. It is noteworthy complaints begin several weeks or months ago (3). Slow and quiet clinical symptoms can cause delay in applying medical institutions (2).

Radiologically, they are usually opacities smaller than 5 cm, multiple (70-77%), bilateral (60-70%), which are located in the lower lobes accompanied by air bronchograms 50% (2). PPL sometimes, as in our case, can be greater than 5 cm, well-circumscribed, medial lesions, cystic, slow and quiet, indolent and it can be interfered with bronchogenic cyst, teratoma, thymic neoplasms, germ cell tumors mimicking cystic disease (2).

PPL can be controversial in histopathologically as in clinically. Immunophenotypic features is variable since PTCL is a heterogeneous disease (5). In our case, HD, TCBL and PTCL was thought cytogically. Presence of Hodgkin and RS-like cells made us away from TCBL and focal positivity with CD 20, CD 79, and CD 30 negativity made us away from HD then led us to think PTCL. However, this can be seen in varying proportions in that diseases. It has therefore been desired to support the cytogenetic diagnosis. As a result, it is diagnosed on the addition morphological, immunohistochemical and cytogenetic studies.

Determination of the prevalence of the disease is important for the treatment and follow-up. Not being a different involvement in thoracic, abdominal and pelvic CT or MRI supports the diagnosis of PPL and can provide additional information about the extent of the disease (2).

An important test for staging and diagnosis of lymphoma is also bone marrow biopsy. However, today it has been reported that bone marrow involvement can be diagnosed by using imaging techniques such as PET-CT and MRI (4). Sufficient size of biopsy specimens is required to diagnose. It is recommended that bronchoscopy is initial for it. It is important to be seen lymphocytic alveolitis in sputum cytology and BAL (1). In most cases, a definitive diagnosis can be performed by thoracoscopy and open lung biopsy (6).

Surgery can be applied to obtain sufficient tissue and perform resection for treatment in lymphomas (6).It is necessary not to be a systemic spread for therapeutic purpose surgery. If surgery is done for PPL; sampling of mediastinal, hiler, intraparenchymal lymphadenopathy is important for staging of the disease. Incisional biopsy can be used for diagnosis in patients with widespread involvement and the lesions which cannot be resected (6).

In PPL, although the choice of treatment is done for tumor histological subtypes, tumor mass and accompanying diseases, the most important factor in them is the histology of the tumor. Although not a common decision today, treatment options include surgery, chemotherapy and radiotherapy (6,7). Surgery is recommended as the first choice for localized tumors, chemotherapy is performed in bilateral, extrathoracic lesions and progressing cases. In the treatment strategies of PTCL the most extensively studied strategies are chemotherapy protocols (7). Also another studied area is autologous stem

cell transplantation protocols. It was reported that five-year survival is 76% in some types of PPL, the average survival time is more than ten years (6).

In conclusion; PPL is quiet and indolent disease, clinical, radiological and histopathological diagnosis are difficult. It can interfere with cystic disease of the lungs and rarely are seen. In the limited disease of PPL, surgical treatment can be thought.

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