Case Report

ISSN: 1735-0344 *Tanaffos 2014; 13(2): 47-49*

TANAFFOS

Primary Composite Lymphoma of the Lung: a Case Report

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Received: 22 December 2013
Accepted: 13 March 2014

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Herein, we report a rare case of primary lung lymphoma in a 61 year-old woman with a history of 6-month nonspecific symptoms like dry cough, fever, chills and weight loss. She was admitted to a hospital and received broad-spectrum antibiotics but discharged without full recovery. In her second hospital admission, a bronchoscopic evaluation and transbronchial biopsy were performed, which were not diagnostic. Finally, an open lung biopsy was done. Immunohistochemical (IHC) staining of the specimen suggested pulmonary Hodgkin lymphoma. Because of disease recurrence, a second bronchoscopy was performed and endobronchial biopsy revealed transformation to anaplastic lymphoma. In the second recurrence, we decided to reevaluate the last biopsy specimens in greater details. Finally, after conduction of several staining patterns, the diagnosis of primary composite lymphoma of the lung was made.

Key words: Pulmonary Hodgkin lymphoma, Anaplastic lymphoma, Primary lung composite lymphoma

INTRODUCTION

Primary lung lymphoma is a rare disease and refers to the proliferation of lymphoid progenitor cells in the airways or the lung parenchyma when there is no extra-pulmonary involvement at the time of diagnosis or within 3 months. Extra-nodal non-Hodgkin lymphomas involving the lungs comprise less than 1% of all non-Hodgkin lymphomas (NHL) and 0.5-1% of all primary lung malignancies (1,2).

Composite lymphoma is a rare disease that has recently gained attention. It refers to the coexistence of more than one lymphoma in a single organ (3).

CASE SUMMARIES

A 61 year-old woman presented with dry cough, fever, chills and progressive dyspnea. She was admitted to a hospital and received antibiotic therapy with no improvement. She was readmitted after two weeks of discharge for further evaluation. She experienced 8 kg of weight loss during 4 months and reported nocturnal fever, chills and sweating. Her past medical history included a diagnosis of myasthenia gravis about 15 years ago treated for 12 years. She also had recently diagnosed diabetes mellitus and maculopapular rash on her cheeks. There was no palpable adenopathy; liver and spleen were normal as well. Vital signs were stable except for tachycardia. A chest CT scan revealed multiple nodular and mass like densities on both right and left sides of the diaphragm (Figure 1).

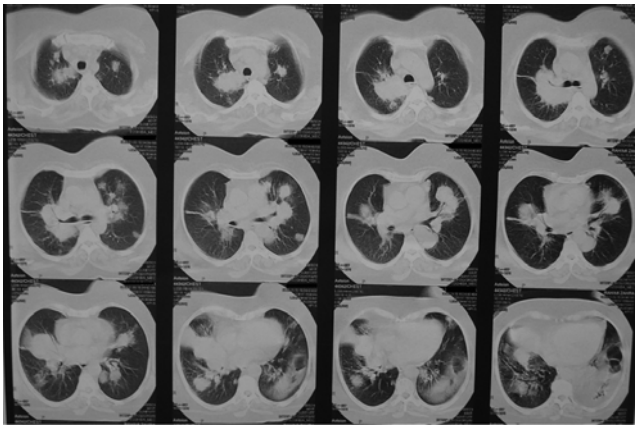

Figure 1. Multiple mass-like densities in both lung fields

No hilar or mediastinal lymph nodes were significantly visible. An extensive workup including endoscopic examination of the gastrointestinal tract, mammography and echocardiography was performed with unremarkable results. A bronchoscopic exam revealed a polypoid lesion in the superior segment of bronchus of the right upper lobe and a biopsy was taken that was reported to be non-diagnostic by a pathologist. Bronchoalveolar lavage cytology was negative for malignancy and acid fast bacilli. Laboratory tests showed neutrophilic leukocytosis with a shift to the left and normal liver and thyroid function tests. Serum protein electrophoresis was normal except for mild elevation of alpha 1 and alpha 2 bands. A bone marrow aspiration and biopsy showed 60% cellularity with a mild increase in megakariocytes, normal myeloid and erythroid cells with normal maturation and no blasts. Lymphoid cells were about 10-12%. ANA, cANCA, pANCA and anti dsDNA were within the normal limits. An excisional biopsy was taken from the right parahilar mass. There was no significant lymph node for biopsy. The pathology report and the results of IHC staining revealed lymphoreticular cells and scattered eosinophils. Some reticular cells were arranged atypically and scattered mononuclear Hodgkin cells were detected besides occasional double and multinuclear Reed-Sternberg cells (Figure 2). IHC staining was positive for CD30 and EMA and negative for CD15 and LCA in Hodgkin cells.

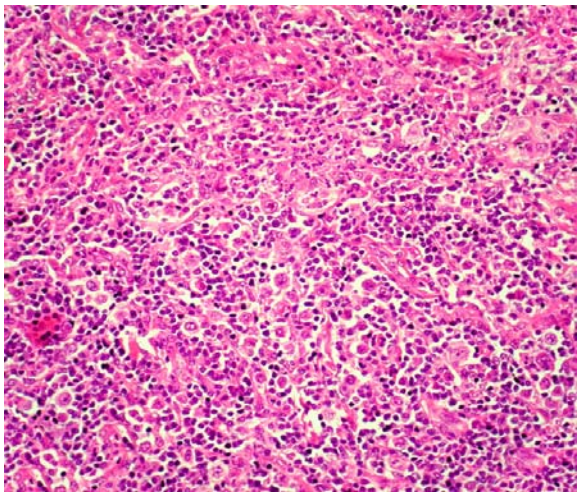

Figure 2. Polymorphic lymphoid cell infiltration along with Reed-Sternberg cells

Treatment: She was treated by 7 courses of ABVD monthly. Repeated chest scan showed pulmonary improvement. Abdominopelvic CT scan was normal. Six months later she was presented with cough and hemoptysis. A chest CT scan showed scattered bilateral nodules and a prominent right hilar mass. A second bronchoscopic evaluation was performed in which a flat velvety lesion on bronchus intermedius was detected and an endobronchial biopsy was taken. The pathology report indicated Hodgkin disease transformation to anaplastic large cell lymphoma that was positive for CD30 EMA LCA and negative for Chromogranin and cytokeratin. She was treated with ESHAP regimen for 5 courses with three-week intervals and at the end of treatment the chest and abdominopelvic CT scans were disease-free. Four months later she presented with dyspnea, cough and hemoptysis. A chest CT scan revealed an irregular mass along with surrounding infiltration and left lower zone infiltration. We decided to re-evaluate her previous tissue specimens. A review of the samples revealed tumoral tissue with nodular lymphocyte proliferation comprising of small lymphocytes and some large cells with large and irregular nuclei resembling Reed-Sternberg cells and less frequent plasmocytes and neutrophils. Large lymphocytes and immunoblasts were prominent along with focal proliferation; some fibrotic tissue was also seen. IHC staining showed widespread lymphocytes positive for LCA. Small and large lymphocytes were positive for CD20. Most small lymphocytes were positive for CD3. Both CD20 positive large lymphocytes and CD20 negative Reed-Sternberg cells were positive for CD30.Only some Reed-Sternberg cells were positive for CD15. A diagnosis of Gray zone lymphoma that is a composite lymphoma of Hodgkin and Non-Hodgkin types with large B cells was made. Considering the positive CD20 cells, a salvage treatment of RICE with Rituximab was prescribed. She went to remission.

DISCUSSION

Primary pulmonary Hodgkin lymphoma is a rare disease and usually presents with lung mass or nodules and rarely with endobronchial lesions. A misdiagnosis occurs in about half the patients. It may involve subpleural and parenchymal lymph nodes but it rises from lung parenchyma and does not arise from the mediastinum (4,-6). The world health organization classifies lymphomas with shared cellular forms between unclassified large B cells and classic Hodgkin lymphomas as combined or gray zone lymphoma (7).

In this case, there was no mediastinal or significant lymph node involvement, characteristic for primary pulmonary lymphoma (9). On the pathologic report, CD15 negative cells raised suspicion about Hodgkin disease but Reid-Stenberg cells are positive for CD15 in 75%-85% of cases (8). Composite lymphoma is more common in old ages but it has been reported in patients 26 to 88 years old. In some cases, there is a family history of non-Hodgkin lymphoma (10). Recently, composite lymphoma has been considered as a rare form of lymphoma, which rises from two or more types of lymphomas with the same morphologic and some immunologic features in a single organ. Pathogenesis is complex and uncertain. Multiple types of composite lymphomas include B cells and T cells, Hodgkin and non-Hodgkin, Hodgkin and T cell and B cell lymphomas (10). Anatomical involvement may be nodal or extra-nodal. Gastrointestinal tract, respiratory system, parotid glands, bone and skin may be involved in extra-nodal disease as the common sites (3,11,12). In this case, despite an existing polypoid endobronchial lesion, a pathologic diagnosis was not possible to be made even after taking two bronchoscopic biopsy samples. The disease relapsed after two courses of chemotherapy and it was interesting that it first emerged as an endobronchial lesion in bronchus intermedius. Considering treatment failure, there was a need for re-evaluation of the pathology report and specimens for a curative treatment. A Rituximab-based regimen was curative and the patient has been disease-free so far.

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