Case Report

An Interesting Case of Lung Mass

Venkata Satya Suresh Attili, C Obula Reddy[1]

Department of Medical Oncology, Yashoda Cancer Hospitals, Hyderabad, [1]Department of Pathology, Kidwai Memorial Institute of Oncology, Bangalore-560 029, India

Address for correspondence: Dr. AVS Suresh, Email: sureshattili@yahoo.com

ABSTRACT

A 46-year-male smoker presented with cough and fever to a chest physician, who on the basis of chest X-ray started him on antitubercular treatment. However, further evaluation suggested that the patient was suffering from a rare disease, i.e. Anaplastic large cell lymphoma of lung. Although lung involvement in cases of lymphoma is observed in as high as 40% of cases, in autopsy series, the exact clinical incidence is not known. One of the largest lymphoma groups reported it to be around 25%. However, primary pulmonary lymphomas have been extremely rare (0.4%), and whenever present they are of (Mucosa Associate Lymphoid tissue) MALT type, with occasional diffuse large cell lymphomas. The anaplastic variant is extremely uncommon. Usually the treatment results are satisfactory with more than 80% of the cases surviving even after 3 years. Here we report the case of anaplastic primary nonHodgkin's lymphoma of lung and review the literature.

Keywords: Primary pulmonary lymphoma, treatment, immuno histo chemistry

DOI: 10.4103/0974-2727.54804

INTRODUCTION

Extranodal involvement in the cases of nonHodgkin's lymphoma (NHL) is a known entity with gastrointestinal tract leading the list, followed by skeletal system, skin, kidneys and other rare organs. However, pure extranodal forms (often referred to as primary) are rare events, and the definition often requires either absence of nodes or the primary site should be the largest one. Primary involvement of lung was observed in around 0.4% of all extranodal lymphomas.[1] Wherever reported, MALT-type lymphomas are more common, followed by diffuse large cell variants. Anaplastic variants are the rarest forms and we report here one such rare case and review the literature.

CASE REPORT

A 46-year-old male smoker presented with a history of dry cough and moderate-grade pyrexia, responding to antipyretic since 3 months. History regarding other system involvement did not suggest any other abnormality. There were no other systemic symptoms. Examination revealed decreased breath sounds at the right lung base. There was no lymphadenopathy or organomegaly. With these symptoms, the patient was provisionally diagnosed as a case of tuberculosis and started on the anti-tubercular therapy. He did not show any improvement and was referred for further management. The blood investigations were within normal range and imaging [Figure 1] of the thorax showed a mass lesion in the upper lobe of the right lung and abdominal scans were within normal range. Fine needle aspiration biopsy from the

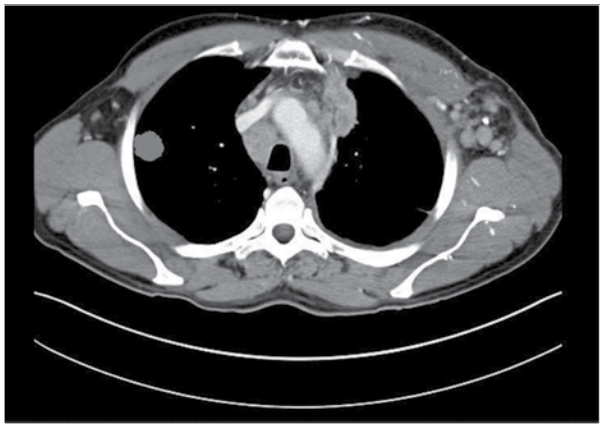

Figure 1: CT scan showing right upper lobe mass

lesion showed atypical cells suggestive of malignancy. For further confirmation, Immunohistochemical (IHC) markers were performed on cell block, which suggested LCA positive, CK (cytokeratin) negative, other markers were inconclusive. The cytogenetics from the lesion suggested t (2 : 5) though Anaplastic Lymphoma Kinase (ALK) was negative. Staging evaluation suggested that it was extranodal lymphoma stage I B E (bone marrow examination showed no involvement). The primary anaplastic large cell lymphoma of lung was the final diagnosis made and patient was started on combinational chemotherapy consisting of cyclophosphamide, adriamycin, vincristine and prednisolone. After six cycles of chemotherapy, the patient is in complete remission.

DISCUSSION

Though lung involvement in cases of lymphoma is observed in as high as 40% of cases, in autopsy series, the exact clinical incidence is not known.[1] One of the largest lymphoma groups reported it to be around 25%. However, Primary Pulmonary Lymphomas (PPLs) are extremely rare (0.4%), and whenever present they are of MALT type, with occasional diffuse large cell lymphomas.[2] It was hypothesized that the high incidence of MALT type of lymphomas in lung are mainly due to the chronic antigenic stimulation (with etiological agents ranging from smoking, autoimmune disorders, infections and occupational exposure to irritants).[1,3] Clinically they often present with incidental diagnosis (asymptomatic) in around 50% of cases, followed by occasional cough and hemoptysis with B symptoms being extremely uncommon.[3] The diagnosis is often delayed and sometimes missed owing to its resemblance with other similar entities like pseudo-lymphoma, lymphoid interstitial pneumonitis and lymphomatoid granulomatosis.[4-8] In such circumstances, IHC features help to differentiate PPL from the above.

One of the largest reported series showed that most of them (54%) are MALT type, followed by diffuse large cell variants (28%) and other rare forms (18%).[3] Primary anaplastic large cell lymphoma is a rare entity among these and to the best of our search; the largest reported series had five cases.[6] Though diagnostic criteria does not exist for this entity, presence of the characters mentioned below are widely accepted.[3]

1. Presence of unilateral or bilateral pulmonary involvement by NHL
2. No evidence of mediastinal adenopathy or extrathoracic disease
3. No past history of lymphoma.

The natural course of this rare disease is usually indolent and responds favorably to treatment, with only few exceptions where the disease is aggressive. Wherever the disease is having an indolent course, usually local modalities like surgery with or without radiation is adequate.[1,3,5,7,8] However, whenever the disease is aggressive, systemic chemotherapy is indicated. Simple alkylating agent based chemotherapy is often enough, with occasional cases requiring anthracyclines. The prognosis is excellent with three-year survival rates of 86%.[3] Pathological type and aggressiveness of the disease at presentation (including duration of symptoms) are usually important prognostic factors and role of Internatinal Prognostic Index [IPI] score in these cases is not studied yet.

REFERENCES

1. Ferraro P, Trastek VF, Adlakha H, Deschamps C, Allen MS, Pairolero PC. Primary non-Hodgkin's lymphoma of the lung. Ann Thorac Surg 2000;69:993-7.
2. Kurtin PJ, Myers JL, Adlakha H, Strickler JG, Lohse C, Pankratz VS, *et al.* Pathologic and clinical features of primary pulmonary extranodal marginal zone B-cell lymphoma of MALT type. Am Surg Pathol 2001;25:997-1008.
3. Kim JH, Lee SH, Park J, Kim HY, Lee SI, Park JO, *et al.* Primary pulmonary non-Hodgkin's lymphoma. Jpn J Clin Oncol 2004;34:510-4.
4. Nicholson AG, Wotherspoon AC, Diss TC, Hansell DM, Du Bois R, Sheppard MN, *et al.* Reactive pulmonary lymphoid disorders. Histopathology 1995;26:405-12.
5. Cadranel J, Wislez M, Antoine M. Primary pulmonary lymphoma. Eur Respir J 2002;20:750-62.
6. Rush WL, Andriko JA, Taubenberger JK, Nelson AM, Abbondanzo SL, Travis WD, *et al.* Primary anaplastic large cell lymphoma of the lung: a clinicopathologic study of five patients. Mod Pathol 2000;13:1285-92.
7. Tian XL, Feng RE, Shi JH, Duan MH, Wang JL, Liu HR, *et al.* Primary pulmonary lymphoma: analysis of 18 cases. Zhonghua Jie He He Hu Xi Za Zhi 2008;31:401-5.
8. Yang SL, Song H, Tang YM. Primary anaplastic large cell lymphoma originated from lung in a case. Zhonghua Er Ke Za Zhi 2007;45:949.

Source of Support: Nil, **Conflict of Interest:** None declared.