Case Report

A 57-Year-Old Woman with Idiopathic Interstitial Lung Disease and Refractory Cough

K. Gowrinath[1], K.R.R. Umamahesh[1] and V. Swetha[2]

Departments of Pulmonary Medicine[1] and Pathology[2], Apollo Speciality Hospital, Nellore (Andhra Pradesh), India

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ABBRIVATIONS USED IN THIS ARTICLE

NHL = Non-Hodgkin's lymphoma

ILD = Interstitial lung disease

CD = Clusters of differentiation

Bcl = B-cell lymphoma

PET-CT = Positron emission tomography-CT

Abstract

Endobronchial non-Hodgkin's lymphoma (NHL) is rare and poses a diagnostic challenge if there is pre-existing lung disease, like idiopathic interstitial lung disease (ILD). We report refractory cough due to isolated endobronchial non-Hodgkin's low grade B-cell lymphoma in a 57-year-old female with ILD. Local diathermy fulguration of endobronchial NHL resulted in remission of cough and there was no recurrence or spread of endobronchial NHL at four years of follow up.

Introduction

Endobronchial non-Hodgkin's lymphoma (NHL) is rare and usually detected as part of widely disseminated disease.[1] In interstitial lung disease (ILD), suspicion of an endobronchial lesion without obstructive radiographic features is difficult and diagnosis may be delayed if new onset symptom, like cough is attributed to progression of pre-existing lung disease or its complications, like secondary infection. NHL in ILD is very rare.[2] We report an unique case of refractory cough due to isolated endobronchial B-cell low grade NHL in an adult female with idiopathic ILD.

Case Report

A 57-year-old married woman presented with dry cough of three months duration with tiredness and occasional wheeze. There was no relief with nebulised bronchodilators, systemic methylprednisolone and antibiotics given at local place. Patient had ILD diagnosed two years back but there was no significant cough. Patient was treated for tuberculous cervical lymphadenitis in childhood. Family history revealed that her mother had asthma. On physical examination, there was no peripheral lymphadenopathy, digital clubbing or pedal oedema. Chest examination showed bilateral inspiratory crackles over lower lobes. Laboratory tools including serology were insignificant. Chest radiograph (postero-anterior view) showed

Corresponding author: Dr K. Gowrinath, Department of Pulmonary Medicine, Apollo Speciality Hospital, Pinakininagar, Nellore-524 004, (Andhra Pradesh), India; E-mail: drkgowrinath@gmail.com

bilateral interstitial opacities. Computed tomography of chest showed eccentric soft tissue mass (1.3cmx0.4cm) within the proximal right main bronchus causing mild luminal narrowing with mediastinal lymphadenopathy (Figure 1). Flexible fiberoptic bronchoscopy showed fleshy growth in the right main bronchial lumen adjacent to the carina (Figure 2). Endobronchial biopsy showed diffuse small lymphocytic infiltration with vascular proliferation (Figure 3). Bone marrow aspiration study was insignificant. Immunoglobulin heavy chain (IGH) clonality analysis was negative. Immunohistochemistry of cell surface markers revealed clusters of differentiation (CD-20) of follicular and interfollicular regions. Other B-cell lymphoma (Bcl) markers, like Bcl2 and Bcl6 were also positive. A diagnosis of low-grade B-cell follicular NHL was made. The whole body positron emission tomography (PET-CT) showed right main bronchial hypermetabolic endobronchial soft tissue and mediastinal lymph nodes. As the low grade B-cell follicular NHL was localised, diathermy fulguration was done. Patient did not have cough later and remained under regular follow-up with yearly PET-CT scan. The CT of chest followed by the whole body FDG PET-CT scan at fourth year of follow-up did not show any recurrence or spread of endobronchial NHL (Figure 4).

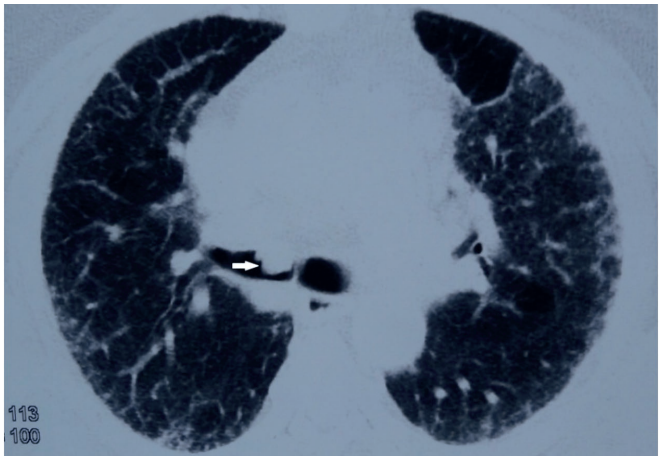

Figure 1. Computed tomography of chest showing an endobronchial tumour (white arrow) within the right main bronchus.

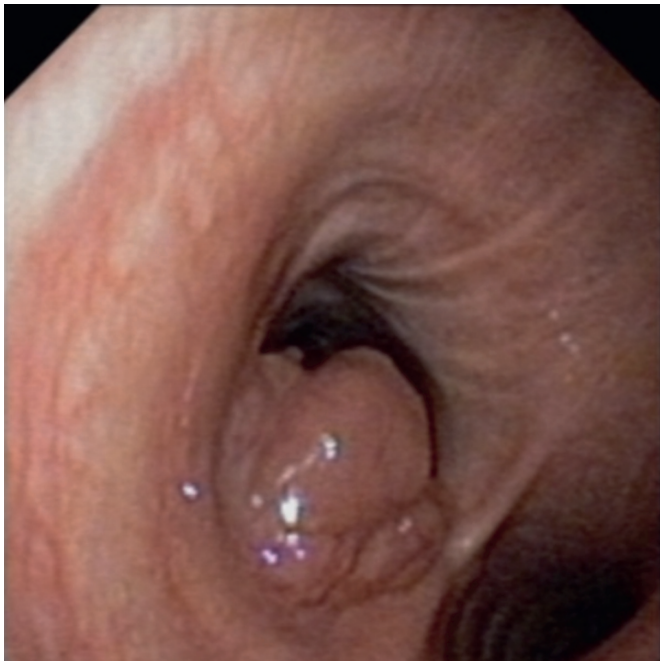

Figure 2. Fleshy growth within the right main bronchus, as seen through flexible bronchoscope.

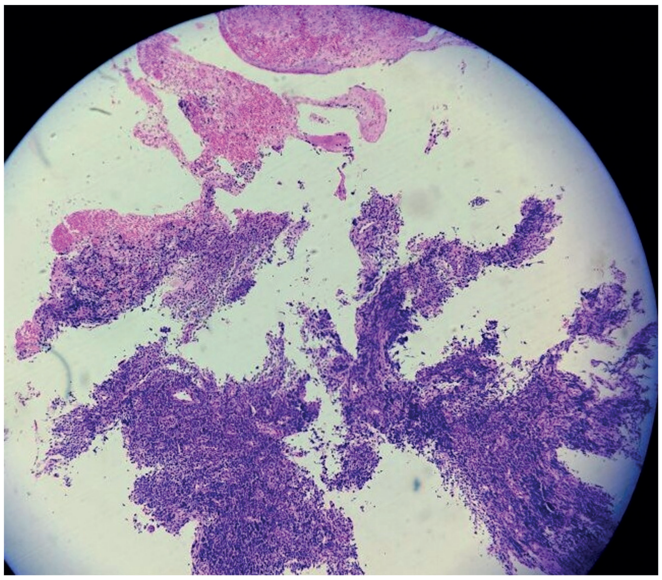

Figure 3. Endobronchial biopsy showing diffuse small mononuclear lymphocytic infiltrate. (H&E, x 4)

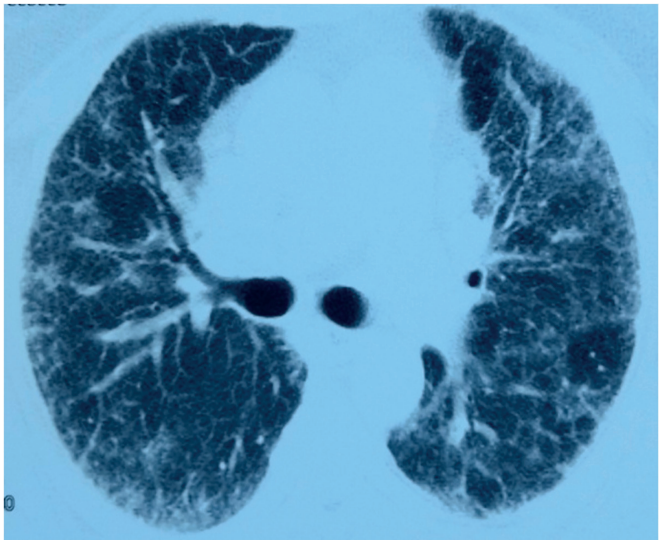

Figure 4. The follow-up CT of chest showing normal right main bronchial lumen.

Discussion

Endobronchial involvement was found in 0.68% cases of NHL and majority of them had obstructive radiologic features.[3] Males are affected more often than women with peak incidence in 6th decade. Cough and wheezing may occur due to airway obstruction by the endobronchial tumour. In our case, diagnosis of endobronchial NHL was delayed as chest radiograph did not show obstructive features and the refractory cough was mistaken as due to progression of pre-existing ILD or secondary infection. In the present case, the endobronchial NHL was Type II as it was localised with mediastinal lymphadenopathy as per earlier classification of endobronchial lymphoma by Rose *et al*.[4] NHL is not a single disease and the specific type must be diagnosed as prognosis and response to treatment varies with each type of lymphoma. An accurate diagnosis of low grade (Grade 1-2) B-cell follicular NHL can be made through morphologic evaluation of hematoxylin and eosin stained tissue section followed by IHC and cytogenetic molecular studies for chromosomal aberrations which have diagnostic, prognostic and therapeutic significance.[5] The neoplastic cells that look alike under the microscope carry different CD markers on their surface and currently the diagnosis of lymphoma always include identification of atleast few of them. In our case, specific diagnosis of low grade B-cell NHL was based upon morphological features and IHC staining. Among the mechanisms of endobronchial involvement of a malignant tumour[6], direct bronchial invasion from adjacent mediastinal lymphadenopathy and lymphatic spread to peribronchial connective tissue was the most likely cause in our case. Very few case reports of isolated endobronchial low grade B-cell NHL are available in the English literature. These include collapse of the right lung in a chronic smoker adult male[7] and an endobronchial NHL at carina with normal chest radiograph which was treated effectively with local irradiation.[8] Rarely, pulmonary B-cell NHL may arise in an area of interstitial pulmonary fibrosis[2] but in our case NHL occurred within the bronchus without involvement of lung parenchyma and we are not aware of any previous similar report in the literature. In our case, occurrence of isolated endobronchial NHL without obstructive radiologic features in pre-existing ILD posed a diagnostic challenge. Initial diagnostic evaluation with CT of chest detected endobronchial lesion and flexible bronchoscopy helped to diagnose NHL and differentiate it from other causes like Hodgkin's lymphoma, carcinoid tumour, primary or secondary carcinoma, inflammatory pseudotumour and infections like fungal or tuberculosis.

Treatment strategy for a localised follicular lymphoma (Ann Arbor stage I-II) is yet to be standardised due to non-availability of well conducted randomised studies but an involved field or involved site radiotherapy is considered to be most appropriate.[9] In our patient, local diathermy fulguration resulted in prompt symptomatic relief without recurrence at four year follow up.

To conclude, CT of chest is the initial study of choice in any case of ILD with refractory cough to detect any endobronchial lesion and long-term remission of symptoms due to an isolated endobronchial non-Hodgkin's low grade B-cell lymphoma is possible with local diathermy fulguration.

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