Endobronchial non-Hodgkin's lymphoma*

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Non-Hodgkin's lymphomas belong to a group of lymphoproliferative malignancies that differ in behavior, treatment and prognostic patterns.

Over the course of the disease, non-Hodgkin's lymphomas may affect the thoracic structures – especially the mediastinum and the pulmonary parenchyma. However endobronchial involvement is extremely uncommon, even in the advanced stages of the disease.

We report a case of non-Hodgkin endobronchial lymphoma and make a review of the literature.

Key words: Lymphoma, non-Hodgkin/complications. Lung neoplasms. Prognosis.

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INTRODUCTION

Non-Hodgkin's lymphomas belong to a heterogeneous group of lymphoproliferative malignancies, the various members of which present differences in behavior, prognosis and recommended treatment. In up to 43% of patients, they may affect the thoracic structures – especially the mediastinum and the pulmonary parenchyma – at any time over the course of the disease. (1) However, endobronchial involvement is extremely uncommon, even in the advanced stages of the disease.

The first report of endobronchial involvement in a case of non-Hodgkin's lymphoma was made by Dawe in 1955. Since then, 55 cases have been reported in the literature.

CASE REPORT

A 24-year-old female non-smoker sought medical attention 3 months after the onset of symptoms, which included dry cough, mild dyspnea, and a weight loss of 3 kg (< 10% of her previous weight). She reported no fever or sweats. Physical examination evidenced decreased vesicular breath sounds in the upper third of the left hemithorax. No palpable enlarged peripheral lymph nodes were found. The patient was submitted to chest X-ray, which revealed an opaque mass in the topography of the upper lobe of the left lung (Figure 1). The subsequent chest computed tomography (CT) scan revealed heterogeneous opacity in the upper two-thirds of the left lung. There was a slight reduction in lung volume and a small pleural effusion on that side. The opacity in the left lung extended across the mediastinum and affected the blood vessels of the cervicothoracic junction and the ipsilateral hilum (Figure 2). The CT scan of the abdomen was normal. Respiratory endoscopy revealed a growing lesion with glossy surface. The lesion was highly vascularized, which caused near total obstruction of the left main bronchus in its most distal portion. There was an opening in the lower lobe bronchus, but it was too small to allow the passage of the fiberoptic bronchoscope (Figure 3). Biopsy of the endobronchial tumor revealed it to be a large B-cell non-Hodgkin's lymphoma. Immunohistochemical tests for chromogranin, synaptophysin and the anti-keratin monoclonal antibodies AE1/AE3 were negative, those for LCA and CD20 were positive, and those for CD3 and CD30 were negative. Laboratory tests revealed that serum lactate dehydrogenase (LDH) was elevated to 1526 U/L (normal range, 240 U/L to 480 U/L). Test results also showed that the patient was suffering from a mild case of anemia:

hemoglobin was 10.4 g/dL (normal range, 11.5 g/dL to 16.4 g/dL); hematocrit was 32.8% (normal range, 36% to 47%); and (first-hour) erythrocyte sedimentation rate was 67 mm/h (normal range, 0 mm/h).

The patient was started on cyclophosphamide, adriamycin, vincristine, and prednisone (CHOP) chemotherapy, which ameliorated the symptoms, eliminating the cough and dyspnea immediately after the first round.

DISCUSSION

Endobronchial involvement is extremely rare in cases of in lymphoma. In 1934, Moolten reported 2 cases of Hodgkin's lymphoma with airway involvement. However, Dawe reported the first case of non-Hodgkin's lymphoma with endobronchial involvement in 1955.

In 1941, Vieta and Crower reported necropsy findings from 55 patients affected by non-Hodgkin's lymphoma and no evidence of endobronchial involvement was found. (8) A similar study by Papaioannau and Watson reported only one endobronchial lesion among 93 cases of primary lymphoma involving the lungs. (9) These studies revealed that endobronchial involvement is very rare in cases of non-Hodgkin's lymphoma.

In patients with non-Hodgkin's lymphoma, endobronchial lesions are most frequently seen in the main bronchi, followed by the lobe bronchi and the trachea. in 9 (30%) of the 31 cases reported by Eng and Sabanathan, involvement of other areas was demonstrated.⁽³⁾

Various mechanisms have been suggested as being responsible for the development of the endobronchial lesion in patients with lymphoma: direct invasion from an adjacent mediastinal or parenchymal lesion, lymphatic dissemination to the peribronchial tissues, and hematogenous dissemination. The most common mechanisms are direct bronchial invasion and hematogenous dissemination. (1)

Rose et al.⁽¹⁾ described two types of non-Hodgkin's lymphoma. Type 1 is characterized by diffuse submucosal nodules in the airways of patients with systemic lymphoma, including considerable lung parenchyma involvement, which may or may not be associated with respiratory symptoms, and rarely showing signs of endobronchial obstruction. Type 2 consists of an endobronchial mass in the central airways. However, in type 2 cases, there is no evidence of systemic lymphoma, although the regional lymph nodes are enlarged. Type 2 is generally associated with symptoms of airway obstruction, such as coughing and wheezing.^(6,10-12)

Patients usually present with nonspecific symptoms. Dyspnea, cough, and wheezing are the most common symptoms, followed by hemoptysis. Asymptomatic patients are rare. (10-15)

In patients with endobronchial involvement, chest X-rays reveal atelectasis in 50% of cases and hilar mass in 20%. (3,16,17) Diagnosis is defined by fiber bronchoscopy followed by the biopsy of the lesion.

Treatment depends on the extent of the disease, as well as on the condition of the patient. Surgical procedures are seldom recommended. Chemotherapy, either alone or in combination with radiotherapy, is the treatment of choice.

Median survival time varies from (a maximum of) 13 months in patients with the generalized form of the disease (type 1) to approximately 3 years in patients with the localized form (type 2).⁽³⁾

The clinical severity of malignant endobronchial lesions is correlated with the possibility of bronchial obstruction and subsequent pulmonary collapse. Through clinical and radiological evidence alone, it is not possible to distinguish malignant endobronchial lesions from other endobronchial diseases such as carcinoid tumor, carcinoma, and lymphoma.

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Figure 1 – Chest X-ray with opaque mass in the upper Lobe of the left lung

Figure 2 – Computer assisted tomography of the chest with heterogeneous opacity in the upper 2/3 of the left lung

Figure 3 – Lesion growing in the left main bronchus