CASE REPORT

Primary anaplastic large cell lymphoma of the lung presenting with acute atelectasis

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Abstract

Non-Hodgkin lymphoma only rarely occurs as a primary lung mass. We report a very rare case of primary pulmonary anaplastic large cell lymphoma presenting with acute atelectasis in a 55-year-old man. Chest computed tomography revealed a consolidated central mass in the left lung with obstructive pneumonia that had developed into total atelectasis. After a bronchoscopic examination failed to yield a definite diagnosis, supraclavicular lymph node biopsy was performed, revealing an anaplastic large cell lymphoma. This case illustrates the need for rapidly locating a possible biopsy site, other than the primary lung mass itself, and the value of empirical steroid treatment for avoiding devastating exacerbation when aggressive pulmonary lymphoma is suspected.

Introduction

Extranodal non-Hodgkin lymphoma (NHL) most frequently occurs in the gastrointestinal tract, and is rarely found in the lung. Lymphomas occur in the lung in only 0.4% of all cases,¹ and NHLs occur in the lung in only 0.3% of cases.² The most common primary NHL in the lung is marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue type (MALT lymphoma),^{1,3,4} whereas anaplastic large cell lymphomas (ALCLs) mainly involve the lymph nodes and skin, and only very rarely occur in the lung. Kim *et al.* reported 26 cases of primary NHL in the lung between September 1995 and June 2003, of which only two cases were of ALCL.⁵ Indeed, to our knowledge, only eight cases of primary pulmonary ALCL in adult patients have ever been reported. Here, we describe a very rare case of primary ALCL in a patient who made a partial recovery from total atelectasis of the left lung.

Case report

A 55-year-old male was admitted to our center after presenting with a cough, yellow sputum, and night sweating for more than a month. Initial chest radiography revealed a large, dense opacity in the left upper and lower lung, suggesting malignancy. Subsequent chest computed tomography (CT)

Figure 1 An initial chest computed tomography (CT) scan reveals a consolidated central mass in the left lung with surrounding ground-glass attenuation. Multiple enlarged lymph nodes are also present in the left supraclavicular area and the mediastinum, suggesting lung cancer with multiple lymph node metastases causing obstructive pneumonia.

Figure 2 After exacerbation, a chest radiography film reveals total atelectasis of the left lung (left). Positron emission tomography (PET) revealed a huge malignant mass accompanied by pleural seeding in left thorax and multiple metastatic lymph nodes with intense fludeoxyglucose (FDG) uptake (right).

showed a consolidated central mass in the left lung with surrounding ground-glass attenuation and multiple enlarged lymph nodes in the left supraclavicular area and mediastinum. These findings were also suggestive of lung cancer, together with multiple lymph node metastases causing obstructive pneumonia (Fig. 1). We started empirical antibiotic treatment for possible pneumonia and performed a transbronchial lung biopsy (TBLB) of the obstructing endobronchial lesion, because TBLB is considered to be a safer and more convenient procedure than lymph node excision in our center. Pathological analysis of the TBLB revealed diffuse infiltration of T lymphocytes, but no obvious evidence of malignancy was noted. We, therefore, performed an excision biopsy of the left supraclavicular lymph node, together with a repeat TBLB of the primary lung mass. While waiting for the biopsy result, the patient's condition deteriorated, and chest radiography showed total atelectasis of the left lung (Fig. 2). After acute exacerbation of the lung lesion, we immediately

Figure 3 Photomicrographs show total effacement of nodular architecture (a. hematoxylin and eosin [H&E] staining, $\times 10$) and diffuse infiltration of large anaplastic cells with frequent mitosis (b. H&E staining, $\times 400$). The tumor cells were positive for CD30 (c. immunohistochemistry, $\times 400$) and destructing the lung parenchyma (d. H&E staining, $\times 40$).

Figure 4 A chest radiography film taken on day 13 of the first cycle of chemotherapy shows that atelectasis of left lung was almost resolved.

administered methyl prednisolone for suspected malignant lymphoma based on the presence of T-cell rich atypical inflammation in the original TBLB histology. Total atelectasis gradually improved over the next three days. Pathological analysis of the lymph node revealed almost total effacement of nodular architecture with diffuse infiltration of large anaplastic cells, which were positive for CD3 and CD30. CD15 and anaplastic lymphoma kinase (ALK) were negative in immunohistochemistry (Fig. 3). These findings are compatible with ALCL. Analysis of the second TBLB yielded similar results to that of the first, except that positive CD30 staining was also noted. Cyclophosphamide, doxorubicin, vincristine, and prednisolone (CHOP) chemotherapy was immediately started, and at day 13 of the first cycle, the atelectasis of left lung was found to have almost resolved (Fig. 4). CT performed after three cycles of CHOP chemotherapy revealed that the tumor in the left lung was much reduced and the patient showed a partial response at the time of this report. Further cycles of chemotherapy are scheduled with the aim of achieving a complete response.

Discussion

Here, we have described a case of ALCL in which a delay between TBLB and a definite diagnosis by LN biopsy was unavoidable because we had to wait for the results of the first TBLB. Only a few weeks after hospitalization, the patient had developed a total atelectasis of the left lung, which could have been fatal if the mass was taken to be a primary lung carcinoma, and empirical steroid treatment to reduce the tumor burden had not been started.

ALCL lymphoma consistently expresses the CD30 antigen, usually has a T-cell phenotype, and frequently follows an aggressive clinical course.^{6,7} It can be classified on the basis of being positive or negative for ALK rearrangement, representing 60% and 40% of cases, respectively. The most common rearrangement is caused by a translocation of the ALK gene on chromosome 2 and the nucleophosmin gene on chromosome 5, t(2;5).^{8,9} ALK rearrangement is an important prognostic factor in CD30-positive ALCL, with ALK rearrangement-negative ALCL having a relatively poor prognosis.⁹

The standard first-line chemotherapy for ALCL usually consists of an anthracycline-based combination regimen, such as CHOP.¹⁰ To our knowledge, only eight adult cases of primary pulmonary ALCL have ever been reported. Among them, Zhang *et al.* reported total atelectasis of the left lung, similar to that which occurred in our case, and which was also caused by primary pulmonary ALCL.¹¹ Barthwal *et al.* described ALCL in a 23-year-old male patient who presented with a collapsed left lung.¹² It is particularly noteworthy that diagnosis is difficult in many cases because of the relatively low tissue yield from bronchoscopic biopsy in primary pulmonary lymphoma patients.¹ Yang *et al.* reviewed 11 cases of primary pulmonary ALCL lymphoma, including pediatric patients, and only one case was diagnosed on the basis of a bronchoscopic biopsy.¹³ Because of the initial indications of lung carcinoma in our case, we tried TBLB twice, but failed to obtain a definite diagnosis. ALCL was confirmed subsequently only by excision biopsy of the left supraclavicular lymph node. Based on our experience with this case, we recommend rapid identification of a possible biopsy site other than the primary lung mass itself. If indicated, empirical steroid treatment could prevent lymphoma-induced acute aggravation.

Disclosure

This work has not been submitted for publication elsewhere except as a brief abstract in the proceedings of a scientific meeting or symposium. No authors report any conflict of interest.

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