High-Grade Primary Pulmonary B Cell Lymphoma Presenting as a Necrotic Mass

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Introduction

High-grade primary pulmonary B cell lymphoma is a rare, aggressive lung malignancy, accounting for less than 0.2% of all primary lung cancers.[1] It often presents as a solitary mass with associated systemic symptoms, most commonly in immunosuppressed patients. We present a case of this unusual malignancy that presented as a necrotic mass in an immunocompetent individual.

Case Summary

A 68-year-old male was admitted for evaluation of a necrotic lung mass found incidentally on an abdominal computed tomogram (CT) performed for evaluation of weight loss and poor appetite (Fig. 1). A subsequent thoracic CT revealed a 10-cm thick-walled right-lower-lobe necrotic mass without central airway involvement. The patient reported several weeks of nonproductive cough, dyspnea, and an 8-kg weight loss. His medical history was notable for depression, gastroesophageal reflux disease, hyperlipidemia, chronic low back pain, and benign prostatic hypertrophy. Past surgeries included a cholecystectomy and bilateral inguinal hernia repairs. His medications included simvastatin, omeprazole, and tramadol. He had never smoked and denied use of alcohol or illicit drugs.

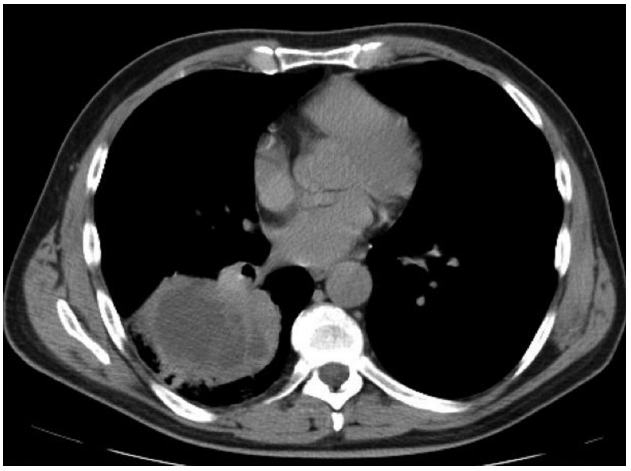

Fig. 1. Computed tomogram shows a right-lower-lobe necrotic mass at presentation.

On physical examination he was afebrile and had normal vital signs, except for an oxygen saturation of 92% on room air. He was thin and ill-appearing. His cardiac and abdominal examinations were unremarkable. Pulmonary examination revealed decreased breath sounds at the right lung base. He had no palpable lymphadenopathy.

Laboratory test results were notable for a white-blood-cell count of 8.3×10^9 cells/L, an erythrocyte sedimentation rate of 59 mm/h, and C-reactive protein of > 100 mg/L. Lactate dehydrogenase was normal at 149 IU/L. His renal and liver panels were remarkable for hypoalbuminemia (26 g/L). Human immunodeficiency virus test was negative. Immunoglobulins, anti-nuclear antibody, anti-neutrophil cytoplasmic antibody, and rheumatoid factor were all within normal limits. CT-guided needle biopsy of the mass revealed only necro-inflammatory debris. A tentative diagnosis of lung abscess was made, and the patient was discharged home on moxifloxacin. A CT 6 weeks later revealed substantial growth of the mass, to 13 cm, and new evidence of airway obstruction (Fig. 2). Bronchoscopy found that the bronchus intermedius was

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The authors have disclosed no conflicts of interest.

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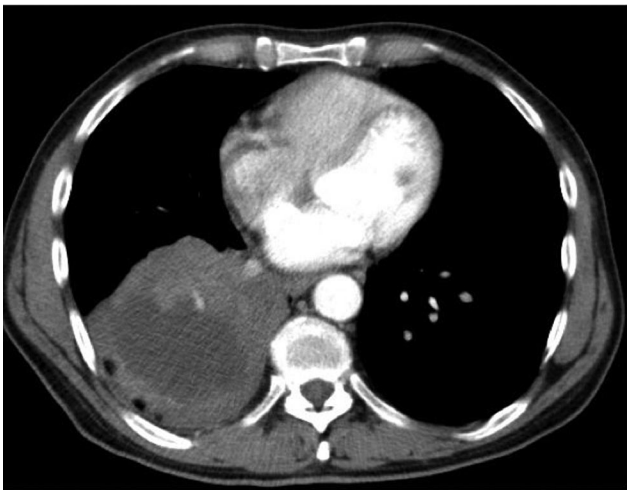

Fig. 2. Computed tomogram 6 weeks after presentation reveals substantial growth of the mass.

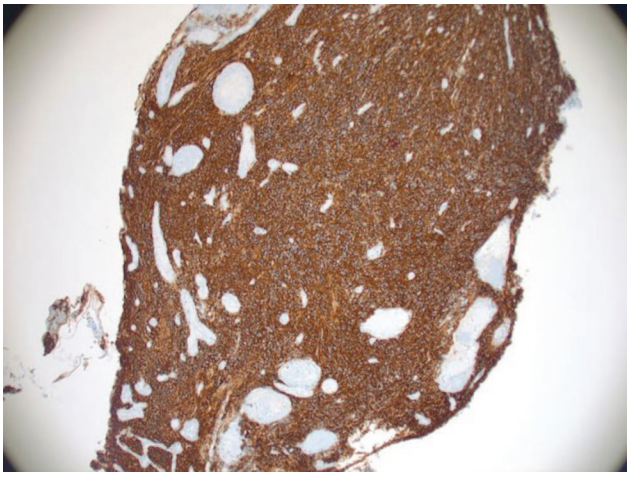

Fig. 3. Endobronchial biopsy shows infiltration of CD20-positive cells.

obstructed, and endobronchial biopsies of that area (Fig. 3) revealed infiltration of large, highly mitotic CD20+ lymphocytes, consistent with a high-grade B cell lymphoma. Given the size of the mass, substantial necrosis, and concern about concurrent pulmonary abscess, lobectomy was performed (Fig. 4). Bone-marrow biopsy found no evidence of disease. Based on the pathology specimen and the absence of disease outside of the lung, the diagnosis was high-grade pulmonary B cell lymphoma.

He recovered from lobectomy and was initiated on R-CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone, and rituximab). Prior to the chemotherapy, positron emission tomography (3 months after the initial CT) found no evidence of disease outside of the right lung, which confirmed the diagnosis of primary pulmonary B cell lymphoma. He had complete remission with his initial chemotherapy, continues to do well, and is being followed

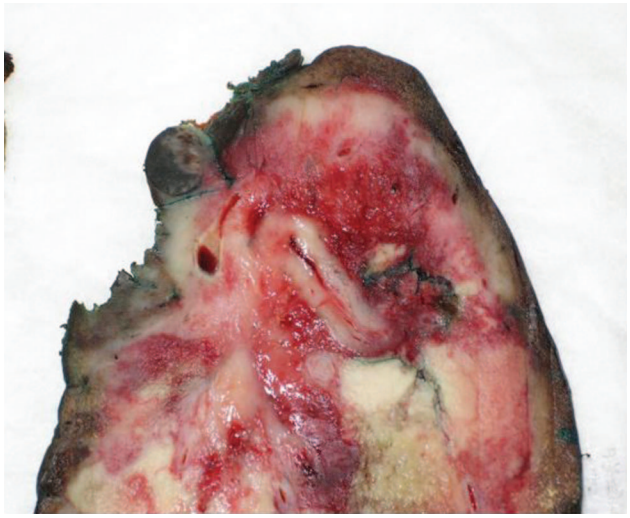

Fig. 4. Gross pathology specimen from right-lower lobectomy shows hepatization of lung tissue.

with serial positron emission tomography to monitor for recurrence.

Discussion

Lymphoma can develop in the pulmonary parenchyma via 3 mechanisms: hematogenous spread from other organs; direct invasion from contiguous mediastinal or hilar lymph nodes that are the primary disease sites; or primary lung involvement.[2] Primary pulmonary B cell lymphoma is rare, accounting for only 0.5–1% of all lung malignancies and less than 1% of non-Hodgkin lymphomas. Primary pulmonary B cell lymphoma is defined by clonal lymphoid proliferation affecting one or both lungs in patients with no detectable extrapulmonary involvement at diagnosis or during the subsequent 3 months. The majority of cases are mucosa-associated lymphoid-tissue lymphoma, which is a low-grade form of B cell lymphoma. High-grade pulmonary B cell lymphoma is much less common, accounting for only 11–19% of cases of primary pulmonary B cell lymphoma. High-grade pulmonary B cell lymphoma generally occurs in patients with immunodeficiency, such as from human immunodeficiency virus or solid-organ transplantation.[1] Our patient underwent evaluation for rheumatologic and immunosuppressive conditions, but none were found.

Patients with high-grade pulmonary B cell lymphoma may be asymptomatic but often report respiratory symptoms, fever, or weight loss. The mean age of presentation in patients who do not have human immunodeficiency virus is 60 years, but cases have been reported from age 30 to 82 years.[1,2] Radiologic studies typically show a single pulmonary mass, often with associated atelectasis, although multiple masses have been reported.[1,3] Cavitation

and necrosis rarely occur and may be mistaken for lung abscess, non-small-cell lung cancer, metastatic disease, atypical infection (mycobacterial, fungal, actinomyces, nocardia), or anti-neutrophil cytoplasmic-antibody-associated vasculitis.[4-6] Pleural effusion is often present.[1] Endoscopy is frequently abnormal, due to tumoral studding and airway stenosis.[1]

Tissue samples from patients with high-grade pulmonary B cell lymphoma demonstrate strong CD20 positivity and high mitotic activity. These characteristic features often allow the diagnosis to be established with small endobronchial, transbronchial, or transthoracic tissue samples.[1] If bronchoscopy or transthoracic biopsies fail to establish the diagnosis, surgical biopsy should be pursued. Once the diagnosis of lymphoma is established, evaluation for extra-pulmonary disease should be undertaken. Recommended diagnostic studies include bone-marrow biopsy and CT of the abdomen, head, and neck. Positron emission tomography is positive in $>$ 90% of cases of aggressive lymphoma, and therefore is useful for both diagnosis and staging.[7]

No current consensus exists on optimal therapy for high-grade pulmonary B cell lymphoma. Favorable outcomes were reported with surgical resection followed by chemotherapy in small series, but current clinical practice guidelines do not recommend surgical resection strictly for debulking.[8,9] Patients with high-grade pulmonary B cell lymphoma require aggressive chemotherapy, and the first-line chemotherapy regimens are CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) and CHOP-like regimens.[9] The efficacy of rituximab for high-grade pulmonary B cell lymphoma is not well established in primary pulmonary lymphoma, but given the established survival benefit in diffuse large B cell lymphoma outside the lung, the addition of rituximab to initial chemotherapy for high-grade pulmonary B cell lymphoma is reasonable.[10] Overall survival is worse in patients with high-grade pulmonary B cell lymphoma, compared to low-grade primary pulmonary B cell lymphoma; the respective median survival times are 8–10 years and $>$ 10 years. Survival time is poorly defined but appears to be much shorter for immunocompromised patients.

Teaching Points

Primary pulmonary lymphoma is a clonal lymphoid proliferation within one or both lungs, without detectable extrapulmonary involvement at the time of diagnosis or in the subsequent 3 months. It is rare, accounting for only 0.5–1% of primary lung malignancies. High-grade primary pulmonary B cell lymphoma is an uncommon, aggressive form that may present as a solitary mass, potentially with necrosis or cavitation. These masses can be mistaken for abscess, non-small-cell lung cancer, or anti-neutrophil cytoplasmic-antibody-associated vasculitis. Systemic chemotherapy with CHOP or a similar regimen is the mainstay of therapy for high-grade primary pulmonary B cell lymphoma. Although often performed, the role of surgical resection is not well defined. The median survival of high-grade primary pulmonary B cell lymphoma is 8–10 years, although it is much shorter in immunocompromised patients.

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