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Unraveling the Complex Presentation of Pulmonary Lymphomatoid Granulomatosis: A Case Study

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ABSTRACT

Lymphomatoid granulomatosis (LYG) is an uncommon lymphoproliferative disorder driven by the Epstein-Barr virus (EBV). This rare condition typically exhibits a progressive clinical course, primarily affecting the lungs. LYG is characterized by the presence of bilateral pulmonary nodules and is pathologically identified by the infiltration of atypical EBV-positive B-lymphocytes, accompanied by reactive T-lymphocytes, with a distinctive angiocentric and angiodestructive pattern. Here, we present a unique case of pulmonary LYG in an 81-year-old female, manifesting as a large mass leading to the complete occlusion of the right main stem bronchus intermedius.

Keywords: Lymphomatoid Granulomatosis; Pulmonary Mass; Angiocentric and Angiodestructive Infiltration; Bilateral Pulmonary Nodules.

INTRODUCTION

Lymphomatoid granulomatosis, an uncommon lymphoproliferative disorder linked to the Epstein-Barr virus (EBV) [1], typically manifests in the lungs, predominantly in males. Its standard radiological appearance includes multiple bilateral pulmonary nodules [2].

The lungs are the primary site of involvement, with additional pulmonary locations such as the skin and brain [3]. Recent studies have shown that all clinically diagnosed Lymphomatoid granulomatosis cases exhibited lung involvement (100%) and tested positive for herpes virus infection (100%) [4]. The disease's development is hypothesized to be linked to an inadequate immune response to the Epstein-Barr virus.

Histologically, Lymphomatoid granulomatosis lesions exhibit angiocentricity, a variably polymorphous

lymphocytic infiltrate, and large, abnormal EBV-positive B cells. Notable vascular changes occur, characterized by lymphocytic inflammation infiltrating the vessel walls. The compromised vessel integrity often leads to extensive necrotic areas, mistakenly referred to as "granulomatosis." This condition is invariably associated with Epstein-Barr virus infection. Lesion grading is based on morphological features and the number of EBV-positive B cells [4]. The proportion of large EBV-positive neoplastic B-cells to mixed reactive T-cells determines the grade of lymphomatoid granulomatosis. Differentiating Grade 3 from Grades 1 and 2 is crucial, as treatment decisions are guided by the disease grade [1]. Grade 1 lesions comprise reactive T-lymphocytes with occasional large EBV-positive neoplastic cells (less than five per high-power field). Grade 2 lesions feature sporadic clusters of large neoplastic EBV-positive cells (rarely up to fifty per high-power field). Grade 3 lesions are characterized by a minimally polymorphous reactive lymphocytic background with readily identifiable large abnormal EBV-positive neoplastic cells. Grade 3 lesions typically exhibit extensive areas of tissue necrosis.

In rare instances, Lymphomatoid granulomatosis presents as a solitary lung mass. Here, we document an exceptional radiological presentation of pulmonary lymphomatoid granulomatosis, appearing as a single mass obstructing the right main stem bronchus [5].

CASE REPORT

An 81-year-old female was admitted to the emergency department complaining of shortness of breath. A CT scan of the chest with contrast revealed a large mass in the right upper lobe, obstructing the right upper lobe bronchus and causing complete collapse of the right upper lobe. The mass had invaded the bronchus intermedius and extended into the mediastinum, as indicated in Figure 1. A PET scan showed high metabolic activity in the right upper lobe mass with mediastinal invasion, confirming the aggressive nature of the lesion.

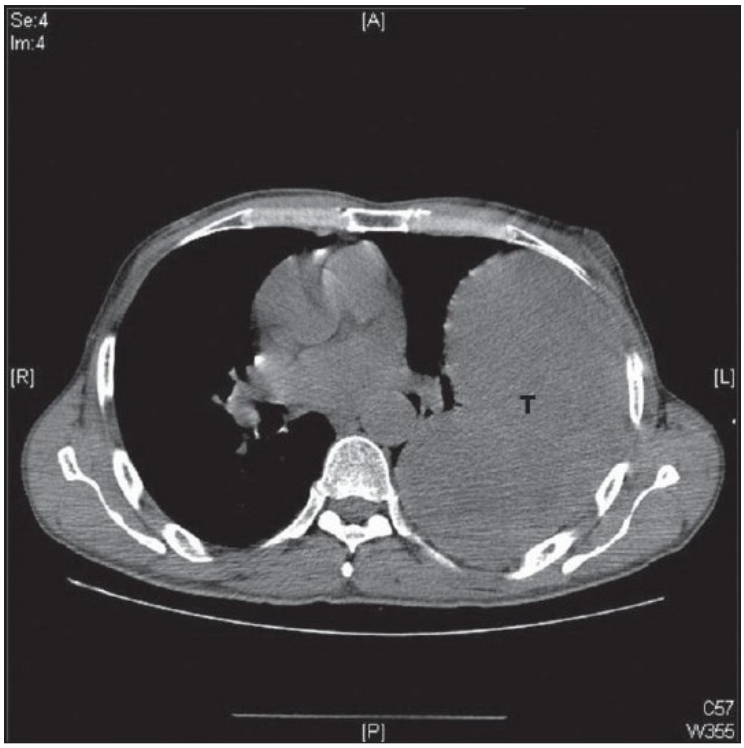

Figure 1: Non contrast CT chest showing right upper lobe mass with endobronchial invasion of right upper lobe bronchus and associated complete distal post obstructive collapse of right upper lobe.

The patient underwent an endobronchial ultrasound-guided cryobiopsy (EBUS) and airway recanalization procedure. Histopathological analysis revealed a prominent angiocentric and angiodestructive inflammatory

infiltrate, comprising small lymphocytes and readily identifiable large atypical lymphocytes. Extensive areas of necrosis were also observed. Immunohistochemical staining for CD3 highlighted various reactive small T-lymphocytes, while staining for CD20 revealed numerous atypical large B cells. The Ki67 expression was high, indicating a high proliferation rate in both the atypical lymphocytes. Epstein-Barr virus-encoded RNA (EBER) in situ hybridization was positive in the large atypical lymphocytes. These findings were consistent with grade 3 lymphomatoid granulomatosis.

A follow-up CT scan was performed after the EBUS biopsy to assess the effectiveness of the airway recanalization procedure. The patient received treatment with dose-adjusted CHOP chemotherapy and two doses of Rituximab, administered at 700 mg and 1400 mg over a period of time. She was discharged from the hospital eleven days after the initiation of treatment. The patient is currently being monitored closely as part of her ongoing care.

DISCUSSION

Lymphomatoid granulomatosis (LYG) is a rare and aggressive B-cell lymphoproliferative disorder associated with the Epstein-Barr virus (EBV) and was first described more than four decades ago. Primarily affecting the lungs, LYG commonly afflicts adult males. It is characterized by the presence of large atypical EBV-positive B cells intermingled with reactive T cells, exhibiting an angiocentric distribution and lymphocytic infiltration. The disease manifests as a spectrum of histological grades, determined by the quantity of EBV-positive B cells. Accurate grading is crucial as treatment decisions and clinical prognosis depend on it. Grade 1 and 2 lesions exhibit an increasing number of EBV-positive B cells (occasionally up to fifty per high-power field) within a polymorphous T cell-rich lymphocytic background. Grade 3 lesions feature abundant EBV-positive B cells mixed with reactive T cells. Notably, lesions with a uniform population of large atypical EBV-positive B cells without a polymorphous background should be diagnosed as Diffuse Large B-cell Lymphoma, Not Otherwise Specified (DLBCL NOS).

Radiologically, LYG typically presents as bilateral pulmonary nodules [2]. Although rare, there are reported cases of pulmonary LYG presenting as a solitary mass or mimicking a lung tumor. Grade 3 LYG lesions often exhibit extensive necrotic areas, leading to a misdiagnosis of malignancy based on imaging. Surgical resections of unilateral pulmonary masses resembling malignancies on imaging have been documented. Thus, it is imperative to consider LYG in the differential diagnosis when evaluating a solitary lung mass radiographically.

Histopathological examination of tissue remains pivotal for an accurate diagnosis. Lesions rich in lymphocytes can mimic malignancy; therefore, endobronchial biopsy or transthoracic needle biopsies are preferred methods to obtain an accurate diagnosis of lymphomatoid granulomatosis.

The treatment approach for LYG involves a combination of steroids, cyclophosphamide, interferon-alpha-2b, and anti-CD20 antibody therapy. The clinical course of LYG varies, ranging from an indolent process to an aggressive large B-cell lymphoma. Prognosis is often grim, with up to 64% of patients succumbing to the disease within the first year of diagnosis. More recently, chemoimmunotherapy regimens incorporating CHOP and Rituximab, along with antiviral agents, have shown promising results, leading to a 5-year overall survival rate of 70%.

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