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Lymphomatoid Granulomatous: Pulmonary Manifestations Of A Rare Entity

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Abstract

Background: *Lymphomatoid granulomatosis (LG) is a rare lymphoproliferative disease predominantly affecting extranodal sites, characterized by its angiocentric lymphoma nature and a notable affinity for blood vessels.*
Objective: *To highlight the clinical and radiographic features of LG, emphasizing its primary pulmonary involvement.*
Methods: *A case report of a 69-year-old male presenting with distinct radiographic findings via conventional chest radiography (XR) and computed tomography (CT).*
Results: *The patient exhibited multiple bilateral cavitated nodular lesions in both XR and CT scans. Despite the diagnostic challenges and a grim clinical course, the disease was confirmed as LG post-mortem.*
Conclusion: *Recognizing LG's distinctive pulmonary manifestations within an appropriate clinical context is pivotal for early diagnosis and intervention, albeit with a median survival of approximately 2 years*

Keywords: *Lymphomatoid granulomatosis. Bilateral pulmonary cavities. RX. Ct. RX. Ct.*

INTRODUCTION

GL was first described by Liebow et al. in 1972 as an angiocentric and angiodestructive lymphoreticular proliferative disease that mainly affects the lung. These authors noted the pathologic similarities between

Wegener's granulomatosis and lymphoma; Despite this, certain differences in the natural history and the clinical and radiological expressions suggested that it could be a separate entity (Aromolo, Simeoli et al. 2023). Recent studies suggest that its pathogenesis is related to Epstein-Barr virus (EBV) infection and is frequently associated with states of immunosuppression. [(1)]. Histologically, it is characterised by an atypical and polymorphous lymphoreticular infiltrate, with marked involvement of the blood vessels and variable degrees of necrosis. [(1)]. Its clinical course is variable. It has a slight predominance in men and is more frequent between the 4th and 6th decades of life. Although remissions have been described (Derbyshire, Fornelli et al. 2023)

CASE PRESENTATION

A 69-year-old male patient who consulted for symptoms of dyspnea, dry cough, hemotoxic sputum, and fever of approximately 20 days of evolution. He presented marked deterioration in his general condition, asthenia, and a loss of approximately 15 kg in the last month. Chest X-ray on admission (Fig. 1) revealed bilateral alveolar-type infiltrates and a cavitated image in the left lung midfield. The initial symptoms were interpreted and treated as severe pneumonia (Derbyshire, Fornelli et al. 2023)

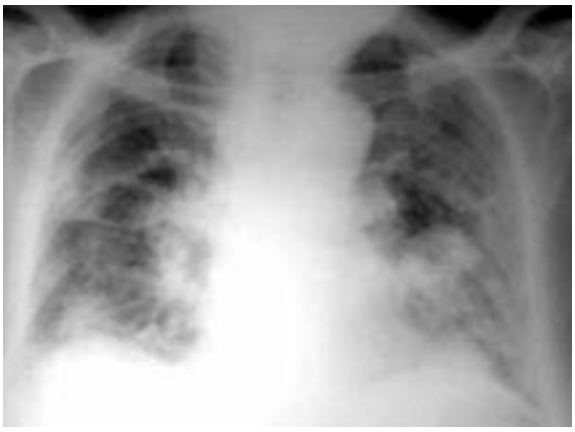

Fig.1. *Chest X-ray on admission. Alveolar-type, bibasal opacities, with a cavitated image in the left lung midfield. The painting was interpreted as severe pneumonia.*

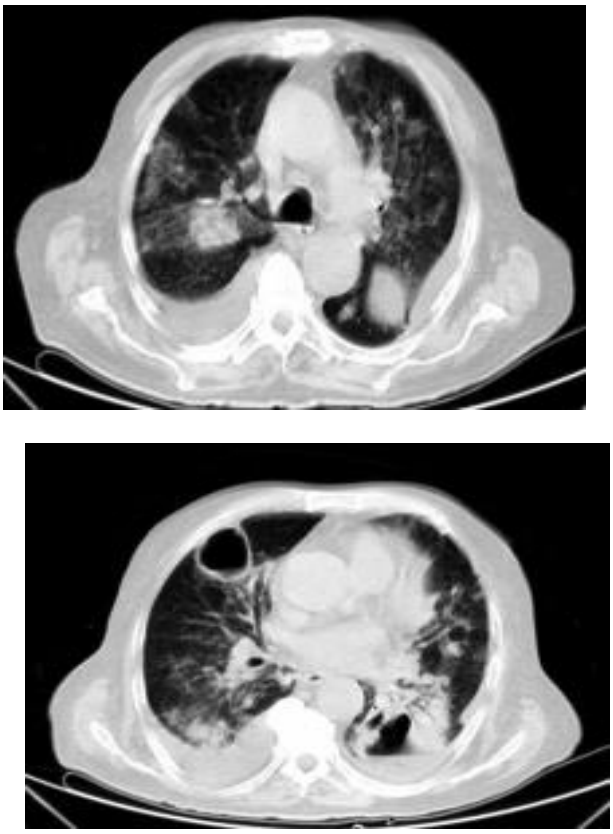

Fig.2. *Chest X-ray (11th day of hospitalisation). Progression with new inÀfilters; nodular images with a solid appearance and others partially cavitated.*

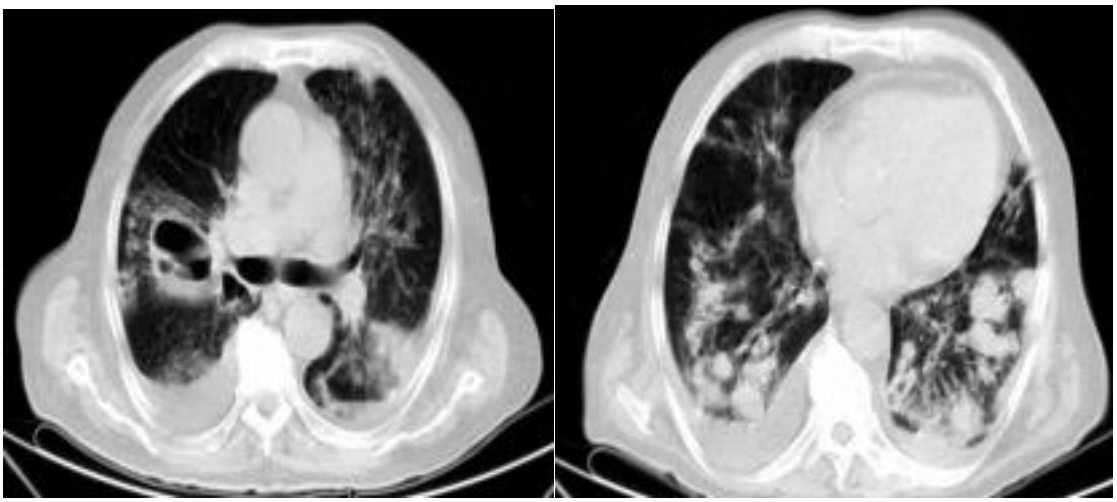

Fig.3. *a,b,c and d. CT with window for lung parenchyma. Multiple nodular opacities, some of them cavitated. It is observed, in addition plus, a focus of consolidation in the left lower lobe and bilateral pleural effusion, predominantly right intracisternal.*

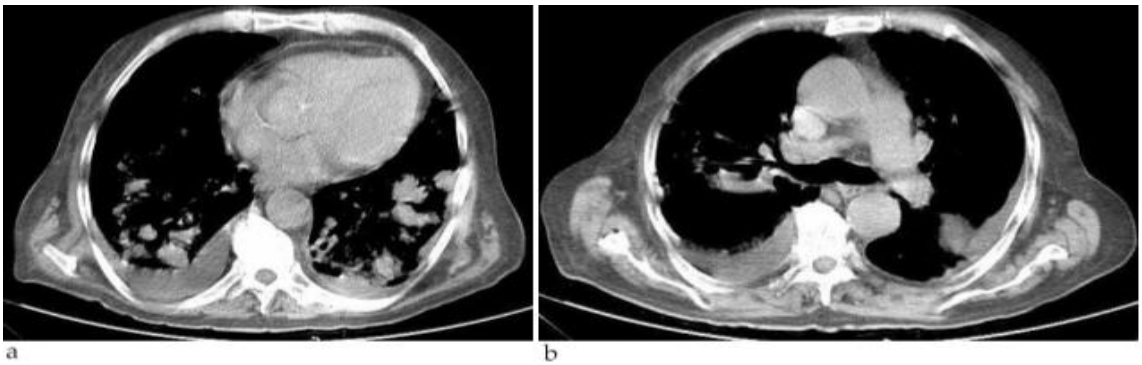

Fig.4.*a and b.CT with a window for mediastinum. Bilateral pleural effusion with left fissural involvement and multiple nodular opacities. Mediastinal lymph nodes are not visualised.*

Tachypneic and febrile, appearing in successive chest X-rays, new infiltrates, nodular images with a solid appearance and others partially cavitated (Fig. 2). HE requested a CT scan (Figs. 3 and 4). The patient remained febrile and in poor general condition despite multiple antibiotic regimens. The disease was progressive, with worsening respiratory failure and death of the patient 45 days after admission. The definitive diagnosis of GL was obtained using a sample of lung parenchyma obtained by autopsy (Girard 2023).

DISCUSSION

GL is a rare lymphoproliferative disease that affects extranodal sites, mainly the lung. It occurs most commonly between the 4th and 6th decades of life, with a slight predominance in men (ratio h:m 2:1) and association with states of immunodeÀciency (Tehzeeb, Divilov et al. 2023). Recent studies relate its development to Epstein-Barr virus (EBV) infection due to its presence in atypical B lymphocytes in most patients with GL[(4)]. Histologically, it is characterised by an atypical lymphoreticular infiltrate with marked infiltration of the blood vessel wall, which can reach a magnitude sufficient to obstruct the vascular lumen. (Kobets, Ahmad et al. 2023)

It is classified into different groups, according to the degree of cellular atypia and necrosis titular, being able to progress to high-grade lymphoma in 13 to 47% of patients· Although the distribution of the disease is multi organic, the most frequently affected site is the lung, presenting at the time of diagnosis with respiratory symptoms (dyspnea, cough, chest pain) and symptoms of general repercussion (fever, depression). to weight loss)Going spontaneous remissions, most present a more aggressive evolutionary course, with disease and progressive respiratory failure[(3)]. Over 60% of patients die within 5 years, with a median survival of 14 months. [(3)]. Radiographic findings include multiple rounded opacities (80%) and diffuse reticulono-dullary infiltrates (Pina-Oviedo, Shroff et al. 2023). Regarding the former, they can conÁuy and Áuctuate in size and deÀnition in successive studies. Cavitation of the same has been described in 20% of cases. Other manifestations, although rare, include hilar lymph nodes and pleural effusions. [(7)]. (Quintanilla-Martinez, Swerdlow et al. 2023)

Differential radiological diagnoses include Wegener's granulomatosis, solid tumour metastasis, sarcoidosis, cryptogenic organising pneumonia, malignant lymphoma, and pseudolymphoma. [8]. In addition, we must consider also consider entities of infectious aetiology, necrotising types of pneumonia, and infection by Your tuberculosis, Pneumocystis carinii, y Aspergillus (So, Shachi et al. 2023). LG does not present characteristic laboratory abnormalities, and its diagnosis is only confirmed after a microscopic examination of a tissue biopsy. Open lung biopsy or video-assisted thoracoscopy is generally required for the methods chosen for taking a biopsy. In cases with skin involvement, taking a bishop whether it simple . The treatment of the disease is still controlled. Spill (Singh, Singh et al. 2023). Cyclophosphamide and prednisone may be useful in those patients with diseases limited to the lung and skin and low histological grade. Multiagent chemotherapy is used in patients with systemic symptoms, involvement beyond the lung and skin, and high histologic grade. Other possible treatments include surgical resection followed by chemotherapy in patients with localised disease and antiviral treatments, such as interferon alfa-2b and ganciclovir, the latter6 in experimentation (Razack, Cariem et al. 2023).

CONCLUSIONS

Lymphomatoid granulomatosis (LG) presents as a rare and aggressive lymphoproliferative disease with a notable predilection for the lungs. While the disease's clinical course is unpredictable, early recognition of its unique pulmonary manifestations is crucial for timely diagnosis and potential intervention. Despite advancements in understanding its pathogenesis, including its association with Epstein-Barr virus (EBV) and states of immunosuppression, the prognosis remains grim with a median survival of approximately 2 years. Differential diagnosis, often challenging given its radiographic similarities with other pulmonary conditions, underscores the importance of histological confirmation. Current therapeutic strategies, though not definitive, encompass a range of treatments from immunosuppressants to multi-agent chemotherapy, tailored to the disease's extent and severity. Future research and therapeutic innovations are essential to improve outcomes for patients afflicted with this rare and aggressive disease.

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