Pulmonary lymphomatoid granulomatosis mimicking lung cancer

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Abstract

Lymphomatoid granulomatosis (LYG) of the lung is an extremely rare, Epstein–Barr virus-related lymphoproliferative disease. We report a case of pulmonary LYG that presented as a large necrotic mass.

Keywords: Lymphomatoid granulomatosis • Lymphoma • Lung cancer

INTRODUCTION

Lymphomatoid granulomatosis (LYG) is a rare, Epstein–Barr virus (EBV)-related lymphoproliferative disease, which was initially described as pulmonary angiitis and granulomatosis [1]. It remained uncertain for years whether LYG represents a type of lymphoma or an inflammatory process [2, 3].

The natural course is extremely variable, but most of the published series report high mortality (40–70%) [2–4].

We report here on an extremely rare case of large necrotising LYG type that was treated by pneumonectomy.

CASE REPORT

A 55-year-old male, ex-smoker, was presented with complaints of 3 months history of weight lost of 16 kg, dry cough and mild exertional dyspnoea. His past medical history included reflux disease and sclerosing cholangitis since 1998. On the chest radiograph, left upper lobe mass was seen. The contrast chest CT scan revealed a large necrotic tumour in the left upper lobe measuring 6.7 × 8.9 cm (Fig. 1a and b). 18F-fluoro-2-deoxyglucose positron emission tomography (FDG-PET) showed a large, thick-walled mass measuring 8.8 cm in maximum diameter in the left upper lobe (SUV max 22.1).

Figure 1 (a) CT without contrast cavity and (b) CT with contrast—showing a large cavitating mass that abutted the major fissure and mediastinum. (c) PET and (d) PET-CT—showing a large mass measuring 8.8 cm diameter in the left upper lobe (SUV max 22.1).

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the left upper lobe (SUV max 22.1) with no other abnormality (Fig. 1c and d). Bronchoscopy showed tumour obstructing the left upper lobe. Endobronchial biopsy consisted of necrotic debris. The cytological examination of bronchial washing showed rare viable tumour cells that were reported as consistent with non-small cell lung carcinoma. Pulmonary function tests showed FVC of 3.25 l (74%) and FEV1 of 2.49 l (71%). EBUS-TBNA followed by mediastinoscopy were performed, both negative for malignancy. Subsequently, the patient underwent thoracotomy. A diagnostic wedge resection was impossible without opening necrotic cavity. As the malignant process abutted the left main pulmonary artery, the only possible access to achieve complete resection was intrapericardial pneumonectomy with resection of mediastinal pleura and part of pericardium. Histology confirmed an atypical lymphoid proliferation with extensive necrosis favouring LYG grade 3, with marked dense secondary inflammatory changes (Fig. 2a–e). Systematic lymph node dissections showed no evidence of malignancy. The resection margin was negative for tumour. One year after surgery, the patient is followed by oncology service without chemotherapy.

**DISCUSSION**

Liebow first described LYG as the triad of polymorphic lymphomatoid infiltrate, angiitis and granulomatosis [1]. The current view is that LYG is a form of EBV-induced B-cell lymphoma rather than a primary vasculitis [2–6]. To this date, ~600 cases of LYG were described. Similar to our patient, most patients are middle-aged adults with predominance of men. Presenting symptoms included: cough, fever, dyspnoea, weight loss and sweating. Acute respiratory distresses and life-threatening haemoptysis have been reported. LYG has been associated with other diseases including autoimmune/systemic diseases (biliary cirrhosis, sarcoidosis, ulcerative colitis and hematopoietic diseases).
Extrapulmonary manifestations of LYG affect skin [25–50%], central nervous system [10–35%], upper airways [10–30%] or other organs [2–4]. The radiological features usually include multiple poorly—defined nodular opacities measuring 1–8 cm in diameter predominates in lower lobes. A cavitating lesion occurs in 20–30% of pulmonary nodules. The role of FDG-PET CT in the diagnosis of LYG is undetermined.

The diagnosis is difficult and endobronchial biopsy or trans-thoracic needle biopsies are often non-conclusive. Lymphoma cells are well known to mimic carcinoma on the cytology specimen. The role of surgery is to obtain diagnostic tissue. Therapeutic resections in unilateral, less advanced cases has been reported [2, 3, 7, 8].

The overall clinical pre-operative assessment directed towards NSCLC and we operated on the patient with intention to treat cancer. We did not perform trans-thoracic biopsy as the CT scan showed the necrotic cavity for which biopsy is often non-conclusive.

Standard treatment of LYG has not been established. Therapy varies from observation, steroids to aggressive chemotherapy depends of grade of the LYG and centre experience [1–7]. Prognosis is poor. The mortality is between 54 and 65%, and deaths occur in first 36 months after diagnosis [1–7].

Conflict of interest: none declared.

REFERENCES