Thoracic Surgery Quiz – Case 3

A 60 year old man presented to our clinic due to persistent cough of 2 weeks' duration without an accompanying fever. On the initial work up a chest X-ray revealed multiple opacities of both lungs as well as a parenchymal mass obstructing the right middle and lower lobar bronchi (fig. 1) and he was admitted for further investigation.

The patient was HIV positive for 15 years. One year before he had been investigated for appetite loss, weight reduction of 8 kg, persistent subfebrility (37.5 °C) of 3 weeks' duration and anemia. Chest X-rays, gastroscopy, bone marrow biopsy and colonoscopy were non-diagnostic.

At the current admission, a diagnosis of tuberculosis was suspected but the subsequent investigation did not confirm it. A chest CT showed multiple bilateral nodular infiltrates without hilar or mediastinal lymphadenopathy (figures 2 and 3). Bronchoscopy was performed but did not reveal any diagnostic abnormalities. A decision was made to proceed with an open mini-thoracotomy to obtain a tissue sample. Histological examination of the lung tissue showed a diffuse large B-cell non-Hodgkin's lymphoma (B-NHL). Immunohistological staining was positive for CD20, bcl-6, bcl-2 and negative for CD30 and CD3.

Comment

High-grade B-NHL represents 11 to 19% of cases of primary pulmonary lymphoma (PPL) in the series that have been published. This histological type often occurs in patients with underlying immunological disorders, such as solid organ transplant recipients, HIV infection and Sjögren's syndrome. The Epstein-Barr virus has been implicated in the development of some of these high-grade B-NHL that affect the lung. The possibility of a common precursor B cell for both HD and B-NHL has been described in several papers and is based on published reports of HD transformed to B-NHL.

The manifestations of the disease vary according to the immune status of the patient. In the case of HIV, highly active anti-retroviral therapy (HAART) causes a dramatic improvement in the patients' immune status associated with a rising CD4 count, declining viral load and a decreased incidence of opportunistic infections.

In high-grade B-NHL PPL the lymphoid tissue infiltrates the vascular and pleural structures and numerous necrotic zones can be observed. Definitive diagnosis is achieved through histological examination of the lesion. In most cases the tissue to be examined is obtained surgically. Immunohistochemical examination is required to exclude the presence of sarcoma, melanoma or carcinoma and it demonstrates the clonal nature and the B phenotype of the infiltration. The treatment of choice is surgical intervention followed by combination chemotherapy as usually used in high-grade lymphomas. High-grade B-NHL PPLs-B have a decidedly less favorable prognosis than low-grade lymphomas, with an average survival of approximately 3 years.

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Diagnosis: Primary pulmonary lymphoma, diffuse large b-cell type