

Primary Pleural Lymphoma: Case Report

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ABSTRACT

We report a case of a 70-year-old female with a history of prolonged exposure to biomass during childhood, non-smoker and no history of exposure to people with tuberculosis who presented with progressive dyspnea. Chest CT showed circumferential and nodular suitable pleural thickening with involvement of the mediastinal pleura and ipsilateral pleural effusion. An ultrasound-guided pleural biopsy was performed with a Trucut needle. The Histopathology result of the pleural biopsy determined an infiltration due to non-Hodkin lymphoma of lineage B. Immunohistochemistry CD20 positive, CD3 Positive. This case is thought to be a very rare case of primary malignant lymphoma arising in the pleura without a history of chronic pleural inflammation or immunocompromise.

Keywords: Primary pleural lymphoma, pleural biopsy, pleural effusion.

INTRODUCTION

Pleural disease in non-Hodgkin's lymphoma is well documented and commonly presents with pleural effusions in 20% of patients¹. However primary pleural lymphoma accounts for <0.5% of all non-Hodgkin lymphoma and ~2.5% of primary chest wall tumors^{2,3}. Two types of primary pleural lymphomas have been described the body cavity-based lymphoma in patients with HIV and the pyothorax associated pleural lymphoma in those with tuberculosis. Primary pleural non-Hodgkin's lymphoma in an immunocompetent patient without a history of chronic pyothorax is extremely rare⁴. This last type of presentation is what our patient manifests.

CLINICAL CASE

A 70-year-old female with a history of prolonged exposure to biomass during childhood, non-smoker, no history of exposure to people with tuberculosis, and drug allergies denied. She had no past medical history of HIV infection, tuberculosis, pyothorax or autoimmune disease. His condition began two years ago with asthenia, fatigue, cough with white expectoration, weight

loss, and dyspnea, and he was admitted to the pulmonology department with a diagnosis of right pleural effusion. Clinically, with occasional cough, 92% saturating without supplemental oxygen. The patient was afebrile, tolerating diet, without hemodynamic or neurological compromise and without alteration of ventilatory mechanics. Chest CT showed circumferential and nodular suitable pleural thickening with involvement of the mediastinal pleura and ipsilateral pleural effusion. No mediastinal lymphadenopathy or lesions in the contralateral lung parenchyma were observed. (Fig. 1). The blood count was normal.



Figure 1: Pleural thickening with mediastinal and costal involvement.

Upon admission, it performed an ultrasound-guided pleural biopsy with a Trucut needle. Subsequently, a 7 Fr small lumen intrapleural catheter was placed in the right safety triangle. Due to evidence of septa in thoracic USG, intrapleural fibrinolysis was performed with 10 mg of alteplase to improve pleural drainage. The Histopathology result of the pleural biopsy determined an infiltration due to non-Hodkin lymphoma of lineage B. Immunohistochemistry CD20 positive, CD3 Positive. (Fig. 2).

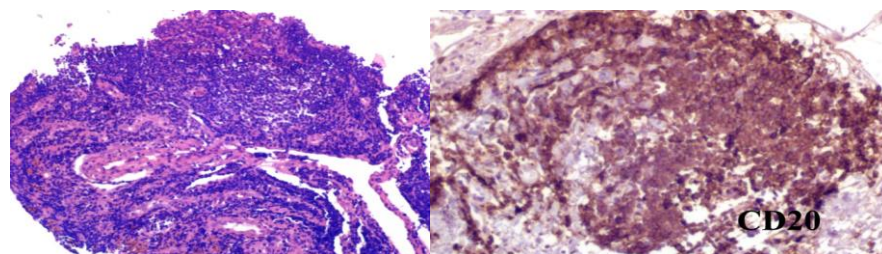


Figure 2: CD3 and CD20 positives.

Cytochemical and cytological of pleural fluid: Glucose 92 mg/dL, Total proteins 1.62 g/dL, Ph 8, Leukocytes 0/mm³, Pleural fluid/serum protein ratio 0.2. There was no bacterial growth and no malignant cells were identified. Subsequently, complete drainage of the pleural cavity was completed with chemical pleurodesis with povidone-iodine. After completing the diagnostic approach, she was referred to the hematology service to start chemotherapy.

DISCUSSION

Only two forms of high-grade lymphoma involve the pleura as primary neoplasms: primary effusion lymphoma (PEL) and pyothorax associated lymphoma (PAL)⁵. Fluid overload-associated large B-cell lymphoma (FO-LBCL) is a new entity described in the fifth edition of the World Health Organization Classification of Hematolymphoid Tumors. It refers to malignant lymphoma present with symptoms of serous effusions in body cavities (pleural, peritoneal, and pericardial) in the absence of an identifiable tumor mass⁶. It is thought that longstanding

pleural inflammation is an important factor in the development of lymphoma in the latter scenario. It has been postulated that there is chronic stimulation of B cells in the pleural cavity because the most common malignant lymphoma arising in the pleura has been reported to be diffuse large B-cell NHL³.

Although any type of lymphoma can involve the pleura, diffuse large B-cell lymphoma has been reported as most frequent, followed by follicular lymphoma, with rates of approximately 60% and 20%, respectively. A small series of case reports have identified other types of pleural lymphoma, such as extra-nodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (EMZL/MALT-type) and mantle cell lymphoma. Therefore, primary pleural non-Hodgkin's lymphoma (NHL) in an immunocompetent patient without a history of chronic pyothorax is extremely rare⁷.

Other possible etiologic factors reported include history of autoimmune disease, such as Sjögren's syndrome, rheumatoid arthritis, chronic lymphocytic thyroiditis, and Epstein-Barr virus (EBV) infection. In body cavity-based lymphoma, also referred to as primary effusion lymphoma, human herpes virus-8/Kaposi's sarcoma herpes virus infection is always present⁸. An approach to correct diagnosis of pleural tumors depends on the pattern of involvement focal or diffuse, unilateral or bilateral, and calcified or noncalcified. The role of imaging is to identify pleural thickening, differentiate benign and malignant pleural thickening, and identify the cause if possible⁹. On Computed Tomography (CT) scan, malignant pleural thickening is nodular (>1 cm), shows circumferential involvement, and involves the mediastinal pleura. On imaging, benign pleural thickening appears as a diffuse involvement of pleura¹⁰.

Pleural effusion (PE) is a common sign associated with various disorders, including tuberculous pleurisy, congestive heart failure, malignancy, and pneumonia. PE caused by primary and metastatic tumors of the pleura is categorized as malignant pleural effusion (MPE), Lung cancer is the most frequently associated with MPE, followed by breast, ovarian, lymphoma, mesothelioma and gastrointestinal cancer. Effusion cytology and pleural biopsy are the gold standards for MPE. A pleural biopsy or thoracoscopy is required when effusion cytology is negative, and malignancy remains the suspected cause of PE¹¹. In our case we resorted to an ultrasound-guided biopsy with a Trucut-type needle to obtain the diagnosis since the cytological test was negative. On the other hand, according to the BTS guidelines for the management of pleural disease, it is recommended that pleural drainage be performed with a small caliber tube (14F or smaller).¹²

Treatment paradigms focus on the palliation of dyspnea, which can be achieved with serial thoracentesis, an indwelling pleural catheter placement, chemical pleurodesis, or surgical interventions such as mechanical pleurodesis. Placement of an indwelling pleural catheter and performance of chemical pleurodesis is necessary for MPE with known or suspected expandable lung in the absence of prior definitive therapy. IPC alone is recommended for patients with nonexpandable lungs, failed pleurodesis, or in cases of loculated effusions, as these lungs are less likely to re-expand to allow for pleural symphysis¹³.

Chemotherapy is based on the CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) or more intensive regimens such as MACOP-B (methotrexate with leucovorin rescue, doxorubicin, cyclophosphamide, vincristine, prednisone, and bleomycin), with a

significant benefit due to addition of immunotherapy with rituximab (R). A potential benefit of densification by a biweekly application of R-CHOP was raised by an exploratory analysis of 50 patients with PMBCL included in the phase-III UK NCRI R-CHOP-14 versus -21 trial, where a trend to better progression-free survival and overall survival detected after R-CHOP-14 ¹⁴.

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