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Case report

Rare case of primary extranodal marginal zone lymphoma of the thorax

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ABSTRACT

Primary lymphoma presenting a solitary lesion of the chest wall is extremely rare, as the majority of chest-wall tumors arise from metastasis. We report a case of a 64-year-old man with no history of HIV infection or pyothorax who presented with dry cough, right pleuritic pain and dyspnea. A computed tomography scan revealed an irregular pleural mass invading his right chest wall with pleural effusion. CT-guided needle biopsy revealed extranodal marginal zone B-cell lymphoma. The patient was treated with chemotherapy and radiotherapy. The patient has 9 years of follow up with 2 relapse's episodes.

1. Introduction

Primary pleural lymphoma is extremely rare, accounting for only 0.3–1% of all extranodal lymphomas [1]. Two types are described in the literature; primary effusion lymphoma in the setting of human immunodeficiency virus (HIV) infection, and pyothorax-associated lymphomas, with a strong Epstein-Barr virus association. The clinical symptoms are generally non-specific and can lead to a delay in diagnosis. Non pyothorax or HIV associated lymphoma are very rare. We report a case of primary pleural marginal zone B-cell lymphoma without history of pyothorax or HIV infection with a follow-up of 9 years, and review of the literature.

2. Case presentation

The patient was a 64-year-old male, suffering from rheumatoid arthritis. His symptoms were dry cough, right pleuritic pain, dyspnea and asthenia but had no fever, night sweats or weight loss. Physical examination was consistent with a right pleural effusion and revealed no other notable findings. Chest radiography showed right pleural effusion (Fig. 1). Blood tests showed a biologic inflammatory syndrome with elevated C-reactive protein. Chest computed tomography (CT) showed a large right pleural effusion, pleural thickening and a suspicion of a medullary endocanalar extension without lymphadenopathy. Pleural puncture revealed an exudate with mixed formula. Gram's stain, Ziehl Nielsen stain and cultures of pleural fluid were negative. All bacteriologic samples were negatives. Cytological examination of the pleural fluid and pleural biopsy using an Abram's needle didn't contribute to any diagnosis. He underwent CT-guided needle biopsy of the pleural mass. Histopathological and immuno-histochemical examinations revealed monotonous infiltration of B-cells expressing CD-20 antigen (Fig. 2). In the absence of any evidence of lymphoma outside the pleural space, the diagnosis of primary pleural marginal zone B-cell lymphoma was made.

Serology for the human immunodeficiency virus (HIV) was repeatedly negative.

Research for HHV8 and EBV infection was not performed. Bone marrow biopsy didn't show any abnormalities. Magnetic resonance imaging was performed and showed pleural thickening extended from D4 to D7, to vertebral bodies (D4 to D7), and to posterior arches and soft tissue. The patient underwent staging CT of the brain and the abdomen that demonstrated no evidence of metastasis.

The patient was treated with six cycles of cyclophosphamide, doxorubicin, vincristine, prednisone (CHOP) regimen with radiotherapy. Chemotherapy was well tolerated with no major toxicity. His chest pain resolved completely and CT became normal. Follow-up CT showed complete resolution of the pleural mass and effusion. The patient had no relapse and a continuous follow-up in our department. After 6 years (on 2014), a CT scan of the thorax showed pleural thickening and moderate pleural effusion on the right side, without lymph node swelling in the mediastinum. Pleural biopsy revealed B-cells expressing CD-20 antigen. Exclusive radiotherapy was started. Chest CT performed

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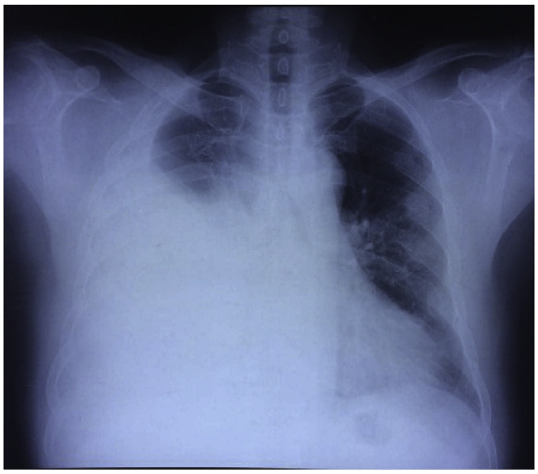

Fig. 1. Chest X ray: right pleural effusion.

four months later showed complete response.

However, 3 years later (on 2017), pleural thickening without pleural effusion, or significant mediastinal lymph node were observed on contrast-enhanced CT of the thorax (Fig. 3). He underwent CT-guided needle biopsy. Histopathological and immuno-histochemical examinations revealed infiltration of B-cells expressing CD-20 antigen. The diagnosis of relapse of a primary pleural marginal zone B-cell lymphoma was made. The patient was addressed to the hematology department.

3. Discussion

Primary pleural lymphoma occurring in immunocompetent patients without history of chronic tuberculosis, empyema or HIV infection is extremely rare [2–4]. It accounts for about 0.3–1% of extranodal lymphoma [1]. King et al. [5] reported that primary chest wall malignant lymphoma is very rare, representing only 2.4% of primary chest wall soft tissue tumors.

The mechanism of this rare type of lymphoma is likely a stimulation of B-lymphocytic cells in the pleural cavity, which is seen in long-standing chronic pleural disease [6]. Other postulated mechanisms for primary pleural lymphoma include antecedent of autoimmune disease, such as Sjogren's Syndrome, rheumatoid arthritis, malignant lymphoma (elsewhere in the body), chronic lymphocytic thyroiditis, thyroid lymphoma and Epstein-Barr virus (EBV) infection [6]. In fact, our patient was a 64-year-old male, suffering from rheumatoid arthritis. The chronic stimulation of B lymphocytes and the decrease in the number of circulating T lymphocytes may lead to lymphoid cell hyperplasia [7].

Primary pleural lymphoma has diverse clinical manifestations but a lack of specificity. Our patient presented a pronounced dyspnea, which was caused by a right-sided pleural effusion diagnosed as pleural lymphoma. Two forms of high-grade lymphoma involve the pleura as primary neoplasms are reported in the literature: primary effusion lymphoma (PEL) in HIV patients and pyothorax associated lymphoma (PAL) [8]. Both have well characterized clinical and pathological features. Our case shows typical histological and immunohistological findings of pleural marginal zone B-cell lymphoma. Therefore, primary pleural non-Hodgkin's lymphoma (NHL) in an immunocompetent patient without a history of chronic pyothorax is extremely rare [1,7].

Primary pleural lymphoma needs to be differentiated from other pleural diseases like pleural mesothelioma, which can occur diffusely at any place in the pleura without enlarged mediastinal and hilar lymph nodes. Pleural plaques were seen in less than 4% of patients presenting pleural lymphoma [9].

CT has traditionally been used to evaluate the disease and extent of spread. Aquino et al. [10] reported that extrapleural involvement was commonly (30%) presented as a tumour, enlarged lymph nodes, or direct invasion by the adjacent parietal pleural mass. Yookyung et al. [11], reported in 2 cases, an extrapleural space involvement by direct invasion of the parietal pleural masses. Intercostal arteries were well visualized in the conglomerated pleural and extrapleural masses on axial contrast-enhanced CT images, representing the “pleural sandwich sign”. This sign has not been reported in other pleural tumors, including metastases, malignant mesothelioma, and solitary fibrous tumors that usually do not have extrapleural lesion at the initial presentation or quickly invade intercostal vessels when they invade the extrapleural space. Therefore, this characteristic CT sign would be helpful to differentiate primary pleural lymphoma from other pleural tumors.

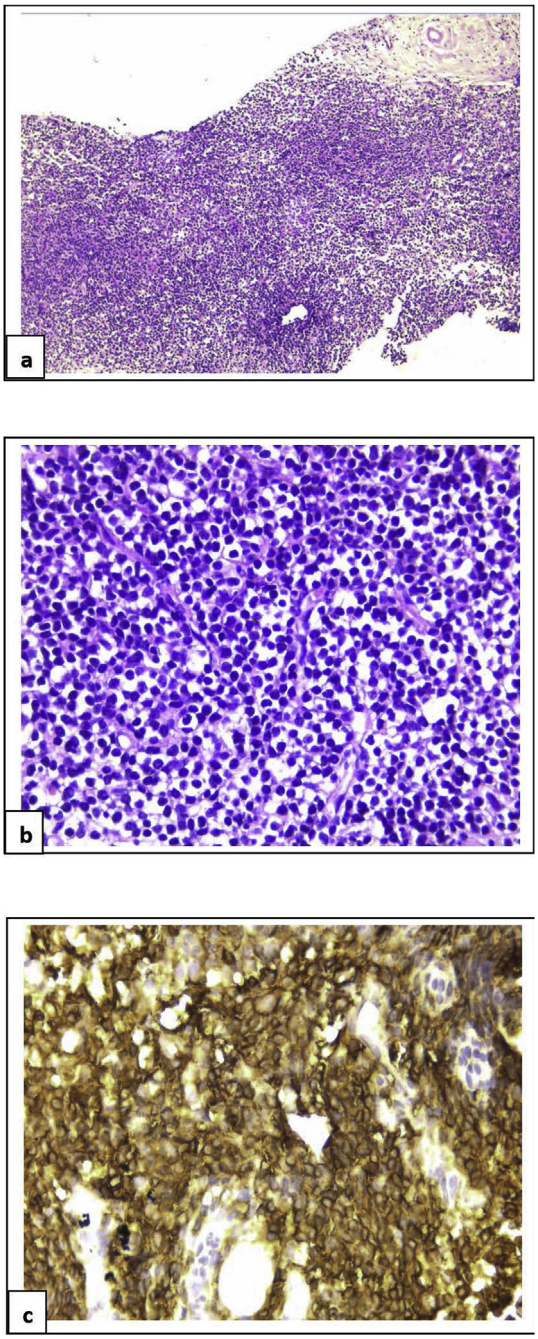

Fig. 2. Histopathological and immuno-histochemical examinations: a: a diffuse infiltrate of small lymphoid cells (Hematoxylin & Eosin x100) b: monotonous small lymphoid cells with incisored nuclei (Hematoxylin & Eosin x400) c: the tumour cells stain for CD20.

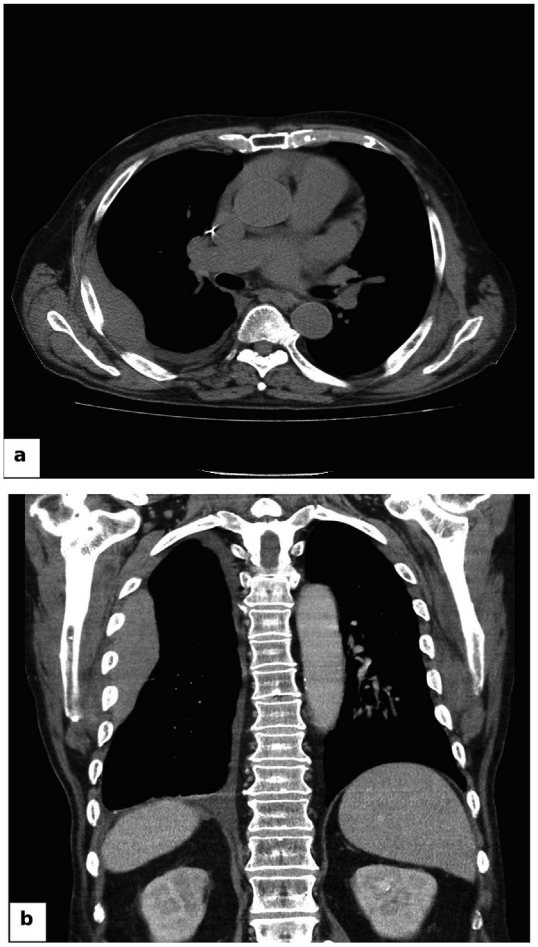

Fig. 3. Chest CT scan: a: axial CT scan image b: coronal CT scan image Pleural thickening without pleural effusion, or significant mediastinal lymph node.

However, additional modalities such as magnetic resonance imaging (MRI) and positron emission tomography (PET)/CT have emerged. PET/CT is more accurate in the overall staging of lymphoma than CT alone and can be used to evaluate treatment response [12].

Our patient had initially primary pleural marginal zone B-cell lymphoma with total resolution, then had 2 relapses of the disease. The patient was treated with six cycles of cyclophosphamide, doxorubicin, vincristine, prednisone (CHOP) regimen with radiotherapy. However, there is no standard treatment regimen for primary pleural lymphoma. Different treatment modalities are proposed for extranodal marginal zone B-cell Lymphoma like chemotherapy, radiotherapy or surgery. In order to conserve lung function and reduce the risks of operation, chemotherapy should be considered as a first line option for the treatment of pulmonary marginal zone B-cell lymphoma [13]. The most common regime is cyclophosphamide, doxorubicin, vincristine and prednisolone (CHOP) [14]. It remains controversial whether patients with lymphoma located only in the chest wall should undergo surgical resection. Romagurea et al. [15] reported that surgical debulking is associated with improved survival in Stage I–II diffuse large-cell lymphoma. However, Pulmonary extranodal marginal zone B-cell lymphoma is an indolent disease and exhibits a favorable prognosis, with a five-year survival rate of ~90% [16,17], but extra-pulmonary lesions and lymph node involvement are poor prognostic factors [16]. Aosaza et al. [18] found that non-pyothorax associated lymphoma (NPAL) is slightly better than PAL: 5 of the 21 historical patients died, 1 recurred after remission, and the remaining 15 experienced remission following operation, chemotherapy, or radiation therapy. The present case has a survival of 9 years at list. We suppose that it is related to the good Performance Status (PS) and the preserved respiratory function of our patient [19].

4. Conclusion

We describe this very rare case of primary pleural extranodal marginal zone B-cell lymphoma without history of HIV infection or pyothorax. It is difficult to differentiate it from other malignant pleural diseases. No standard treatment regimen is defined for this particular entity.

Declarations of interest

None.

Authors' contributions

All authors participated in the preparation of the manuscript. Naceur Rouatbi and Samah Joobeur conceptualized and designed the manuscript. Ahmed Ben Saad and Rim Khemakhem performed manuscript writing. Nouha Ben Abdeljelil and Manel Njima performed the histopathologic examination. Asma Migaou performed the literature review. Saousen Cheikh Mhamed and Nesrine Fahem revised the english version. All authors read and approved the final version of the manuscript.

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