INTRODUCTION

The B-cell lymphoma of the marginal zone, also called Mucosa-Associated Lymphoid Tissue (MALT) Lymphoma, is an extranodal lymphoma that stems from a series of epithelial tissues, including stomach, salivary glands, lung, small intestine, thyroid and other organs, and is included within the non-hodgkin Lymphomas (NHL) [1].

When it presents as a pulmonary condition is called Bronchus-Associated Lymphoid Tissue (BALT) Lymphoma. This Lymphoma has a tendency to be localized during long periods of time in the tissue of origin, but has potential of systemic spread and transformation to a high-grade B cell lymphoma. We describe a BALT Lymphoma case with 13 years of evolution until its diagnosis.

CASE PRESENTATION

A 75 year-old female was referred to the Central University Hospital of Asturias, Oviedo, in March 2000, because of an abnormal chest X-ray (CXR) as a casual finding. The patient at this time was asymptomatic (no chest pain, night sweats or fever). She had no known toxic habits nor surgical or medical background of interest. The chest X-ray showed a left lower lobe (LLL) alveolar infiltrate. She was diagnosed with community acquired pneumonia and received antibiotic treatment for 10 days. In her follow-up visit, she remained asymptomatic, but due to the persistence of the lesion 2 months after the pneumonia diagnosis, a computed tomography (CT) scan of the chest was performed. The CT scan revealed a LLL alveolar consolidation and associated bronchiectasis. On January 10, 2001, a bronchoscopy was conducted and no endobronchial lesions were found, being the bronchial aspirate (BAS) and brushing technique negative for malignancy and infection. With the diagnosis of bronchiectasis in the LLL, follow up visits continued for a period of 3 years, in which the patient was clinically stable and the initial lesion in the LLL remained unchanged.

The patient remained asymptomatic and did not return for subsequent follow up visits until October 2013; when her primary care physician referred her back to our hospital due to dyspnea and left pleuritic chest pain. The physical examination was normal (no palpable peripheral adenopathies), except for pulmonary auscultation, where diminished respiratory sounds and crackles were found at the left lung base. Laboratory tests (complete blood count and biochemistry parameters) were within normal ranges. The serum lactate dehydrogenase (LDH) level was 230 UI/L. A spirometry showed moderate obstruction to air flow (FVC 1920-72%, FEV1:1260-68%, FEV1/ FVC 65%).

The chest X-ray (Figure 1) showed an increase in size of the LLL lesion compared with the CXRs of 2000. The CT scan revealed (Figure 2) an increase in the density of the alveolar consolidation accompanied by air bronchogram and bronchiectasis in the postero-basal segments of LLL with no mediastinal or hilar adenopathies. In November 2013, a new bronchoscopy with transbronchial biopsy was performed, but a diagnosis was not achieved, therefore, a CT image guided lung biopsy was performed 2 weeks later. The microscopic examination revealed tumor tissue with a small size of monomorphous lymphoid cellularity. Immunohistochemical staining demonstrated that the cells were positive for CD45, CD20 and CD79. The CD138 was positive with more Kappa than Lambda positive cells.
With the final diagnosis of Bronchus-Associated Lymphoid Tissue (BALT) Lymphoma, the patient was referred to the hematology department in January 2014. The following studies were completed: serum protein electrophoresis (gamma monoclonal peak: 22.9 g/L), Immunofixation (monoclonal component IgM Kappa) and bone marrow aspirate (mature lymphocytosis: 16.25%; plasmatic cells: 2.25%; flow cytometry: plasmatic cells: 1.5% with clonality and normal immunophenotype). At the end of January 2014, treatment was started with Rituximab and Chlorambucil (6 cycles) ending in June 2014. Five months after the last course of treatment, a CT scan showed that the pulmonary lesion had significantly reduced its size (about 50%). In November 2014, the patient was in good condition without any evidence of disease progression. The patient remains in follow-up.

DISCUSSION

The NHL affects thoracic structures, mainly the mediastinum and lung parenchyma in more than 43% of the patients at some time during their evolution [2,3]. The BALT lymphoma represents 1% of all lymphomas [4] and more than two-thirds of pulmonary NHL. The BALT lymphoma is seen in its vast majority in 55-60 year old adults, without differences in incidence between sex [5]. The pathogenesis of BALT Lymphoma may be associated with chronic immune stimulation driven by infection or autoimmune disorder. However, the etiology of this entity is unclear [6].

The radiological presentation of BALT lymphoma in the CT scan is variable. It can occur as a single nodule or multiple nodules, parenchymal consolidation, focal or diffuse interstitial infiltrate, atelectasis and/or pleural effusion [7]. Of all the forms of presentation, the most common is the alveolar consolidation such as occurred in our case. According to Bae et al [8] the CT findings show only nodules or parenchymal consolidation (33%), multiple nodules or areas of consolidation (43%), bronchiectasis or bronchiolitis (14%) and diffuse interstitial pattern (10%). Several radiological patterns can coexist and the involvement can be bilateral in 60% of the cases. The pleural effusion is the less frequent radiological manifestation (9%) [9]. In our case the radiological manifestation was a unilateral alveolar consolidation without systemic involvement and without endobronchial lesion.

Normally, the parenchymal lung lesions have a slow growth, as seen in our case. The patient’s symptoms will depend on the degree of radiological affection and the presence or not of endobronchial lesion. The most common symptoms are cough, dyspnea, chest pain, bloody sputum and obstructive pneumonitis symptoms. However, it is common that patients do not present symptoms at the time of diagnosis.

Regarding treatment, chemotherapy may be proposed to patients with contraindications to surgery, elderly patients, or those who have progression after surgery. Regimens are based on alkylating agents (eg, Chlorambucil or Cyclophosphamide), Fludarabine, or Rituximab both as a single agents and combined with chemotherapy [10]. In our case, the patient has responded very well to Rituximab and Chlorambucil.

We must not forget primary pulmonary lymphoproliferative processes spite their low frequency; they course with multiple forms of presentation and should be included in the differential diagnosis of bronchopulmonary diseases that present with alveolar injury.

REFERENCES


