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Rituximab monotherapy become an optimal treatment for primary pulmonary malt lymphoma

Mr. Editor:

Extranodal marginal zone lymphoma or mucosal associated lymphoid tissue (MALT) is the most common form of marginal zone lymphoma, constituting approximately 5% of all B-cell lymphomas. It is originated outside of lymph nodes and the most frequent locations are stomach, small intestine, salivary glands, thyroid and lung [1].

MALT lymphoma with lung involvement has been linked to prolonged exposure to toxic substances like tobacco, infections and autoimmune diseases, developing a chronic antigenic stimulation of lung tissue. Its incidence is very low, being 3.6% of all extranodal lymphomas and 0.4% of non-Hodgkin lymphoma. Clinical presentation is usually non-specific and in most cases, is a chance finding in a thoracic image test [2]. The diagnosis is based on histopathological analysis of tissue obtained by biopsy, where a diffuse infiltration of clonal B cells is observed. Those infiltrated B cells are positive for CD20, CD43, CD79a surface markers and negative for CD5 and CD103 markers [3] .

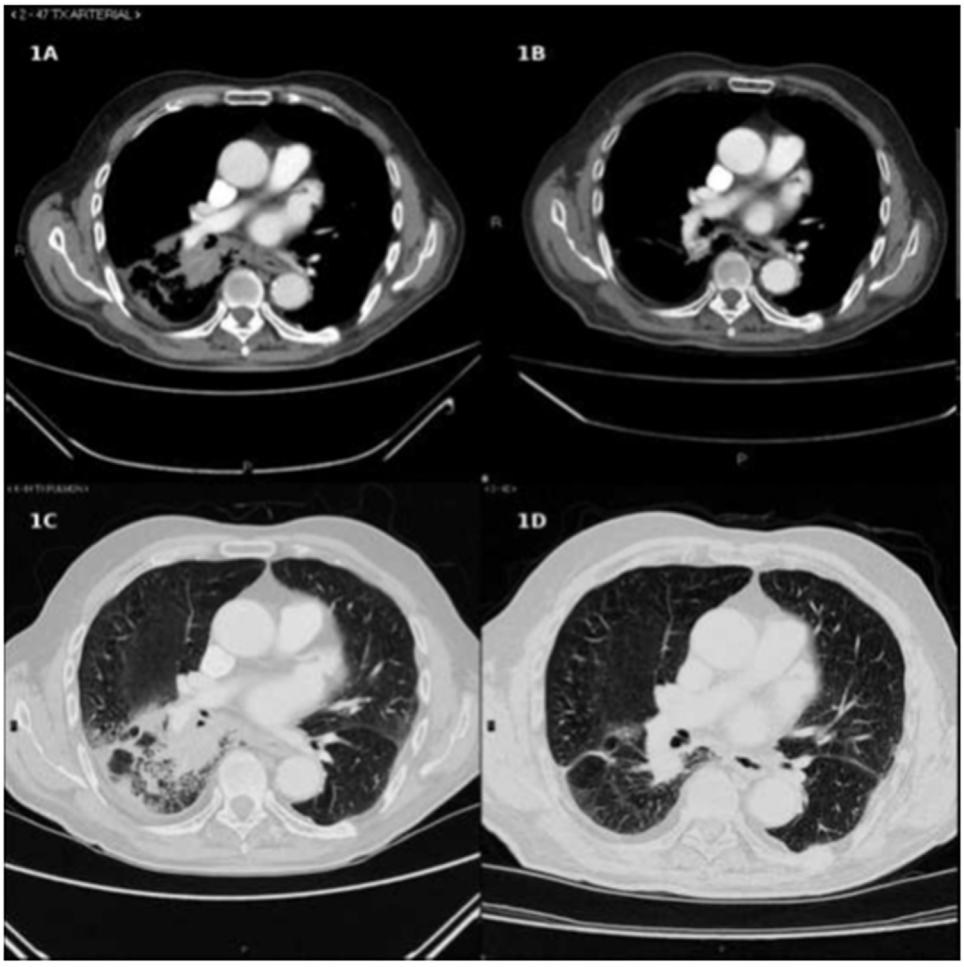

Fig. 1. A, C) Image 1A (mediastinum window) and 1C (lung window) where mass is shown in top right lobe surrounding right main bronchus. B, D) Image 1B (mediastinal window) and 1D (window lung) after four doses of rituximab monotherapy where the mass is almost disappeared.

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We present the case of an active smoker 82 year old man. He was admitted in February 2016 in our hospital with symptoms of dyspnea during the last 4 months, with right hemitorax pain, cough, expectoration and occasional blood traces in sputum. Blood count, biochemistry and serology were performed and all parameters were in normal range (including LDH and b2-microglobulin). In arterial gasometry he presented partial respiratory failure. The chest X-Ray showed a parahilar mass with associated micronodular pattern, loss of volume in the right lower lobe (RLL) and aortic elongation. In complete CT, a mass of about 35 mm in size in right upper lobe (RUL) surrounding the right main bronchus is observed, with right paratracheal, subcarinal and hilar lymphadenopathies of up to 20 mm, suggestive of bronchogenic carcinoma (Fig. 1a). Fiber bronchoscopy and mass biopsy were performed and reported as infiltration by low-grade B-lymphoma of the marginal zone with immunophenotype: $CD20^{+}$, $CD3^{-}$, $CD43^{-}$, $CD5^{-}$, CYCLINE $D1^{-}$, $BCL6^{-}$, $CD10^{-}$ and $CD23^{-}$.

The patient was reported to Hematologist, adding more probes as PET- CT, where hypermetabolic uptake in the right parahilar mass of RUL is observed, as well as metabolic activity in subcarinal region and ipsi and contralateral mediastinum (SUVmax 8). All together, we concluded that is a B-LYMPHOMA from the MARGINAL EXTRANODAL PRIMARY PULMONARY ZONE stage II 2-E (Ann-Arbor). Thus, he started treatment with rituximab monotherapy, receiving 4 cycles. After treatment evaluation using fiber bronchoscopy, no lesions suggestive of malignancy were observed, and a significant decrease in size − 6 mm of major axis- was observed in CT images (Fig. 1b). Due to this good response, it was decided to administer another 4 cycles of rituximab monotherapy, with excellent tolerance and without any adverse effects. In May 2016 the patient finished treatment, being completely asymptomatic and re-evaluating the response through PET, where hypermetabolism in the right lung was not observed. After 12 months of surveillance, the patient remains stable from hematological criteria.

Normally, treatment of pulmonary MALT lymphoma is not well defined. Usually it is based on surgical resection and occasionally radiotherapy on the affected area for localized stages. Moreover, chemotherapy or immunotherapy is applied for advanced stages or contraindication of the previous ones. Long-term results of these patients are excellent in most of the cases, with an overall survival of 70% − 100% at 5 years [4].

Herein, we demonstrated that Rituximab is a good election for patients with MALT lymphoma who express CD20. Rituximab offers a good quality of life and low toxicity (generally minor and transient side effects) for these patients [4,5]. Those are key points to be taken when deciding the best therapeutic option in malignant but indolent neoplasms such as MALT lymphoma.

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