Ulcerative colitis associated with extranodal marginal zone B-cell thyroid lymphoma of mucosa-associated lymphoid tissue and Hashimoto thyroiditis: description of a case

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Ulcerative colitis (UC) is a chronic and relapsing large bowel disease which predisposes to the development of large bowel cancer and other malignancies [1,2]. Primary thyroid lymphoma (PTL) is a rare but quite significant malignant tumor of the thyroid gland arising in a proportion of cases on the ground of Hashimoto thyroiditis [3]. The majority of cases are extranodal marginal zone B-cell lymphomas of mucosa-associated lymphoid tissue (MALT), diffuse large B-cell lymphomas or a combination of both [4].

The development of PTL during the course of UC has very rarely been described. Herein, we describe the case of a woman with long-standing UC who developed extranodal marginal zone B-cell MALT lymphoma. The patient, aged 46, suffered from total UC since the age of 26. During all these years, the disease was running with exacerbations of mild-to-moderate severity every one or two years, that were settled promptly. However, at least two exacerbations (the last one in 2008), were so severe that she required hospitalization, full doses of IV corticosteroids, nil by mouth and fluid and electrolyte replacement. From her past history, she mentioned appendicectomy at the age of 18. In 2005, 13 years after the onset of UC, she had undergone surgical drainage of a perineal abscess. Six months later, a total thyroidectomy was performed because of thyroid enlargement accompanied with clinicolaboratory signs of possible thyroid malignancy. Histology revealed the presence of extensive lesions of autoimmune Hashimoto thyroiditis and stromal fibrosis. A further immunohistochemical study using the CD20ass, Clg(κ,λ,γ,μ,α), CD21, CD5, CD23, CD45RO, CAM5.2 and PCR:IgH indices, revealed the presence of a polyclonal lympho-hyperplastic lesion [Clg(κ)/IgH] with characteristics compatible with extranodal B-(CD20ass+) non Hodgkin lymphoma arising from the cells of the marginal zone of MALT lymphoma. During the next years, she was under substitution treatment with thyroxin 0.1 mg daily. Up to 2012, there have been no indications of local recurrence or distal metastases. Now, she is on maintenance treatment with per os mesalamine. The thyroid function has returned to normal levels.

It is the author’s intention not to add immunosuppressives, and certainly not, biologic agents, in case of a future severe flare-up of UC in this patient. An exacerbation could be treated with appropriate doses of corticosteroids. The addition, or not, of immunosuppressives would be a matter of detailed discussion with the patient.

The combination of UC with PTL is extremely rare. García Arroyo et al [5] described a series of 6 patients with PTL of the type of large-cell lymphoma. One of these patients was concurrently suffering from UC [4], representing probably the only published case of a combination of UC and PTL so far. Obviously, there is no etiological link between UC and PTL and the development of PTL in this patient might be considered as development by chance. It has been suggested, however, that both antigenic stimulation on the setting of Hashimoto thyroiditis and aberrant somatic hypermutation are important factors contributing to the pathogenesis of PTL [5]. On the other hand, thyroid disease of the type of autoimmune thyroiditis and hypo- or hyperthyroidism have frequently been associated with UC [6,7], suggesting that autoimmunity is a significant factor in the whole process of both situations.

In conclusion, extranodal marginal zone B-cell MALT lymphoma could develop in the course of long-standing UC.

References