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A rare lymphoproliferative disorder associated with immunomodulating therapy in Crohn's disease

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Key words: Lymphomatoid granulomatosis. Lymphoproliferative disorder. Azathioprine. Crohn's disease. Inflammatory bowel disease.

Dear Editor,

A 15 mm erythematous-violaceous lesion in the right infra-clavicular region was identified in a 60-year-old male with Crohn's disease (CD) who had been treated with azathioprine for 24 months (Fig. 1). A biopsy study identified lymphomatoid granulomatosis (GL) and Epstein-Barr virus (EBV). An extension study was also performed via computed tomography, proteinogram and marrow biopsy and there were no significant findings. The patient was treated for one year with rituximab after excision of the lesion and withdrawal of azathioprine. There was no recurrence after 26 months of follow-up.

GL is a B-cell lymphoproliferative disorder associated with EBV and four cases have been described in patients with CD treated with azathioprine. The disease usually occurs in males (2:1 ratio) with pulmonary and cutaneous involvement (1-3). The median survival from diagnosis is 14 months (4) and is usually associated with myelo and lymphoproliferative syndromes and immunodeficiency conditions (2,4).

Dermatological lesions usually appear as erythema and subcutaneous nodules, with the classic histopathological triad of transmural lymphocytic vasculitis, large CD20+

atypical B lymphocytes on a background of polymorphic CD3+ T cells and necrotic foci in lymphoid aggregates (1,2). These lesions do not form authentic granulomas (3) and are classified into three grades based on the number of CD20+ B cells and EBV RNA. Grades I and II have a slower course, whereas grade III is more aggressive and has a worse prognosis. The evolution of the disease is very variable, and is able to self-limit or progress to a diffuse B cell lymphoma (1,2). Treatment is usually a combination of rituximab, methylprednisolone, vincristine, doxorubicin or dexamethasone. Early recognition and study of GL is crucial in order to start treatment quickly in patients with CD, treated with azathioprine.

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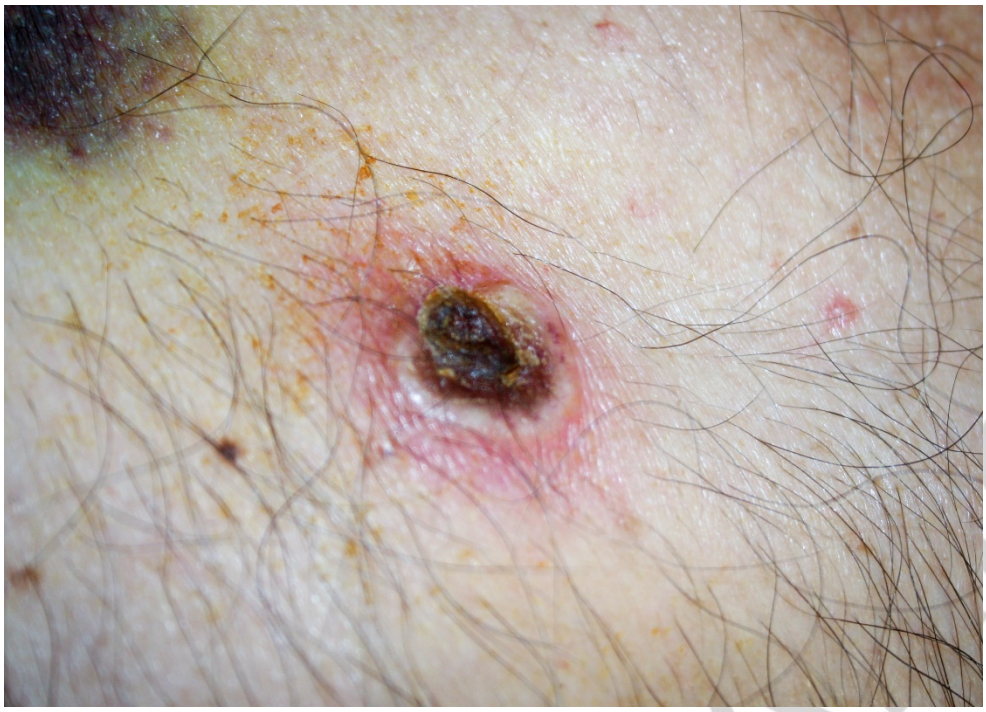

Fig. 1. Lymphomatoid granulomatosis.