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## Hepatosplenic T-cell lymphoma and inflammatory bowel disease

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*Dear Editor,*

A 48-year-old male with a diagnosis of ulcerative colitis 18 years previously, under immunosuppressive treatment with azathioprine during the last six years due to corticosteroid dependence, was admitted to the Emergency Department due to fever of one week's evolution. Blood tests showed thrombocytopenia, C-reactive protein (CRP) 96.9 mg/l, ferritin 3,021 ng/ml and hypertriglyceridemia. Blood and urine cultures were negative. Viral serologies (hepatitis B and C, human immunodeficiency virus [HIV], parvovirus, cytomegalovirus [CMV], herpes simplex virus [HSV]), atypical bacteria (*Borrelia*, *Chlamydia*, *Coxiella*) and screening for latent tuberculosis were also negative. Thoracoabdominal computed tomography (CT) scan only showed splenomegaly. The bone marrow aspirate revealed immature lymphoid cells and a hemophagocyte figure, fulfilling the criteria of hemophagocytic syndrome, starting corticosteroid therapy at a dose of 1 mg/kg. Subsequently, the existence of an

intrasinusoidal CD3 + CD5- lymphoid infiltrate and a FISH study with isochromosome 7q was reported, which is a characteristic pattern of hepatosplenic T-cell lymphoma (HSTCL). The study was completed with liver biopsy, finding 70 % infiltration of T lymphocytes (50 % gamma-delta), therefore the diagnosis was confirmed. Chemotherapy (cyclophosphamide, doxorubicin, vincristine, etoposide) was started with the aim of considering hematopoietic stem cell transplantation. Unfortunately, the patient died six months later.

## Discussion

HSTCL is a rare and aggressive subtype of peripheral non-Hodgkin T-cell lymphoma. Of the total cases reported, 10 % appear in patients with inflammatory bowel disease under immunosuppressive treatment with thiopurines and particularly in combination therapy with anti-TNF (1,2). Its incidence is unknown, but it is estimated at 0.1/1,000 patient-years and 0.3/1,000 patient-years for thiopurines and combination therapy, respectively (3). Despite aggressive therapies such as transplantation, the reported five-year survival is less than 16 % (4). To date, two systematic reviews on HSTCL have been carried out. Both conclude that most cases appear in males under 35 years of age with prior exposure to thiopurines for at least two years, especially in combination with anti-TNF (2,4). Two cases have been reported in our center, one of them was previously described (5).

In conclusion, we believe that although the low incidence of HSTCL should not influence the prescription strategies for thiopurines or anti-TNF, it is essential to adequately inform the patient of the benefits and possible risks before starting any immunosuppressive treatment.

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