LETTERS

Alpha heavy chain disease: a rare lymphoma hard to diagnose

Dear Editor,

A 30-year-old male presented having diarrhoea and abdominal pain since 14 months. Previous investigations identified no disease. Computed Tomography showed mesenteric and retroperitoneal lymphadenopathy with diffuse inflammation and intestinal dilatation. Duodenal and ileac biopsies revealed immunoproliferative small intestinal disease (IPSID), a rare lymphoma also called alpha heavy chain disease[1].

The bowel lamina propria was infiltrated mainly with plasma cells expressing monoclonal cytoplasmic truncated alpha heavy chain without light chain restriction. Histopathology is shown in Figure 1. Hypogammaglobulinemia, due to defective heavy chain assembly, mild anemia and hypoalbuminemia were present. Bone marrow aspiration was normal.

The patient underwent six cycles of bi-weekly chemotherapy with cyclophosphamide (750 mg/m^2), doxorubicin (50 mg/m^2), vincristine (1.4 mg/m^2) and prednisolone (50 mg/m^2/day) (CHOP). However, his symptoms deteriorated with massive mesenteric lymphadenopathy. He denied further treatment and died hospitalized in his country, after 4 months.

IPSID is variant of extranodal marginal zone lymphoma of mucosa associated lymphoid tissue (MALT), with defective alpha heavy chains secreted, observed in Mediterranean young adults with peak incidence in second and third decades. The malignant cells have lymphoplasmacytic phenotype[2]. Salem and Estephan proposed a novel staging system, combining the complementary former Galian and Salem systems and integrating cytological grade, submucosal involvement and anatomical stage of the lymphoma overall[3].

Middle East studies indicate decline in its incidence over the last 3 decades. This is explained by improved socioeconomic and hygienic conditions (vaccinations programs, decrease in malnutrition and intestinal infections), but also because of increased antibiotics use and decreased incidence of *Campylobacter jejuni*, which has been linked with IPSID, through chronic antigenic stimulation. Antimicrobial therapy with tetracycline, ampicillin or metronidazole is effective in early-stage IPSID, which may completely remit, showing the importance of early diagnosis. However, many patients experience transformation to diffuse large B-cell lymphoma (DLBCL), or distant metastases and fatal outcomes are frequent[2]. We conclude that IPSID transformed to DLBCL in our patient, because of his clinical course. Treatment with anthracycline regimens results in remission and longer survival[2].

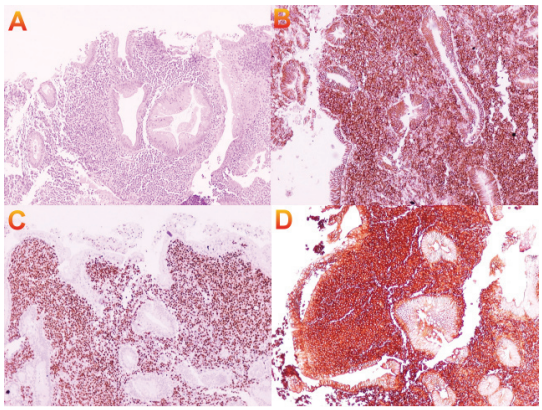

Figure 1: Small intestinal biopsy. Flattened mucosa diffusely infiltrated by plasma cells (A), positive to CD138 (B) and MUM1/IRF4 (C) antigens with cytoplasmic expression of IgA heavy chain (D) (A: Hematoxylin & Eosin, x100, B-D: IHC, x100).

Conflict of interest

None declared.

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Diamantidis MD[1], Kostopoulos I[2], Kaiafa GD[1]

[1]Haematology Department, First Propedeutic Department of Internal Medicine
[2]Department of Pathology
AHEPA General Hospital, Aristotle University, Thessaloniki, Greece

Corresponding author: Michael D. Diamantidis, MSc, PhD, Haematologist, e-mail: diamantidis79@yahoo.gr