Mediterranean lymphoma, an uncommon case of iron-deficiency anemia

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Clinical Image

A 57 year-old spanish man presented six-month period of iron-deficiency anemia. Physical examination was unremarkable. Upper endoscopy revealed mucosal edema and erythema beyond papilla of Vater with high mucosal friability. Specimen biopsies showed lymphoplasmacytic massive mucosal infiltration (Fig. 1) suggestive of immunoproliferative small bowel disease (IPSID). No association with Campylobacter jejuni was demonstrated, although Helicobacter pylori infection was detected in gastric biopsies. To complete the workup of the IPSID, bone marrow, computed tomography and capsule endoscopy studies were performed, confirming proximal small bowel involvement (Fig. 2). Helicobacter pylori eradication followed by 6 month-period of tetracycline treatment was indicated.

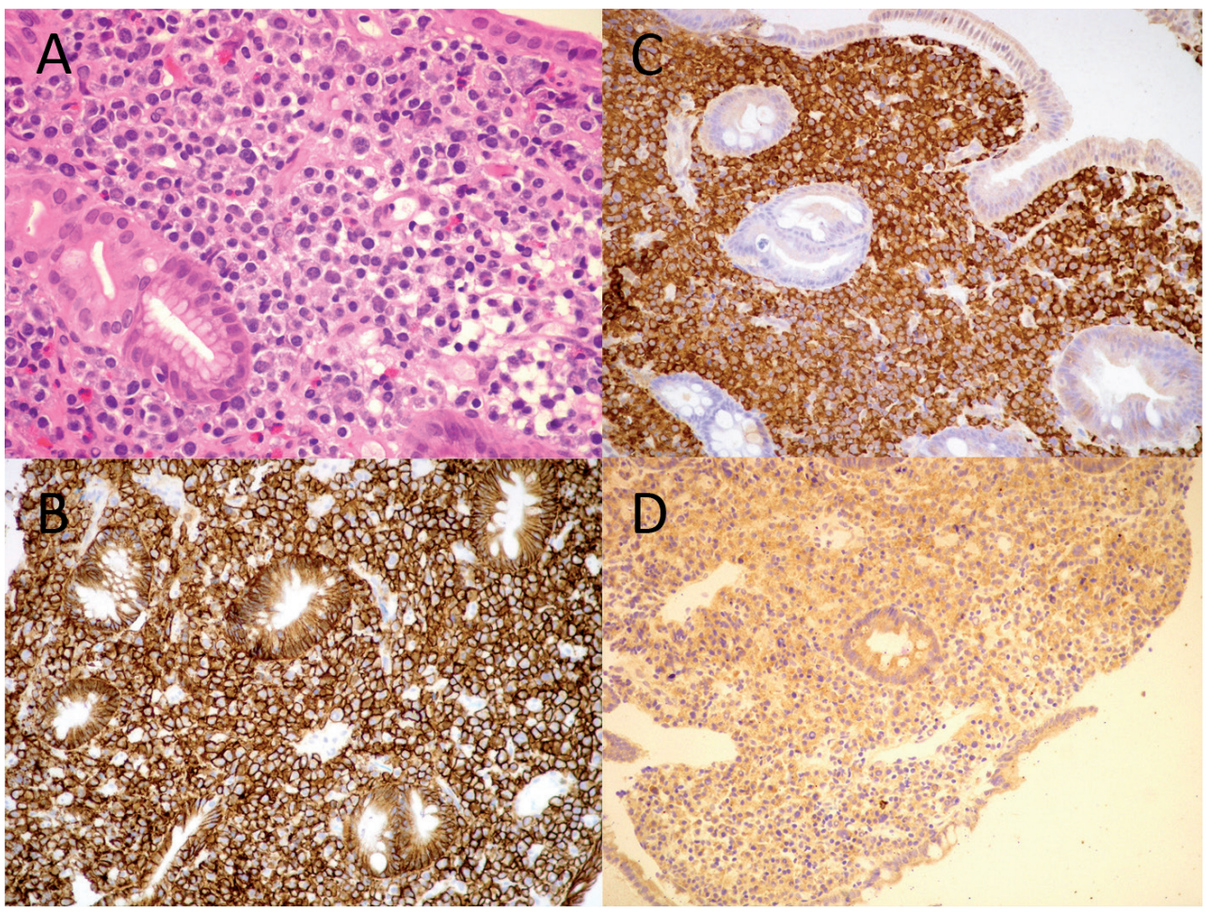

Fig. 1. — IPSID diagnostic is based on histological and immunochemistry studies : (A) Massive mucosal infiltration of lymphoplasmacytic (H-E, 40x) cells staining positive for CD138 (B), lambda-light chain (C) and negative for Kappa-light chain (D)..

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Conclusions

IPSID, also known as Alpha-chain disease or Mediterranean Lymphoma, is a variant of the B-cell lymphoma of mucosa-associated lymphoid tissue (MALT). Prevalence is higher in the Middle East and Africa and has been related to Campylobacter jejuni infection. IPSID is characterized by an excessive production of plasmatic cells with truncated alpha heavy chain (1). At early stages, it mainly involves the proximal small intestine in a diffuse and continuous pattern, resulting in malabsorption and diarrhoea. However, progression to lymphoplasmacytic and immunoblastic lymphoma is usually frequent (advance-stage), invading the intestinal wall, mesenteric lymph nodes and metastasize to distant organs (2). Antibiotic therapy (tetracyclines) and eradication of any concurrent infection (eg, parasites, viruses, Helicobacter pylori, Campylobacter jejuni) is the usual treatment limited to early-stage disease (3), while chemotherapy is reserved for those who do not respond adequately to antibiotics or those with advanced disease at presentation. CHOP regimen (Cyclophosphamide, Vincristine, Doxorubicin, and Prednisone) is the chemotherapy schedule used (4). Rituximab can be added if abnormal cells express CD20 (R-CHOP).

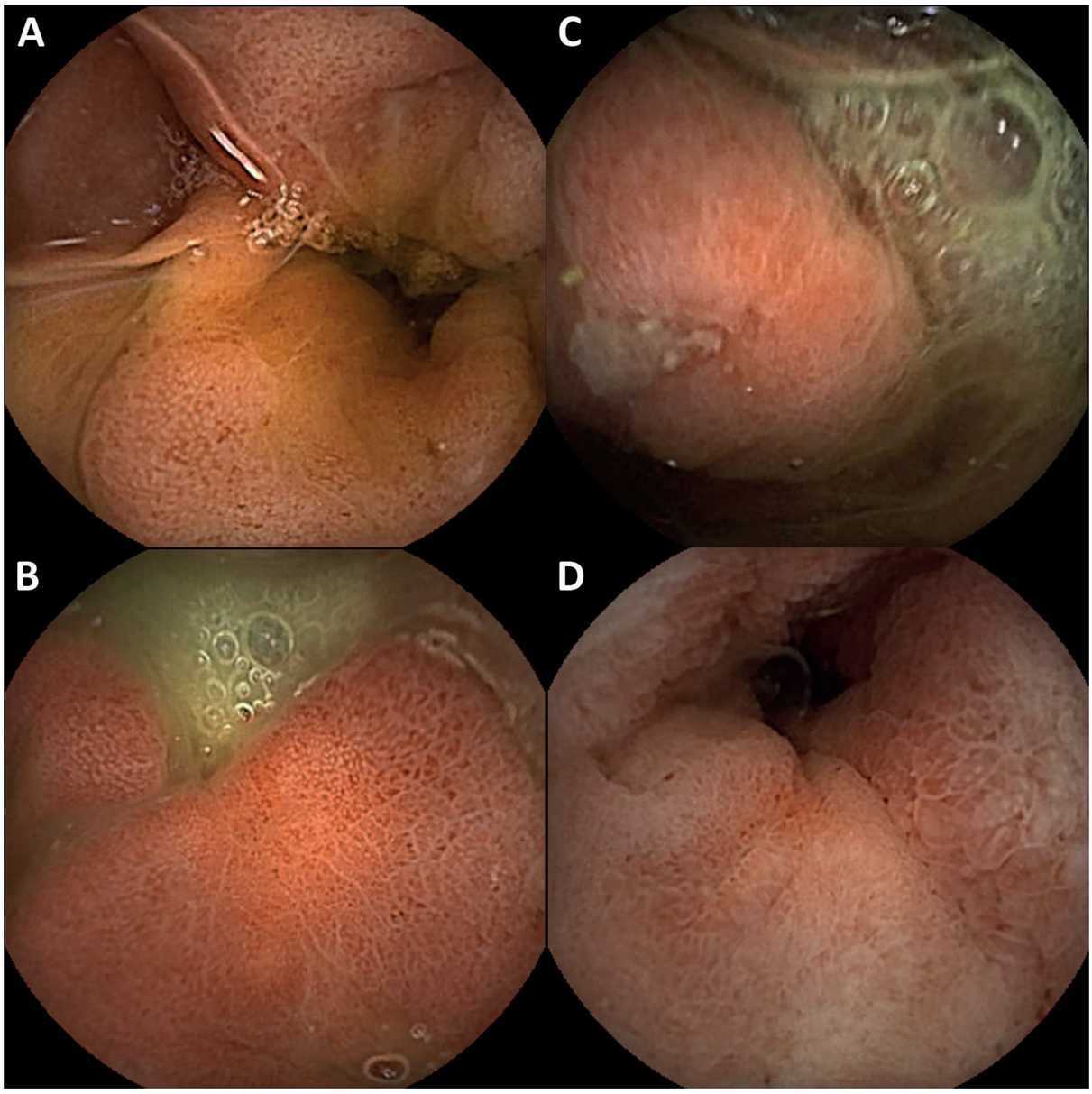

Fig. 2. — Mucosa of small bowel under the view of capsule endoscopy: mucosal edema (A) and erythema (B) in duodenuml with ulcerated (C) and atrophic mucosa in jejunum (D)..

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