A not-so-innocent kiss

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Case presentation

A 64-year-old man was admitted to the hospital with a 3-day history of fever. Two months earlier, he had experienced a similar ten-day episode of fever and nightly sweating during a trip to Hungary. Apart from tiredness and subtle weight loss over the last year, the patient was symptom-free. Relevant medical history included gout and a past infection with Epstein-Barr virus (EBV). The patient was febrile, 38°C, but further clinical examination was completely normal. Laboratory findings showed an elevated white blood cell count of 13.250/µL [4200 - 9800/µL] and a CRP of 190mg/L [0 - 3.2mg/L]. On CT scanning of the abdomen, a 5 cm long pathological wall thickening of the terminal ileum was noted [Figure A, arrow]. Colonoscopy confirmed the abnormal mucosal lining of the terminal ileum, showing a whitish polypoid nodularity [Figure B]. Biopsies were taken for histopathological examination [Figure C, Hematoxylin-eosin staining; Figure D, CD30 staining; Figure E, EBER staining].

What is the diagnosis?

A) Terminal ileitis caused by Crohn's Disease
B) Nontyphoidal Salmonella ileitis
C) MALT (mucosa-associated lymphoid tissue) lymphoma of the terminal ileum
D) Hodgkin Lymphoma
E) Primary intestinal lymphangiectasia

Diagnosis

Answer D. The endoscopic and histopathological features were compatible with EBV-related Hodgkin Lymphoma. Histopathological examination showed mucosal infiltration of atypical cells with an abundance of eosinophilic cells and the presence of Reed-Sternberg cells [Figure C, arrow], which stained positive for CD30 [Figure D], MUM1 and EBER [Figure E], partially positive for CD15 and CD79a and negative for CD3, CD20, CD68, LCA and broad-spectrum cytokeratin. The lesion was diagnosed as a mixed cellularity, primary extranodal, classical Hodgkin Lymphoma (cHL). The positive EBER staining assumed a past infection with EBV as underlying etiology of the disease. On PET scanning, no further extra-intestinal involvement was observed, and bone marrow biopsy turned out negative. Classical Hodgkin lymphoma generally presents with peripheral lymphadenopathies and frequently invades the gastro-intestinal tract at a later stage. Primary extranodal cHL, however, is extremely rare, and is observed in less than 1% of Hodgkin Lymphoma cases. Risk factors are inflammatory bowel disease, past infection with EBV, as was present in this case, and a context of immunosuppression, e.g. a post-transplant status.(1,2) However, these conditions do not need to be fulfilled. The diagnostic criteria for primary gastrointestinal cHL were met, being (a) the presence of a predominant gastrointestinal lesion, (b) the absence of peripheral lymphadenopathies, and (c) the absence of liver, bone marrow and spleen involvement.(2,3) The patient was started on Adriamycin-Bleomycin-Vinblastine-

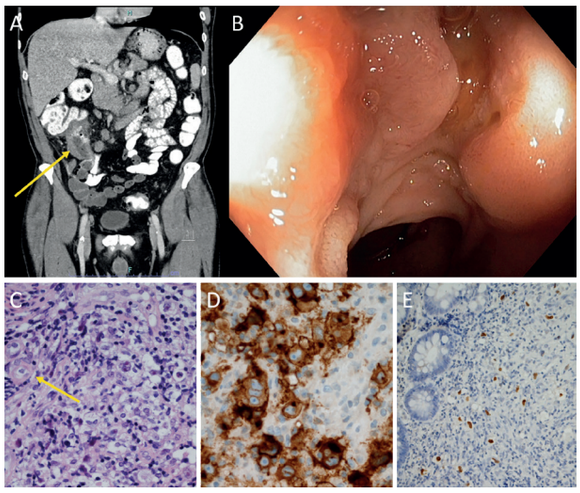

Figure 1. — Radiographic, endoscopic and histopathological features of the disease. Panel A, CT abdomen, coronal plane ; Panel B, colonoscopic image of the terminal ileum ; Panel C, Hematoxylin-eosin staining (x20 magnification) showing lymphoid hyperplasia, irregular and prominent nucleoli, and an abundance of eosinophilic cells and histiocytes, arrow indicating Reed-Sternberg cell ; Panel D, CD30-staining (x20 magnification), positive cytoplasmatic staining pattern; Panel E, EBER-staining (x20 magnification), positive nuclear staining pattern.

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Dacarbazine (ABVD) twice-weekly, with follow-up PET scanning after every 2-4 cycles of chemotherapy.

Contributor statement

JB, CVdB and PVH did clinical assessment and investigation. PVH was responsible for care of the patient. MAVC was responsible for histopathological analysis. CVdB wrote the draft of the report, and JB and PVH revised the report. All authors approved the final version.

Competing interests

None declared.

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