Intravascular Large B-Cell Lymphoma Within Appendix Presenting As Acute Abdomen: A Challenging Diagnosis for Hematologists

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To the Editor,

Intravascular large cell lymphoma (ILCL) is a rare subtype of non-Hodgkin lymphoma characterized by the proliferation of lymphoma cells within the lumen of small vessels. The clinical diagnosis of ILCL is challenging due to the absence of an obvious lymphadenopathy or detectable mass (1). The disease is usually diagnosed post-mortem or incidentally in patients with diverse signs and symptoms related to organ dysfunction caused by occlusion of blood vessels (2). Herein, we report a case of intravascular large B cell lymphoma (ILBCL) presenting as acute abdomen. The diagnosis was rendered following the histopathologic examination of the appendectomy specimen. To our knowledge, this is the first report of ILCL of B-cell origin with appendix involvement.

58-year-old male patient was referred to our hospital to investigate the etiology of pancytopenia. His laboratory tests demonstrated values of Hb 10.9 g/dL, WBC 800/uL, platelet 48.000/uL, LDH 625 U/L. Bone marrow biopsy revealed dyshematopoiesis, especially in megakaryocytic and granulocytic cell lines, hypercellular bone marrow including patchy CD20+ B-cell infiltration in the peritrabecular zone. Intravascular pattern of infiltration was not prominent. Cytogenetic analysis resulted as complex karyotype, and FISH analysis showed trisomy 7 and 21. B-cell lymphoid infiltration in the bone marrow was identified as hematogone-like atypical B-cell proliferation, but further work-up including PET/CT was planned for the differential diagnosis of lymphoproliferative disorders. On the 15th day of his initial admission, the patient suddenly developed abdominal pain. Upon finding rebound tenderness and dilated appendix with thickened, hyperenhancing wall on CT scan, the patient was consulted to the general surgery department and underwent emergent surgery.
Laparoscopic appendectomy with suspicion of acute appendicitis. In gross examination, the appendix was within normal limits in size and shape. The histopathological investigation of appendix was reported as “high-proliferative, neoplastic B-cell infiltration in the vascular structures at the wall and serosa of the appendix” (Figure 1). Histological findings of acute appendicitis were absent. Meanwhile, PET-CT which was performed prior to surgery was reported as “splenomegaly, hypermetabolic lesion on cecum at right hemipelvis (abscess?), intermediate hypermetabolism at axial skeleton and bilateral humerus-femur” after the operation. In light of these findings, the patient was diagnosed with “intravascular large B-cell lymphoma” and given 6 cycles of R-CHOP and intrathecal methotrexate for central nervous system prophylaxis. Bone marrow biopsy after 4 cycles of R-CHOP showed normal cytogenetics without any lymphocyte infiltration and complete metabolic remission detected on interim PET-CT. Follow-up of the patient has been uneventful 12 months after the completion of treatment.

Although rare cases of ILBCL of the gastrointestinal tract have been previously described (3-6), to our knowledge there have been no reports of ILCL of B-cell origin with appendix involvement in the English literature. The only reported case of ILCL involving the appendix was of T-cell origin and described by Malicki et al in 1999 (7). In 2004, gastrointestinal involvement has been reported as 8% (3 cases) in a series of 38 patients with ILCL by Ferreri et al. (8) Since then, there is limited case reports, in which the gastrointestinal tract was the primary diagnostic site of ILBCL; three in the colon, two in the gastroduodenum, two in the duodenum, two in the ileum and one in the stomach (3-6). Treatment of ILBL includes both systemic therapy with anthracycline based regimens and therapy directed at the central nervous system (CNS) since CNS involvement is frequent (9). Clinical outcomes for patients with ILBCL have improved in the rituximab (R) era. In the largest retrospective analysis, higher rates of overall and progression free survival have been reported in patients treated with R- chemotherapy compared to chemotherapy alone (66% and 56% vs 46% and 27%, respectively) (10). Our patient is the first presentation of ILCL of B-cell origin which was diagnosed by an appendectomy. Early diagnosis on appendectomy and timely initiation of appropriate treatment allowed the successful response in this case. As a take-home message, late-onset acute appendicitis should require careful histological examination to exclude associated systemic disease such as inflammatory bowel disease, pseudomyxoma or lymphoma, as in the presented case.

**Keywords:** Intravascular large B-cell lymphoma, Appendix

**References**


Figure 1. A-B. Infiltration of lymphoid cells in a small and large vessel in the periapendiceal area (H&E). C-D. CD20 immunohistochemistry, highlighting the B-cell phenotype of the neoplastic cells (CD20).