INTRAVASCULAR LARGE B-CELL LYMPHOMA: A REPORT OF TWO CASES

A Lapus, M Swaby, A Richmond, J Nguyen, M Condron, X Duan, M Uthman, M Wang, ND Nguyen, R. Brown, L Chen
Department of Pathology & Laboratory Medicine.
The University of Texas Health Science Center--Houston Medical School

The University of Texas
Medical School at Houston
A part of The University of Texas Health Science Center at Houston

BACKGROUND

Intravascular large B-cell lymphoma (IVL) is a variant of extranodal diffuse large B-cell lymphoma with the malignant cells essentially confined to small vessel lumina. Clinical signs are protean and the histology can be subtle, leading to a delayed diagnosis with increased mortality potential. We present two patients with IVL:
(1) Presenting as acute cholecystitis and unexplained anemia and
(2) As suspected CNS autoimmune vasculitis.

DESIGN

Case 1: The patient is a 43 year old woman presenting with abdominal pain and anemia; a cholecystectomy was performed for suspected cholelithiasis. A grossly unremarkable acalculous gallbladder was received for routine processing. Wright-stained peripheral blood smears were obtained for anemia workup. Immunophenotypic analysis by flow cytometry was also performed on peripheral blood.

Case 2: The patient is a 69 year old man presenting with episodic loss of consciousness, right lower extremity weakness, slurred speech, and personality changes. Imaging studies were suggestive of CNS vasculitis and a biopsy was performed. A frontal lobe biopsy was received for routine processing.

H&E sections and a panel of immunohistochemiscal stain were obtained for both of cases.

RESULTS

Case 1: H&E sections showed multiple small vessels filled with aggregates of medium to large atypical lymphoid cells with increased mitoses (fig 1A, 1B). Co-expression of CD20/CD5 was demonstrated by immunostain (fig 1C). Proliferative index by Ki-67 was 80-90% (200X, 1D). The lymphoma cells were negative for Cyclin-D1, CD3, TdT (fig 1A-E), and cytokeratin. There were lymphoma cells in peripheral blood (fig 1G), confirmed by flow cytometric analysis (fig 1H, 1I).

Case 2: H&E sections showed clusters of large atypical lymphoid cells within small vessels, with CD20 expression (fig 2A, 2B). They were negative for CD3 (fig 2C), CD7, TTF-1, pancytokeratin, synaptophysin and GFAP. Lymphoma cells were also present in aspirate smears (photo not shown).

Pathologic Findings-Case One

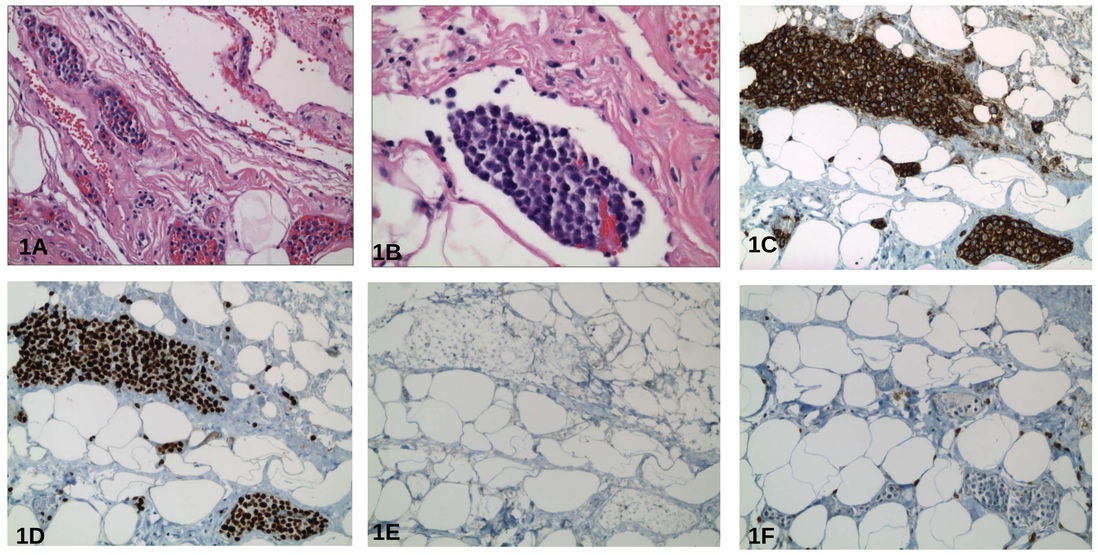

Fig 1A-F: Intravascular lymphoma of gallbladder. H&E section showed large atypical lymphoid cells within the vascular lumens (200X, 400X. 1A-B). Lymphoma cells were positive for CD20 (1C) (200X) and CD5 (200X, 1D). Lymphoma cell were negative for CD3 (200X, 1E) and TdT (200X, 1F). .

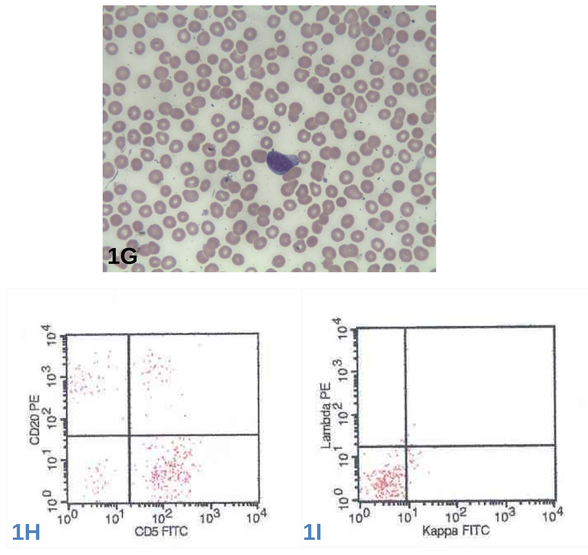

Fig 1G-I. Atypical large lymphoid cells in peripheral blood smear (400X. 1GA). Flow cytometry showed co-expression of CD20/CD5 (1H), and lack of light chains expression (1I).

Pathologic Findings-Case Two

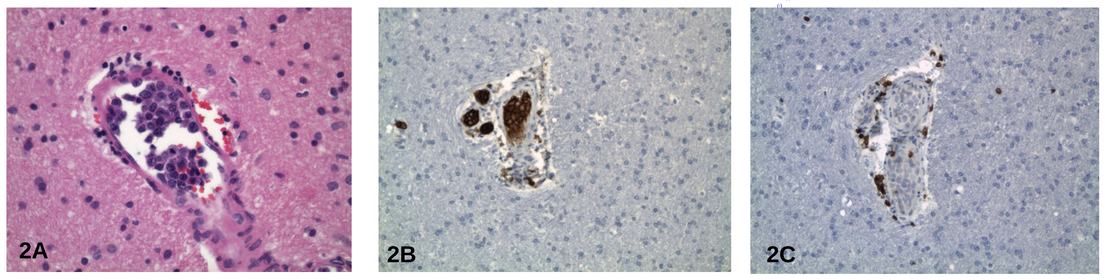

Fig 2A-C: Intravascular lymphoma of the brain. H&E showed large atypical lymhpoid cells within the vascular lumen (200X, 2A). The lymphoma cells were positive for CD20 (200X, 2B) and negative for CD3 (200X, 2C).

CONCLUSIONS

Highly variable clinical presentations plus subtle pathologic changes may contribute to a delayed diagnosis and poor outcome in IVL patients. In our two cases, the most characteristic findings are dilated small vessels with lymphoid cells. However, in our second patient, these changes were subtle, owing to the paucity of blood vessels in the biopsy. The presentations of 'cholecystitis' and 'vasculitis' were likely due to vascular obstruction by malignant cells. Significantly, IVL disseminated into the peripheral blood is rarely seen, but was present in our first patient.