Lymphology 29 (1996) 166-169

PRIMARY NON-HODGKIN LYMPHOMA PRESENTING AS ILEOCOLIC INTUSSUSCEPTION

P.H. Gerst, M. Niazi, V. Narasimha, R. Saraiya, E. Albu

Departments of Surgery (PHG,VN,RS,EA) and Pathology (MN), Bronx-Lebanon Hospital Center, Bronx, New York USA

ABSTRACT

We report the case of an HIV-seropositive patient with non-Hodgkin lymphoma of the small intestine who presented with an ileocolic intussusception. This lesion fulfilled the diagnostic criteria for primary gastrointestinal lymphoma. Such a neoplasm in an immuno-compromised patient is usually more aggressive and less responsive to treatment than in an HIV-seronegative patient.

Primary gastrointestinal lymphoma is rare, constituting 5% of all lymphomas and 1 to 4% of all malignancies of the alimentary tract (1). Only 1 to 2% of all primary gastro-intestinal malignancies arise in the small intestine; of these, fewer than 30% are lymphomas (2). Common extra-nodal sites of involvement include the central nervous system, bone marrow, bowel, skin, lung, and liver (3). Malignant lymphoma has been recognized as a manifestation of the acquired immunodeficiency syndrome (4). HIV-seropositive patients tend to develop atypical and aggressive lymphomas.

CASE REPORT

A 42-year-old man developed colicky right-sided abdominal pain of four days duration associated with bilious vomiting. He had lost considerable amount of weight and was very weak. Five years earlier, at the time of admission for a gunshot wound of the abdomen, he had tested negative for human immunodeficiency virus (HIV).

On admission he was severely dehydrated and hypotensive. He had marked distention of the abdomen and visible peristalsis. Direct and rebound tenderness were present on the right side of the abdomen. The patient's stool was guaiac-positive. He had a moderately elevated serum transaminase level but a normal white blood cell count. Abdominal radiographs showed markedly distended loops of small bowel with multiple air-fluid levels. A CT scan demonstrated a mass in the right lower abdomen suggesting an ileocolic intussusception *(Fig. 1A,B)*.

After resuscitation, laparotomy disclosed 2.5 liters of straw-colored ascitic fluid, a hard macronodular liver, and a 14x7 cm mass representing an irreducible ileocolic intus-susception *(Fig. 1C)*. The mesentery of the ileum proximal to the intussusception contained several enlarged lymph nodes. A second large mass was located in the ileum *(Fig. 1D)*. An ascending segmental colectomy with ileal resection and end-to-end anastomosis was performed 10 cm proximal to the second ileal mass.

After operation, the patient's liver function continued to deteriorate and despite aggressive treatment he died two weeks later. Histopathologic examination of the mass that had led to the intussusception showed non-Hodgkin lymphoma of the diffuse, large-cell

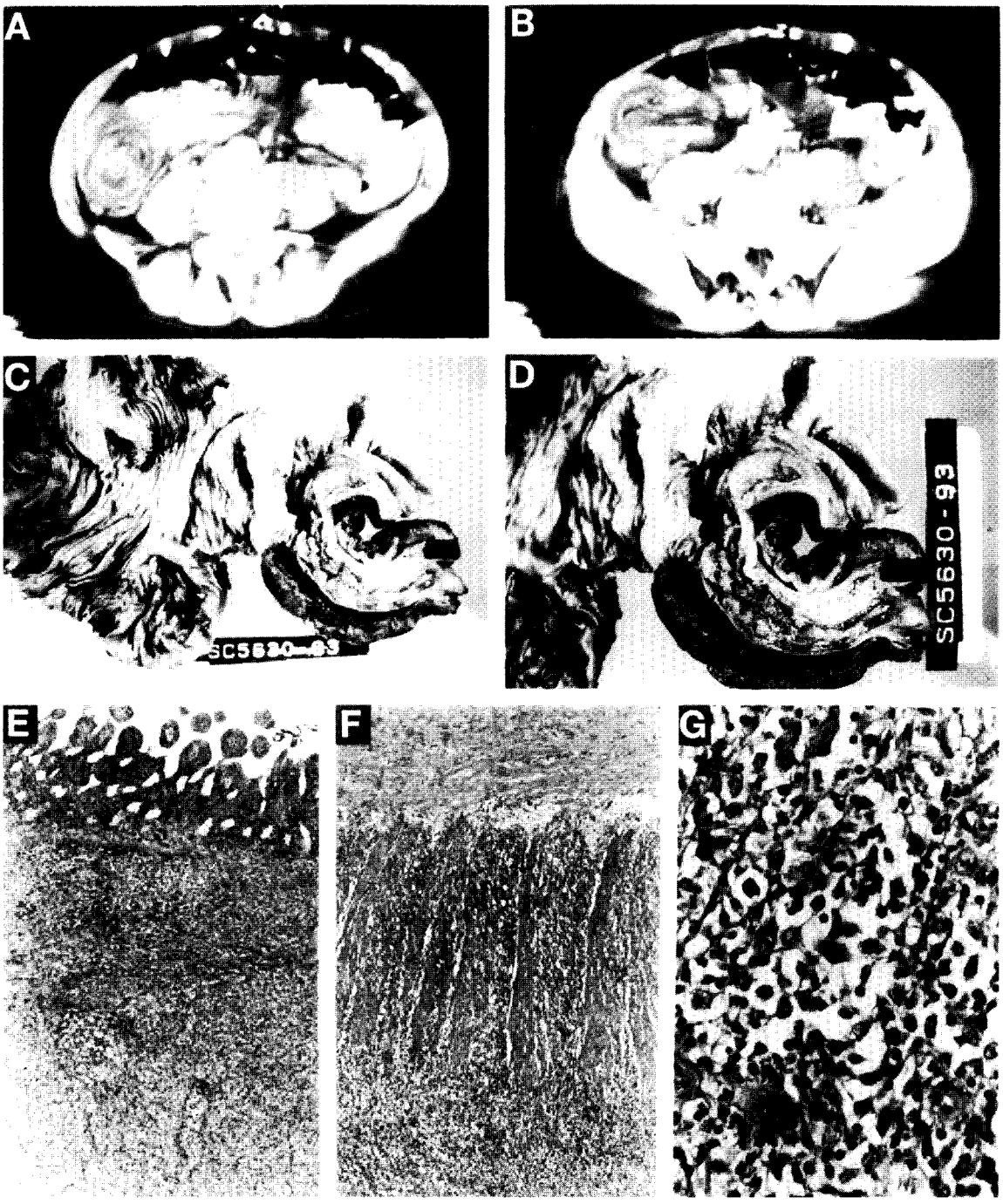

Fig. 1. HIV-seropositive patient with non-Hodgkin lymphoma presenting as an ileocolic intussusception (A,B). Computer tomographic scan of abdomen showing ileocolic intussusception (arrows) (C,D). Sagittal section of the resected specimen showing the ileocolic intussusception (E,F). Photomicrograph (low power) showing lymphomatous infiltrate involving the mucosa and wall of ileum (G). Photomicrograph (high power) showing large cells, immunoblastic type.

type *(Fig. 1E,F,G)*. Immunoblastic immunophenotypic studies suggested a B-cell lymphoproliferative disorder. Serum antibody testing after the patient's death was positive for HIV.

COMMENT

HIV-positive patients are at high risk of non-Hodgkin lymphoma (3). Lymphoma occurs in 4 to 10% of HIV-infected individuals and often is of the B-cell type (5). There is no gender predilection (6). Characteristically, lymphoma in an HIV-seropositive patient demonstrates high-grade histology, rapid growth, widespread involvement, poor response to chemotherapy, and short duration of patient survival (5).

A high incidence of malignant tumors has long been recognized in patients with primary immune defects such as X-linked lymphoproliferative and Wiskott Aldrich syndromes. Such tumors are a major feature of patients with acquired immunodeficiency, although no direct etiological association has been established between lymphoma and a specific agent.

Studies have shown that HIV sequences are absent from the clonal B-cell expansions in hyperplastic lymphadenopathy associated with AIDS-related complex (ARC). This finding suggests that HIV is not directly responsible for the malignant transformation (7). The role of Epstein-Barr virus (EBV) transformed B cell clones in the genesis of lymphoma in AIDS patients has been strongly suggested. The state of HIV-induced immunosuppression and massive EBV infection in ARC appears to favor the expression of multiple EBV-transformed B cell clones. This feature is thought to increase the probability of additional genetic alterations, specifically translocation of the C-myc gene, resulting in malignant transformation of these B cells. In support of this hypothesis, Knowles et al (8) have shown that introduction of an activated C-myc gene into EBV-infected lymphoblasts from patients with AIDS leads to their malignant conversion.

Since the beginning of the AIDS epidemic, there has been a significant increase in the incidence of non-Hodgkin lymphoma within the at-risk population, including homosexuals, drug abusers, and hemophiliacs (9).

Upper gastrointestinal series and abdominal computer tomographic scan have a high degree of diagnostic sensitivity. Use of pan-endoscopy increases the likelihood of accurate diagnosis to more than 90% (5). Differential diagnosis of terminal ileal disease in patients with AIDS should include lymphoma, mycobacterium avium intracellulare, and Kaposi sarcoma. Estrin et al (9) conclude that endoscopic bowel evaluation of AIDS patients is a useful diagnostic modality that may obviate the need for laparotomy.

Management of non-Hodgkin lymphoma in immunocompromised patients still has not been properly defined. The prognosis is poor irrespective of the modality of treatment. The overall five-year survival of patients with gastrointestinal lymphoma is 45%. This percentage declines to only 28% in patients with small bowel lymphoma and to 14% when such lymphomas involve regional lymph nodes (4). Ziegler et al (3), in a multi-institutional study of 90 homosexual men with non-Hodgkin lymphoma, found a 53% response rate to either chemotherapy and/or radiotherapy, but with a relapse rate of 54%. Moreover, the overall morbidity and mortality in this study was 91%.

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Paul H. Gerst, M.D.
Department of Surgery
Bronx-Lebanon Hospital Center
1650 Selwyn Avenue, Suite 4A
Bronx, New York 10457 USA