Acute febrile illness and a splenic mass in a 15-year-old West African immigrant

Jennifer Ahmed MA¹, Timothy Mailman MD^{2,3,4,5}, Joanne M Langley MD MSc^{2,5,6}

Within 24 h of arrival from West Africa, a 15-year-old boy presented to the emergency department with periumbilical abdominal pain, intermittent bloody diarrhea, vomiting and fever of three days duration. He had lived in refugee camps for over 10 years. His physical examination was remarkable for fever (40.7°C), thin body habitus (height and weight below the fifth percentile for age), scleral icterus and hepatosplenomegaly (liver palpable 2 cm below the right costal margin, span 11 cm; spleen tip 5 cm below the left costal margin). The abdomen was somewhat tender to palpation but soft, and his bowel sounds were normal.

Investigations showed rare malaria parasites in the peripheral blood smear; numbers were insufficient for speciation. Stool was negative for *Salmonella*, *Shigella*, *Escherichia coli* 0157, *Campylobacter*, *Aeromonas* and *Yersinia*. Serology results for *Leptospira*, hepatitis A and HIV were negative. Results for hepatitis B surface antigen were positive. Cultures of blood and urine showed no growth. Serology for Epstein-Barr virus (EBV) showed the presence of immunoglobulin G to viral capsid antigen, but no immunoglobulin M was identified. A single-step Mantoux test was negative.

Imaging of the liver and spleen showed a normal-looking liver, but revealed the presence of three splenic masses. Computed tomography identified two cystic lesions diagnosed as peripheral splenic infarcts, and one large noncystic mass

Figure 1) Computed tomography of the abdomen of a 15-year-old male immigrant from West Africa with hepatosplenomegaly and multiple splenic lesions

that seemed to originate in the spleen and extend outward (Figure 1).

A spleen biopsy was performed. What is the diagnosis?

¹Faculty of Medicine; ²Departments of Pediatrics; ³Pathology; ⁴Microbiology and Immunology, Dalhousie University; ⁵IWK Health Centre;

⁶Department of Community Health and Epidemiology, Dalhousie University, Halifax, Nova Scotia

Correspondence: Dr Joanne M Langley, Canadian Center for Vaccinology, 5850 University Avenue, PO Box 9700, Halifax, Nova Scotia B3K 6R8. Telephone 902-470-8498, fax 902-470-7232, e-mail jmlangle@dal.ca

Received and accepted for publication January 24, 2008

DIAGNOSIS

Pathology of the splenic biopsy showed focal areas of necrosis and apoptotic tumour cells. Immunoperoxidase staining confirmed these cells to be B lymphocytes, and karyotyping revealed a reciprocal translocation between the long arms of chromosomes 8 and 22. These findings were consistent with a diagnosis of Burkitt's lymphoma (BL).

DISCUSSION

The differential for the splenic mass included bacterial, fungal or helminthic abscess, tuberculous granuloma and malignancy. All cultures of the splenic lesion were sterile, and no parasites were seen. Biopsy findings were consistent with a diagnosis of BL.

BL is a form of non-Hodgkin's lymphoma (1) that exists in three forms (endemic, sporadic and HIV-related), which share common cytomorphology but different clinical features (2). The endemic form is found in tropical Africa and Papua New Guinea (3). Sporadic BL occurs worldwide. Endemic BL is primarily a disease that occurs in children; it is the most common childhood cancer in tropical Africa, although it can be seen in adults (usually younger than 30 years of age).

Although sporadic BL mainly involves tumours of the abdomen (the ileocecal region) (1), endemic BL in Africa is associated with tumours of the jaw. Associated clinical features of endemic BL in children may include failure to thrive, abdominal swelling and/or pain, fever, proptosis, ascites, splenomegaly and/or hepatomegaly (3). HIV-related BL typically presents

with abdominal involvement and is diagnosed more often in adults than in children (4). In addition to its association with HIV, BL has also been linked with EBV and malaria (1,5,6). A single EBV gene, EBNA-1, is usually expressed in tissue from African BL patients (6). Incidentally, EBV was first discovered in tissue from BL patients (7). Malaria is thought to decrease regulation of T cells infected with EBV and, thus, enhance B cell growth.

Our patient's presentation did not fit the classical descriptions of BL. In addition, the initial presentation was suggestive of an infectious etiology, and asymptomatic infections with both malaria and hepatitis B were indeed found. A wide range of symptomatic and asymptomatic infections should be considered in ill immigrants (8).

REFERENCES

1. Wright DH. What is Burkitt's lymphoma? *J Pathol* 1997;182:125-7.
2. Kessel B, Moisseiev E, Korin A, et al. Burkitt's lymphoma: An unusual cause of obstructive jaundice. *Asian J Surg* 2005;28:301-4.
3. Meremikwu MM, Ehiri JE, Nkanga DG, Udoh EE, Ikpat O, Alaje EO. Socioeconomic constraints to effective management of Burkitt's lymphoma in south-eastern Nigeria. *Trop Med Int Health* 2005;10:92-8.
4. van den Bosch CA. Is endemic Burkitt's lymphoma an alliance between three infections and a tumour promoter? *Lancet Oncol* 2004;5:738-46.
5. Jarrett RF. Viruses and lymphoma/leukaemia. *J Pathol* 2006;208:176-86.
6. Cohen JL. Epstein-Barr virus infection. *N Engl J Med* 2000;343:481-92.
7. Epstein MA, Achong BG, Barr YM. Virus particles in cultured lymphoblasts from Burkitt's lymphoma. *Lancet* 1964;15:702-3.
8. Canadian Pediatric Society. *Children and Youth New to Canada: A Health Care Guide*. Ottawa: Canadian Pediatric Society, 1999.