Hemophagocytic Syndrome: An Unusual Manifestation of Acute Human Immunodeficiency Virus Infection

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Abstract
An 18 year-old heterosexual man was hospitalized because of fever, chills, a sore throat, and a dry cough for 8 days. He had sexual intercourse with a new partner within the 4 months prior to admission. At admission, the patient presented a clinical picture compatible with hemophagocytic syndrome (HPS). The presence of hemophagocytosis was subsequently determined pathologically from bone marrow and lymph node specimens. An exhaustive diagnostic work-up failed to reveal any causative etiology, the symptoms improving after 2 doses of intravenous immunoglobulin (IVIG) infusion, given at a dose of 0.5 mg/kg each, the 2 doses being administered 1 week apart. Three months subsequent to the patient’s initial presentation, acute human immunodeficiency virus (HIV) infection was diagnosed, and the patient received highly active antiretrovirus therapy (HAART) from the time of diagnosis. The patient remained well for the following 2 years. HPS in the advanced stages of HIV infection has previously been described, but HPS during seroconversion of an acute form of the infection is rare. We most definitely suggest, however, that acute HIV infection be included in the list of potential causes of HPS. IVIG therapy appears to be an appropriate therapeutic modality, and HAART also is effective, for prevention of recurrence of HPS in a patient with acute HIV infection. Int J Hematol. 2003;78:450-452.

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1. Introduction

Hemophagocytic syndrome (HPS) has previously been described in association with a number of diseases [1] and has been recognized as being involved in the course of human immunodeficiency virus (HIV) infection [2,3]. It would appear, however, that most of the reports of HPS revealed that these cases occurred in the advanced stages of HIV infection, at which time patients also suffered from concomitant infectious or malignant diseases [2,3]. Therefore, the role of HIV as a causative agent of HPS remains to be determined. We describe the case of a patient in whom HPS occurred during the acute stage of HIV infection. It appeared that no other etiology, apart from HIV infection, could be implicated as the causative agent.

2. Case Report

An 18-year-old heterosexual man visited our hospital reporting 2 days of fever, chills, sore throat, and dry cough. The initial physical findings revealed only an injected throat. Subsequent blood count showed a white blood cell (WBC) count of 9.7 × 10^9 cells/L, a hemoglobin level of 16.2 g/dL, and a platelet count of 184 × 10^9/L. The patient was discharged on the same day with prescriptions for symptomatic treatment. Six days later, blurred vision developed in addition to persistence of the previous symptoms. At second presentation, the patient was conscious and apparently well oriented. His blood pressure was 83/42 mmHg, pulse rate was 87 beats/minute, respiratory rate was 20 breaths/minute, and the highest oral temperature was 40°C. Remarkable findings at physical examination were enlarged tonsils coated with purulent exudate. The WBC count dropped to
measured 2.8 × 10^9 cells/L, with 74% neutrophils, 20% lymphocytes, and 6% monocytes. Empirical antibiotic with amoxicillin/ clavulanate was administered, and the patient was admitted. When the history was traced, there appeared to be nothing particular apart from the patient’s having participated in sexual intercourse with a new partner during the preceding month.

When the history was traced, there appeared to be nothing particular apart from the patient’s having participated in sexual intercourse with a new partner during the preceding month. During hospitalization, a spiking fever persisted, and generalized lymphadenopathy developed bilaterally over the neck and the axillary and inguinal areas. The largest lymph node was detected was located over the left inguinal region and neck and the axillary and inguinal areas. The largest lympheralized lymphadenopathy developed bilaterally over the inguinal region and neck. The patient received highly active antiretroviral therapy (HAART). No recurrence of HPS was found in the subsequent 2 years.

### 3. Discussion

HPS was first described by Risdall et al in 1979 as an immune disorder induced by various viral infections among immunocompromised patients [4]. Since that report, many other infectious diseases and illness etiologies have been reported to have been diagnosed in association with HPS.