Case Report

A Rare Case of Pleural Effusion Burkitt’s Lymphoma with Tumor Lysis Syndrome in an HIV, HHV8, EBV, and HCV Negative Young Male Patient

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Abstract

We report an unusual case of a 25 year-old male with Burkitt’s like disease associated with Primary Effusion Lymphoma (PEL), who was negative for HIV, HHV8, HCV, and EBV. The pleural effusion contained lymphoma cells with B-cell lineage and c-myc gene rearrangement. The patient had a rapidly progressive and fatal course and died from tumor lysis syndrome. This is the second reported case in the literature of such unique characteristics. Further understanding of the molecular and pathogenetic mechanisms underlying this disease is warranted.

Case Report

A 25 year-old previously healthy Caucasian male presented with 3 months of progressive shortness of breath, 45 pound weight loss, night sweats and generalized weakness. Symptoms persisted despite 3 courses of antibiotics. There was no family history of malignancy. He denied a history of IV drug use or being sexually active. On exam he was afebrile and had an oxygen saturation of 97% on 4 liters nasal canula. There were decreased breath sounds at the left base. There was moderate pitting edema of both legs. Laboratory evaluation revealed hemoglobin 16.0 gm/dL, WBC 6.6, platelet count of 118,000, prothrombin time of 22.9, INR of 1.9, total bilirubin of 2.8, direct bilirubin of 1.1, AST 158, ALT 105, bicarbonate 13, anion gap of 21, BUN/creatinine of 49/2.4. A hepatitis panel was non-reactive, mononucleosis screen was negative, and HIV and Quantiferon gold for tuberculosis were negative.

An ultrasound of the lower extremities revealed a right popliteal DVT. A non-contrast CT of the chest, abdomen, and pelvis showed a moderate to large left pleural effusion, absence of the right kidney, and mild to moderate ascites (Figure 1).

Thoracentesis of the left pleural effusion showed neoplastic B-cells with strong positive CD20 staining, focal positive CD10 staining, and negative CD99 staining consistent with a B-cell lymphoma. Fluorescence in-situ hybridization (FISH) results were positive for a rearrangement involving MYC (77.5% of cells) and positive for a MYC/IGH rearrangement (76.5% of cells). These findings were characteristic of Burkitt’s lymphoma. An immunoperoxidase stain for HHV8 was negative. Bone marrow biopsy showed orderly hematopoiesis in all cell lines with no...
Primary effusion lymphoma (PEL) is a rare type of Non-Hodgkin’s lymphoma [1]. It was first reported in immunodeficient, Human immunodeficiency virus (HIV) patients [2]. PEL usually involves body cavities, however involvement of bone marrow and lymph nodes has been described [3]. It represents 4-5% of all AIDS-related lymphomas [4].

Discovery of HHV-8, or Kaposi’s sarcoma-associated herpes virus DNA sequences in AIDS-related lymphomatous effusions helped differentiate PEL from other types of lymphoma [5]. However, later on PEL was also reported in patients who were HIV-negative but HHV-8 positive [6] suggesting strong etiologic basis of this gamma virus for PEL. Epstein bar virus (EBV) has been reported to be strongly associated with PEL associated in patients who are HIV-positive [4,6].

Nador et al reported HIV-positive PEL patients who were HHV-8 negative but had c-myc gene rearrangements and had morphologic and cytologic features of Burkitt’s or Burkitt’s like lymphoma [7].

There now have been several reports of PEL now occurring in HIV-negative, HHV-8 negative patients [8,9]. Four cases of HIV-negative, HHV-8 negative, PEL have been associated with Hepatitis C virus (HCV) infection [10,11]. HCV is a hepatotropic virus however the pathogenetic mechanism underlying association with PEL remains unclear.

In conclusion, we report a rare case report of a young patient with Burkitt’s related PEL who was negative for HIV, HHV8, EBV and HCV raising the possibility that there may be yet unidentified viruses which may be involved in the pathogenesis of this disease. Alteration in the c-myc gene may also activate molecular pathways leading to malignant lymphomatous effusions. Furthermore, signal transduction abnormalities, cellular adhesion molecule, and continuous antigenic stimulation as well as regional, geographic and other epidemiologic factors are involved in development of HHV8+, HIV-, EBV-, HCV-, c-myc altered development of Burkitt’s related PEL.

REFERENCES
