Introduction

Primary effusion lymphoma (PEL) is a rare type of non-Hodgkin’s lymphoma, which is characterized by serous effusion in body cavities without detectable tumor masses or lymphadenopathy. It is usually found in human herpes virus 8 (HHV8) and human immunodeficiency virus (HIV) positive patients, and tumor cells usually show a B-cell–lineage immunophenotype.

Case presentation: In the current case, a 44-year old male patient suffering from shortness of breath was hospitalized. He was diagnosed with pleural effusion on the right side. Following a cytological examination of pleural fluid, it was revealed that the patient had high-grade T-cell lymphoma. CD3, CD16, CD8 were positive; whereas CD20, CD30, and CD56 were negative; Ki67 was close to 100 %. HHV8 and HIV were negative, Epstein-Barr virus (EBV) was positive.

Discussion: The disease had an aggressive progression, and the patient was deceased the day after the diagnosis. The current case indicates an atypical PEL case due to its aggressiveness, which had T-cell phenotype with HHV8 negative, HIV negative and EBV positive.

Conclusion: Atypical PEL should be taken into consideration when dealing with aggressive PEL cases.

Key words: Primary effusion lymphoma, T-cell, EBV.

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In the current case, an HHV8-unrelated PEL-like lymphoma with T-cell phenotype, which shows HIV negativity, EBV positivity, and aggressive progression, is reported.

Case presentation

A 44-year-old male was admitted to the hospital with fatigue, night sweating, coughing, shortness of breath, and abdominal swelling. The patient had a 60 pack/year history of smoking, and was diagnosed with chronic obstructive lung disease one year before. The physical examination showed no respiratory sounds in the basal level of the right lung and advanced abdomen distension. Chest X-ray and computed tomography (CT) showed pleural effusion on the right side (Figures 1,2). At the same time, pericardial effusion was present in the heart.
CT and abdominal ultrasound scans showed abdomen widespread ascites and omentum thickening. The patient did not have any mass or lymphadenopathy. Blood tests results were normal (Table 1).

The patient underwent paracentesis and transudative ascites was taken. An omentum biopsy was performed showing chronic inflammation without caseification. The patient underwent thoracentesis and serofibrinous fluid was taken. The adenosine deaminase (ADA) level was higher in the pleural fluid (ADA: 60 IU/L). The patient’s clinical and laboratory findings initially suggested tuberculosis peritonitis, and an antituberculosis treatment was initiated. After cytological examination, the patient’s pleural fluid was interpreted as “high-grade T-cell lymphoma” (Figure 3).

The immunohistochemical examination showed CD3 (Figures 4), CD16, and CD8 expression, while TdT, CD20, CD30, CD56, and pan-cytokeratin expression was not present. Ki67 index was close to 100 % (Figure 5).

The immunohistochemical examination of the pathological specimens showed that HHV8 was
negative. Serological examination showed that HIV and HCV were negative, whereas EBV was positive. The patient had massive pleurisy, widespread ascites, and pericardial effusion at the same time, and the patient's general condition deteriorated gradually. Despite thoracentesis, the patient’s pleurisy continued to increase. The patient developed acute respiratory distress syndrome and finally severe respiratory failure. He was intubated and taken to the intensive care unit. However, the patient did not survive despite all interventions. It took two weeks to make the diagnosis. The patient was deceased one day after the diagnosis. A specific treatment was not started.

Discussion

PEL was identified as a distinct entity for the first time by Nador et al. in 1996(5). It was classified among B-cell lymphomas according to the WHO classification in 2001(6). According to the WHO classification in 2008, PEL was classified as a subgroup within the diffuse large B-cell lymphomas (DLBCL)(7).

The association between PEL and HHV8 is common, and HHV8 is believed to play a major role in PEL pathogenesis. HHV8 is a type of virus that plays a role in the aetiology of Kaposi’s sarcoma, PEL, and Castleman’s disease(8,9). Latency-associated nuclear antigen (LANA-1) is a latent protein of HHV8 and also one of its important antigens, and the antibodies produced against HHV8 indicate the presence of this virus. LANA-1 has been shown to bind to tumor suppressor proteins p53 and retinoblastoma in PEL cells to inactivate them, thus inhibiting apoptosis(10,11). HIV positivity is common in HHV8-related PEL cases, and these patients are generally young or middle-aged homosexual or bisexual male patients(1,2,4,5). HCV and EBV positivity is also seen in PEL, and these viruses are believed to play a role in the disease pathogenesis(5,12). PEL can also be seen in patients with a suppressed immune system, who underwent organ transplantation, and those who have been diagnosed with cirrhosis(5).

PEL is a disease with a poor prognosis, especially in HIV-positive patients. Median survival is 4 months, while one-year survival is 17 % in HIV-positive patients(5). On the other hand, HHV8-unrelated PEL-like lymphoma has a slightly better prognosis compared to PEL, and the median survival is 6-10 months, whereas one-year survival is 35%(5,6,7,13,24).

The disease had an aggressive progression in the current case. The patient was deceased one day after the diagnosis, without any specific treatment. We believe that high Ki67 expression (Ki67 index was close to 100 %) was also responsible for this outcome.

Adiguzel et al. examined the clinical and pathological features of 31 HIV-negative HHV8-unrelated PEL-like lymphoma patients. The mean age of the patients was 66.9 years. Forty-two percent of the patients were HCV-positive, whereas 19.4 % of the patients were EBV-positive. Ninety percent of the cases had B-cell phenotype, whereas the remaining 10 % had T-cell phenotype. In this case series, there was only one case that was EBV-positive, HCV-negative, and had T-cell phenotype, and the patient was deceased 26 months after the diagnosis(14). The current case was EBV-positive, HCV-negative, HHV8-unrelated, PEL-like lymphoma with T-cell phenotype; however, the disease had a severely aggressive progression, and the patient was deceased the day after the diagnosis.

HHV8, HIV, and HCV were negative in the current patient, while EBV was positive. EBV positivity is usually seen in patients infected with HIV. EBV co-infection has been shown in 80% of the PEL patients who are infected with HIV (5). In the current case, HIV was negative while EBV was positive.

To our knowledge, our case is the first HHV8-unrelated PEL-like lymphoma case that was HIV-negative, EBV-positive, and had T-cell phenotype and aggressive progression. Atypical PEL type should be taken into consideration when diagnosing patients with PEL showing an aggressive progression ending with a rapid death.

References


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