PET/CT presentation of primary effusion lymphoma-like lymphoma unrelated to human herpes virus 8, a rare NHL subtype

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ABSTRACT

We present a 71-year-old female with human herpes virus 8 (HHV8)-unrelated primary effusion lymphoma (PEL)-like lymphoma. Dyspnea and pericardial effusion led to pericardiocentesis, diagnosing diffuse large B-cell lymphoma. She underwent positron emission tomography/computed tomography (PET/CT), which demonstrated hypermetabolic pericardial, pleural, and ascites fluid without lymphadenopathy elsewhere. Malignant fluid in the absence of lymphadenopathy is a hallmark of PEL. PEL is associated with immunodeficiency states such as acquired immunodeficiency syndrome (AIDS) and infectious agents such as HHV8. Our patient had no such history and had not received immunosuppressive chemotherapy. We present the PET/CT findings of this rare case of HHV8-unrelated PEL-like lymphoma.

Keywords: Human herpes virus 8, malignant pericardial effusion, positron emission tomography/computed tomography, primary effusion lymphoma

INTRODUCTION

Primary effusion lymphoma (PEL) is a rare form of non-Hodgkin’s lymphoma characterized by malignant fluid accumulation in the absence of lymphadenopathy. Typical sites of accumulation include pleural, pericardial, and ascites fluid. PEL is associated with immunodeficiency states such as acquired immunodeficiency syndrome (AIDS) and infectious etiologies such as human herpes virus 8 (HHV8). The World Health Organization (WHO) uses the PEL term for only HHV-related PEL. In our case, analysis of pericardial fluid was negative for HHV8. The term HHV8-unrelated PEL-like lymphoma has been used in the literature to describe such cases.

CASE REPORT

We present the case of a 71-year-old female with no significant past medical history who originally presented to the clinic with shortness of breath, abdominal bloating, and lower extremity edema. The symptoms appeared gradually over a few months. Subsequent transthoracic echocardiogram revealed a pericardial effusion. She then underwent pericardiocentesis with resultant marked improvement in her dyspnea. Post-pericardiocentesis the patient reported only residual orthopnea and night cough when lying down. Analysis of the aspirated pericardial fluid revealed monoclonal B-cell lymphocytes compatible with diffuse large B-cell lymphoma, which prompted staging positron emission tomography/computed tomography (PET/CT). Of note, the fluid was negative for HHV8. Immunologic blood work was negative for hepatitis A, hepatitis B, and hepatitis C infection. Serum polymerase chain reaction (PCR) was negative for cytomegalovirus (CMV) and Epstein–Barr virus (EBV).

The 18-fluorodeoxyglucose PET/CT (18-F FDG PET/CT) was performed using 19.6 mCi of intravenously administered radiotracer on a GE Discovery STE PET/CT scanner (GE Healthcare, Waukesha, WI, USA). The reconstruction diameter was 50 cm. The patient was imaged 1 and 2 h postinjection.

The PET/CT demonstrated uniformly increased radiotracer uptake in a moderate pericardial effusion (standardized uptake value (SUV) maximum 3.2), which increased in metabolic activity on delayed imaging (SUV maximum 3.6). Of note, the activity was within the fluid density space of the pericardial effusion.
and not limited to solely the pericardium itself [Figure 1]. Additionally, there were moderate bilateral pleural effusions and abdominal ascites, which demonstrated similar persistent and uniform radiotracer uptake [Figures 2 and 3] [Table 1]. These findings are compatible with malignant involvement, particularly given this patient’s history of prior pathology-proven malignant pericardial effusion.

Of note, the PET/CT also demonstrated no significant hypermetabolic cervical, thoracic, abdominal, or pelvic lymphadenopathy despite the above-described FDG-avid fluid collections [Figure 4].

The patient received chemotherapy and follow-up restaging PET/CT demonstrated interval resolution of the hypermetabolic pericardial, pleural, and peritoneal effusions [Figure 5].

**DISCUSSION**

PEL is a rare subtype of non-Hodgkin’s lymphoma characterized by malignant effusions in the body cavities such as the pericardial, pleural, and peritoneal cavities in the absence of lymphadenopathy or organomegaly. PEL was first described in 1989 in a patient with HHV8 and human immunodeficiency virus (HIV) infection. When PEL occurs in the absence of HHV8, the term HHV8-unrelated PEL-like lymphoma has been used in the literature.

The described PET/CT findings in our case of HHV8-unrelated PEL-like lymphoma are similar to the previously reported PET/CT findings in traditional PEL. Therefore, the distinction between PEL and HHV8-unrelated PEL-like lymphoma requires analysis of the fluid and testing for the presence of HHV8. The distinction between these two closely related clinical entities is important because HHV8-unrelated PEL-like lymphoma has a more favorable prognosis than traditional PEL.

The B-cell markers obtained from flow cytometric analysis have also been found to be helpful in the differentiation between PEL and HHV8-unrelated PEL-like lymphoma. The cells in PEL are usually negative for B-cell markers such as cluster of differentiation (CD)19, CD20, and CD79a. In our patient, flow cytometry was positive for CD20 and CD79a.

**CONCLUSION**

We describe the PET/CT findings of HHV8-unrelated PEL-like lymphoma, which has only once been previously reported. The presence of malignant fluid accumulation in the absence

![Figure 1](image1.png) Malignant pericardial effusion. Axial PET/CT images demonstrate 18-F FDG radiotracer uptake in moderate pericardial effusion (arrows). Analysis of pericardiocentesis fluid revealed diffuse large B-cell lymphoma. PET = Positron emission tomography, CT = Computed tomography, 18-F FDG = 18-fluorodeoxyglucose

![Figure 2](image2.png) Whole body MIP. Coronal whole body MIP image obtained 1 h after the intravenous injection of 19.6 mCi 18-F FDG. Note the absence of lymphadenopathy or hepatosplenomegaly. MIP = Maximum intensity projection, 18-F FDG = 18-fluorodeoxyglucose

![Figure 3](image3.png) (a and b) The 18F-FDG accumulation over time. Metabolically active bilateral pleural effusions (crosshairs) at (a) 1-h and (b) 2-h postinjection. 18-F FDG = 18-fluorodeoxyglucose

**Table 1: Standardized uptake value maximum over time**

|                | SUV<sub>max</sub> |
|----------------|------------------|
|                | 1 hour post injection | 2 hours post injection |
| Pericardial effusion | 3.2             | 3.6               |
| Pleural effusion   | 1.5             | 2.0               |
| Abdominal ascites  | 3.6             | 4.9               |
| Pelvic ascites     | 5.1             | 6.8               |

SUV: Standardized uptake value
of lymphadenopathy should raise suspicion of PEL. The imaging features of HHV8-unrelated PEL-like lymphoma appear to be similar to previously reported PEL PET/CT findings. Therefore, analysis of the fluid with particular attention to the presence of HHV8 is necessary to make this distinction, which is clinically significant because HHV8-unrelated PEL-like lymphoma portends a more favorable prognosis compared with traditional PEL.

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