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Case Report

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Primary Effusion Lymphoma

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Introduction

Primary effusion lymphoma (PEL) is a type of B-cell Non-Hodgkin's lymphoma (NHL) [1]. It was first described in 1989 as an AIDS-related lymphoma in patients with a human immunodeficiency virus (HIV) infection presenting with weakness and cachexia with a large malignant pleural effusion [2]. PEL was later found to be associated with kaposi sarcoma-associated herpesvirus (KSHV) or Human Herpes Virus 8 (HHV 8) [3,4]. PEL was later classified as a mature large B cell neoplasm without a detectable tumor mass by the World Health Organization [5].

The most common presenting complaints are fever, altered mental status, and dyspnea. Pleural effusion and ascites are seen in 85% and 50% of the patients, respectively [6]. The diagnosis can be challenging in patients with an unknown HIV history. Here we present a 41-year-old male with progressive dyspnea, anemia, thrombocytopenia, and large left-sided pleural effusion. During evaluation, he had a positive HIV test, and his pleural fluid analysis showed atypical lymphoid cells of B-cell origin, with prominent nucleoli and a large nucleus. Pleural staining showed a positive HHV 8 by polymerase chain reaction testing. Flow cytometry was positive for markers of lymphocytic activation CD30, CD38, CD45, CD71, CD138 Latency-associated Nuclear Antigen (LANA), Multiple Myeloma 1 (mum 1), paired box-5 (PAX-5), Human Leukocyte Antigen - DR isotype (HLA-DR).

Case presentation

A 41-year-old male with no significant past medical history presented to the emergency room with complaints of shortness of breath at rest. He reported the symptoms were ongoing for several weeks and were gradually progressive with cough and sputum. He denied any fevers or chills, night sweats, nausea, vomiting diarrhea. He denied any recent travel, drug use. He reported a history of unprotected sexual intercourse several months prior.

On presentation, he was febrile to 101 F, tachycardic to 130's/min, and tachypneic to 32 breaths/min, saturating at 99% on room air. On auscultation, he had absent left-sided breath sounds. His initial labs showed a hemoglobin of 9.1 (n=13.5-17.5 g/dL) and thrombocytopenia to 120 (n= 150-450 k/mm cu). His chest X-ray demonstrated opacification of the left hemithorax. A CT of the chest revealed left pleural effusion with severe compressive atelectasis of the left lung with marked nodular thickening of the pleura (Figure 1). His sexual history along with anemia and thrombocytopenia prompted an HIV test that was positive and his CD4 count was 186 cells/uL.

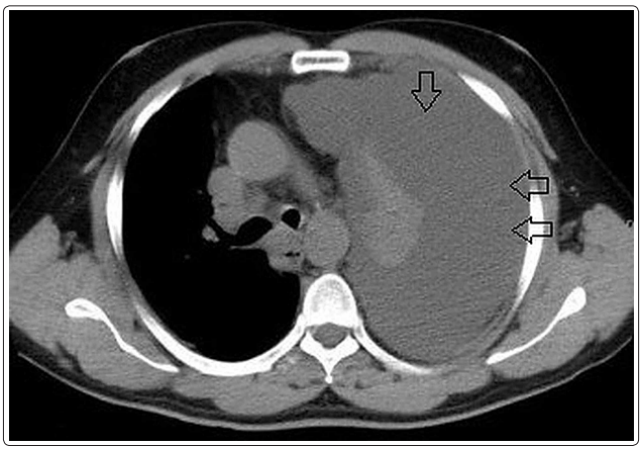

Figure 1: CT of the Chest showing large left sided pleural effusion (Black arrows)

A bedside thoracentesis was performed without any procedure-related complications, and 1500 mL of serosanguinous fluid was removed and sent for analysis. The cytopathology showed poorly differentiated atypical, large, single malignant cells with irregular nuclear membranes, open to marginated nuclear chromatin with prominent nucleoli. Immunohistochemical analysis was positive for CD30, CD38, CD45, CD71, CD138 Latency-associated Nuclear Antigen (LANA), Multiple Myeloma 1 (mum 1), paired box-5 (PAX-5), Human Leukocyte Antigen - DR isotype (HLA-DR).

The pleural fluid analysis also showed human herpesvirus 8 (HHV8), consistent with a diagnosis of primary effusion lymphoma (PEL). He was initially started on cefepime and azithromycin along with a chest tube was placement for pleural fluid removal. He was later started on chemotherapy with etoposide phosphate, prednisone, vincristine sulfate, cyclophosphamide, and doxorubicin hydrochloride (EPOCH) regimen and anti-retroviral therapy with bictegravir, emtricitabine & tenofovir alafenamide for HIV thereafter.

Discussion

Primary effusion lymphoma (PEL), also referred to as body cavity-based lymphoma, is a large B-cell Non-Hodgkin lymphoma localized predominantly in the pleura, pericardium, or peritoneum in immunocompromised patients. PEL corresponds to 1-8% of HIV-associated non-Hodgkin lymphoma cases and 0.5% of all lymphoma cases [6]. Characteristic features of the PEL are absence of an identifiable solid tumor mass and the presence of HHV-8. Approximately 80% of the PEL cases are coinfected with Epstein-Barr virus (EBV) [7]. The most common presenting symptoms include dyspnea, abdominal distention, and joint swelling due to mass effect from the accumulated effusions. Dissemination to distant sites is common, and the survival without treatment is no more than several months after initial diagnosis [8]. The diagnosis of PEL is made on cytological preparations such as liquid-based preparation, cytosporin, cell block of the involved effusion fluid [9]. The neoplastic cells are large, with prominent nucleoli, a round to irregular nuclei, and a varying amount of basophilic cytoplasm, with occasional vacuolated cells [9]. PEL cells express a "null" lymphocyte phenotype as they are indeterminate by immunohistochemistry and do not exhibit typical B-cell or T-cell immunophenotype characteristics.

The most common markers found are CD 30, CD 38, CD 138, CD 71, LANA, mum1, PAX5. There is also a variable expression of HLA-DR, epithelial membrane antigen, and activation antigen [1]. The effusion is also positive for HHV-8 with the expression of LANA-1 by an immunohistochemical stain or by a polymerase chain reaction. It is postulated that the virus encodes various genes involved in inducing cell proliferation and inhibiting apoptosis [1]. PEL has a low incidence and poor clinical prognosis, with a 1-year survival rate of 39%. Patients not on antiretroviral therapy at the time of diagnosis had worse outcomes [6]. Patients on highly active antiretroviral therapy have better prognosis, with a few patients achieving prolonged remission with just antiretroviral therapy alone [8,10].

The accurate diagnosis and of PEL depends on the cytologic evaluation of the specimen. The diagnosis can be challenging in a patient with an unknown HIV history and when the primary site of involvement is the body cavity [11]. Chemotherapy has traditionally been the cornerstone of treatment with aggressive lymphoma regimens such as Etoposide, Prednisolone, Vincristine, cyclophosphamide, doxorubicin (EPOCH) regimen, or cyclophosphamide, doxorubicin, vincristine, prednisone (CHOP) regimen [1]. Development of targeted therapies using Nuclear factor-kappa B (NF-kB), Janus kinase (JAK)- signal transducer and activator of transcription (STAT), and phosphatidylinositol 3-kinase pathways are still ongoing, that will provide further insight into management of PEL [2].

References

1. Narkhede M, Arora S, Ujjani C (2018) Primary effusion lymphoma: current perspectives. Onco Targets Ther 11: 3747-3754.
2. Shimada K, Hayakawa F, Kiyoi H (2018) Biology and management of primary effusion lymphoma. Blood 132:1879-1888.
3. Cesarman E, Chang Y, Moore P S, Said J W, Knowles D M (1995) Kaposi's sarcoma-associated herpesvirus-like DNA sequences in AIDS-related body-cavity-based lymphomas. N Engl J Med 332: 1186-1191.
4. Nador R G, Cesarman E, Chadburn A, Dawson D B, Ansari M Q, et al. (1996) Primary effusion lymphoma: a distinct clinicopathologic entity associated with the Kaposi's sarcoma-associated herpes virus. Blood 88: 645-656.
5. Norris FRCPA D (2012) WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues (4th Edition) - Lymphoma Classification in the Third Millennium. QML Pathol Newsl 2012: 1-12.
6. Borie R, Wislez M, Antoine M, Cadranel J (2017) Lymphoproliferative Disorders of the Lung. Respiration 94: 157-175.
7. Lurain K, Polizzotto M N, Aleman K, Manisha Bhutani, Kathleen M Wyvill, et al. (2019) Viral, immunologic, and clinical features of primary effusion lymphoma. Blood 133: 1753-1761.
8. Chen Y-B, Rahemtullah A, Hochberg E (2007) Primary effusion lymphoma. Oncologist 12: 569-576.
9. Patel S, Xiao P (2013) Primary effusion lymphoma. Arch Pathol Lab Med 137: 1152-1154.
10. Hocqueloux L, Agbalika F, Oksenhendler E, Molina J-M (2001) Long-term remission of an AIDS-related primary effusion lymphoma with antiviral therapy. AIDS 15: 280-282.
11. Xiao J, Selvaggi S M, Leith C P, Fitzgerald S A, Stewart J (2013) Kaposi sarcoma herpesvirus=human herpesvirus-8-negative effusion-based lymphoma: Report of 3 cases and review of the literature. Cancer Cytopathol 121: 661-669.