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Case Report

A Rare Presentation of Plasmablastic Lymphoma in a HIV-negative Male Status-post Liver Transplantation

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Abstract

Introduction: Plasmablastic lymphoma, a rare and aggressive form of Non-Hodgkins lymphoma, rarely presents as a retroperitoneal mass. There are no case reports in the literature of plasmablastic lymphoma presenting in living, HIV-negative patient status post liver transplantation.

Presentation of Case: We report the case of a 63 year-old HIV-negative male status post liver transplant who presented with weakness and obstructive uropathy. Imaging showed a large mass in the retroperitonium. Biopsy revealed plasmablastic lymphoma. CHOP therapy was initiated and after six cycles, the retroperitoneal mass regressed in size; however, cytology from pleural fluid revealed that the disease remained. It has been 9 months since initial diagnosis and he was started on salvage chemotherapy with ESHAP however he subsequently developed a treatment related myelodysplastic syndrome with trisomy 12.

Conclusions: The present case is used to explore the presentation and treatment of plasmablastic lymphoma, and to review the literature concerning the rarity of this disease in the setting of a HIV-negative patient status post solid organ transplantation.

Keywords: Plasmablastic lymphoma; retroperitoneal mass; transplant; HIV-negative

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Consent: We confirm that the patient has given his informed consent for the case report to be published.

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Introduction

Plasmablastic lymphoma (PBL) is an aggressive and rare form of Non-Hodgkin's Diffuse B cell lymphoma [1-7, 9-12, 15]. This disease is usually considered an acquired immunodeficiency syndrome (AIDS)-related lymphoma but it has also been seen in Human Immunodeficiency Virus (HIV)-negative patients [1-6, 9, 10]. PBL has a strong association with the Epstein Barr Virus [6-8]. It has a predilection for the oral cavity however extra-oral sites including central nervous system, lung, liver, small bowel, testis and lymph nodes have been reported [1, 6, 9-11].

There have been two reported cases of plasmablastic lymphoma presenting as a retroperitoneal mass [10, 11]. The first case involved a HIV-positive American male and the second case described a HIV-negative male from Japan whose diagnosis was made at autopsy. In this case report we describe the first known report of plasmablastic lymphoma in a living, American, HIV-negative male, status-post solid organ transplantation who presented with weakness and obstructive uropathy secondary to a retroperitoneal mass.

Case presentation

A 63-year-old male presented to the emergency department with generalized weakness, urinary retention and diarrhea. His past medical history is notable for end stage liver disease secondary to hepatitis C status post orthotopic liver transplant approximately one year ago treated with Tacrolimus, Ribavirin, and Peginterferon alpha 2a. Work up prior to transplantation revealed previous exposure to the Epstein Barr Virus. He denied any current tobacco, alcohol or recreational drug use however, he did admit to drinking heavily from age 20-27 drinking a 6 pack per day.

On physical exam vital signs were stable and physical exam was unremarkable except for dry mucous membranes, poor skin turgor and an enlarged prostate.

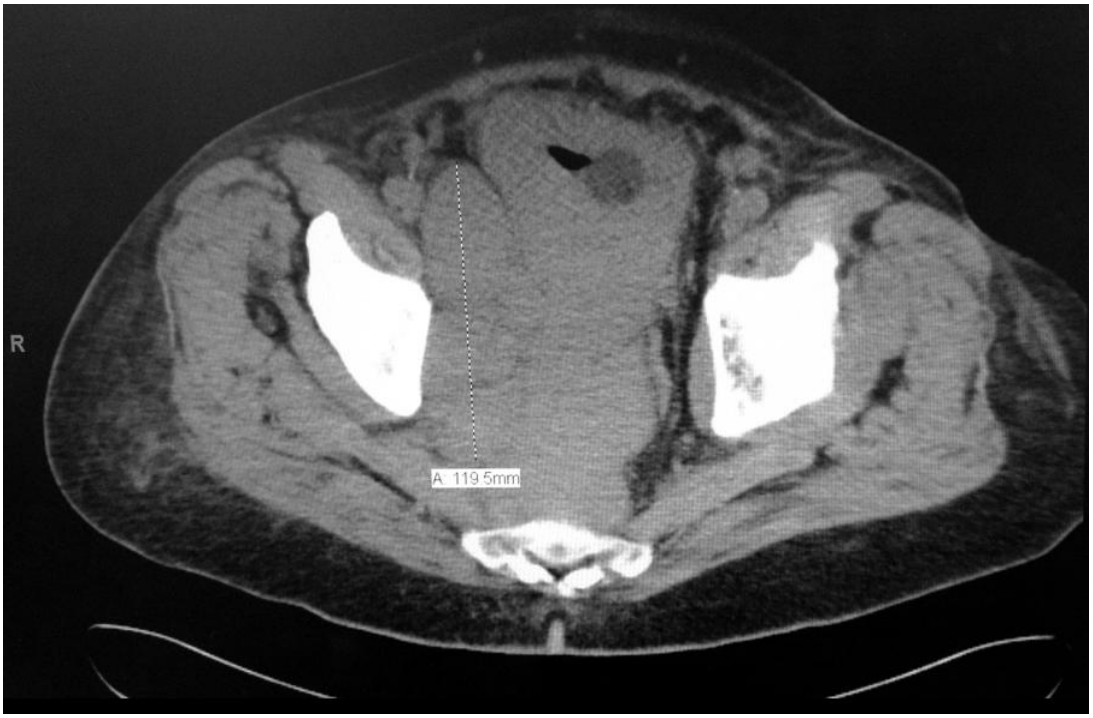

Figure 1 CT Abdomen/Pelvis illustrating a 11.95 cm retroperitoneal mass in the right external iliac region.

Laboratory findings revealed pancytopenia, hyponatremia, and BUN of 48 mg/dL and creatinine of 4.8 mg/dL. A foley catheter was placed and drained 1500 ml of bloody urine. Intravenous fluid hydration was started immediately. He was admitted for evaluation of acute kidney injury and hematologic abnormalities. A bladder ultrasound revealed a distended bladder, bladder wall thickening and an enlarged and heterogenous prostate compatible with prostate hypertrophy. Computer Tomography (CT) of the abdomen and pelvis showed multiple large retroperitoneal masses, largest mass (11.95 cm in diameter) was in the right external iliac region (Figure 1). Histopathology of the pelvic mass revealed plasmablastic B cell lymphoma that was CD138 strong +, CD3-, CD5-, CD10-, CD20-, CD23- CD30- CD79a+, CD117 weak +, Cyclin-D1-, Multiple Myeloma Oncogene-1+, Kappa-, Lambda+, Keratin-, Epithelial Membrane Antigen+, Placental Alkaline Phosphatase-, Human Herpes Virus 8- and Epstein Barr Virus+ (Figure 2). Immunohistochemical stains were negative for activin receptor-like kinase 1 (ALK1). A bone marrow biopsy showed no evidence of lymphoma involvement and a markedly hypocellular marrow.

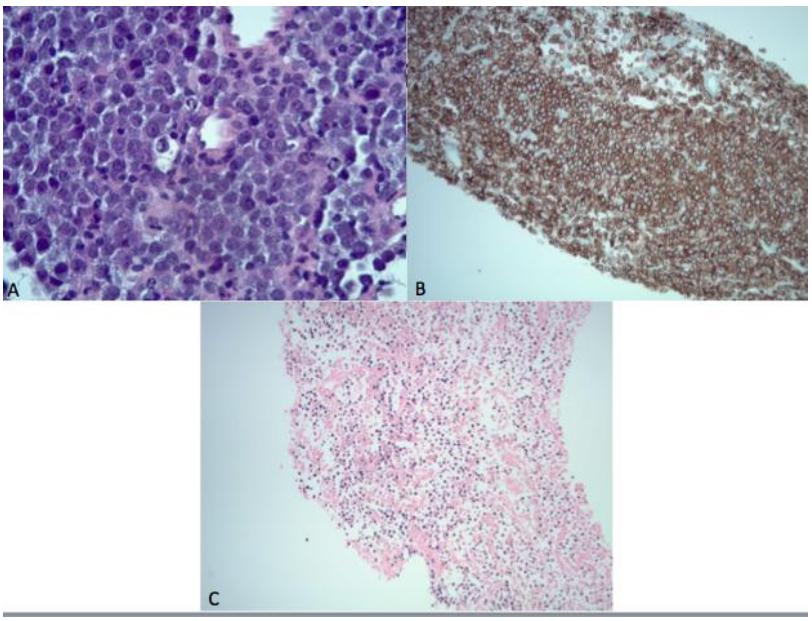

Figure 2 A: Hematoxylin and Eosin, 40x. Tumor is comprised of large cells with scant cytoplasm, nuclei with dispersed (fine) reticular chromatin with minimal to no chromatin clumping and 1 or more large nucleoli
B: CD138, 20x. Tumor cells are diffusely immunoreactive for CD138 by immunohistochemistry.
C: EBER, 10x. Tumor cells express Epstein Barr Virus encoded RNA (EBER) by in-situ hybridization.

An infusaport was placed and a mutli gated acquisition scan (MUGA scan) was done prior to starting treatment. The patient was started on Cyclophosphamide, Doxorubicin, Vincristine and Prednisone (CHOP). After the second cycle of chemotherapy, a staging Positron Emission Tomography- Computed Tomography (PET/CT) showed no evidence for retroperitoneal uptake and the pelvic sidewall mass was approximately 6.5 cm. Standardized Uptake Value (SUV) was only mildly elevated up to 2.6 (Figure 3).

Approximately 6 months later after completion of six cycles of CHOP therapy, the patient received a restaging PET/CT which revealed a significant decrease in size and SUV activity in the right pelvic sidewall mass and no evidence for new lesions. A new large, left pleural effusion was discovered (Figure 3). Cytology from the pleural fluid revealed plasmablastic lymphoma with 71% of all nucleated element being cytoplasmic lambda monoclonal plasma cells. The patient was started on salvage chemotherapy with ESHAP (Etopiside, methylprednisolone, cytarabine and cisplatin).

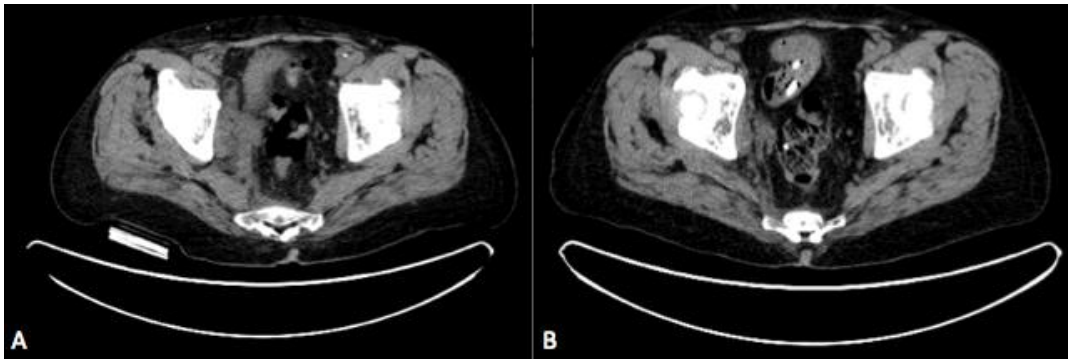

Figure 3 A: Initial PET/CT showing the pelvic sidewall mass measuring approximately 6.5 cm
B: Restaging PET/CT showing a significant decrease in size and SUV activity in the right pelvic sidewall mass and no evidence for new lesions

Discussion

Plasmablastic Lymphoma is a rare aggressive variant of diffuse large B cell Non-Hodgkin's lymphoma. Tumor cells are comprised of terminal B cells that have well differentiated plasma markers such as CD138 and minimal or absent expression of B cell markers CD45, CD20, and CD79a [1-5]. EBV infection has been observed in 74% of published PBL cases [1]. Epstein Barr Virus Encoded RNA In-Situ Hybridization (EBER-ISH) has a positive predictive value of close to 100% in HIV-positive PBL patients. Therefore, it has been suggested to use EBER-ISH to aid in the diagnosis of PBL [6-8]. According to Castillo et al., Patients who had HIV-negative PBL have lower rates of Epstein-Barr virus-encoded RNA expression than HIV-positive patients [6].

Plasmablastic lymphoma accounts for 2.6% of all AIDS related lymphoma [1,7,10]. It has also been described in approximately 76 cases of HIV-negative patients, both immunocompromised from solid organ transplantation as well as in immunocompetent individuals [6]. Delecluse et al. reported PBL in HIV-positive patients is most often found in the oral cavity and was seen in 58% of cases [7]. In HIV-negative patients, oral cavity involvement is lower at 21% of the cases and is more likely to present with extra-oral sites. PBL has been found in the CNS, liver, lungs, testis, subcutaneous tissue of the arm, lymph nodes and retroperitoneum [9-12]. Sixteen cases have been described in literature associated with immunosuppression from anti rejection therapy after solid organ transplant (kidney, heart, lung, and pancreas) [14].

A literature review was performed using Pubmed confirmed the rarity of plasmablastic lymphoma presenting as a retroperitoneal mass. Only two cases have been reported. Dholaria et al. described a case of PBL of the retroperitoneum in an HIV-positive patient and Takahashi et al. described a case of a HIV-negative male from Japan who was diagnosed at time of autopsy with plasmablastic lymphoma [10,11]. There has been no reported cases of plasmblastic lymphoma presenting as a retroperitoneal mass in a living, American, HIV negative patient. Furthermore, there are no reported cases of PBL status post liver transplantation in adults documented in literature.

PBL is associated with early dissemination, poor response to therapy and limited survival [1-6, 9,11]. In HIV negative patients, overall survival is approximately 9 months [6,15]. The National Comprehensive Cancer Network (NCCN), recommends Hyper-CVAD (Cyclophosphomide, vincristine, doxorubicin, and dexamethasone alternating with high dose methotrexate and cytrabine), dose adjusted –EPOCH (etopiside, prednisone, vincristine, cyclophosphamide, doxorubicin) or CODOX-M/IVAC (cyclophosphamide, vincristine, doxorubicin, high dose methotrexate, alternating with ifosfamide, etoposide, and high dose cytrabine) [15]. While NCCN guidelines state that CHOP therapy is not adequate, Castillo et al. reports more intensive

regimens have not shown a survival advantage when compared to CHOP therapy [13,15]. Our treatment plan is similar to one described by Jae Myung Cha et al. in an article titled, A case report with Plasmablastic Lymphoma of the jejunum, in which the patient was treated with six cycles of CHOP and salvage chemotherapy with ESHAP. The patient had a survival of greater than 24 months [12]. In our case, the patient had a near complete response to CHOP chemotherapy with a rapid relapse in an extra-lymphatic site. He subsequently started on ESHAP but after one cycle developed refractory thrombocytopenia and was diagnosed with treatment related myelodysplasic syndrome with a trisomy 12. ESHAP has currently been stopped. More research is needed to establish better treatment guidelines to increase survival.

This is the first case of plasmablastic lymphoma presenting as a retroperitoneal mass in a living, HIV-negative patient and is among one of the extremely rare cases of plasmablastic lymphoma diagnosed after solid organ transplantation. This case is the first described in literature status-post liver transplant in an adult.

Conclusion

In conclusion, the findings of this case suggest that plasmablastic lymphoma should be included in the differential diagnosis of patients who are status-post transplantation presenting with a retroperitoneal mass. The course of this patient's disease and care will be a useful addition to the current literature for determining the treatment and prognosis in HIV-negative patients presenting with PBL.

Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Acknowledgements

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