Plasmablastic lymphoma presenting as painful subcutaneous nodules in a HIV positive patient: A case report

Deea Das, DO, Emilee Welton, DO, Jessica Tyler, DO

Kettering Health Dayton Department of Internal Medicine

Plasmablastic lymphoma (PBL) is a rare subtype of diffuse B-cell lymphoma that typically presents in HIV positive patients. It has been estimated that the incidence of PBL is approximately 2% of all HIV-related lymphomas.This report details a HIV positive patient who was diagnosed with the disease after presenting with an uncommon extranodal manifestation of the disease, painful skin nodules. Skin manifestation of PBL is only seen in about 6% of HIV positive PBL cases. Additionally, this patient was found to have the disease in multiple unusual sites, such as the liver, skeleton, and pleural surface of the lungs. In addition to highlighting less common areas of manifestation of plasmablastic lymphoma, this report will discuss the current prognosis and treatments of this disease.

Case Presentation

A 43-year-old female presented to the emergency department due to numerous tender, subcutaneous nodules on her lower back, abdomen and left leg. She had a known history of HIV, with last known CD4 count of 156. These nodules appeared 1 week prior to her presentation, and no trigger was identified. Infectious Disease was consulted and a CT chest without contrast showed numerous soft tissue masses throughout the chest wall, including both breasts, the pleura, lungs and the pancreatic body. Infectious and autoimmune workup was negative and a core needle biopsy of one of the lesions was done. The patient was then discharged from the hospital with pain medication.

A few days later, the patient returned to the hospital due to enlarging nodules, new nodules appearing , and intractable pain. Her initial biopsy resulted, demonstrating aggressive lymphoid neoplasm with plasmacytic differentiation, favoring PBL. PBL was then later confirmed when FISH studies showed MYC-IgH fusion t(8;14). The patient was admitted to the hospital and Hematology/Oncology was consulted. PET/CT showed extensive malignancy with dominant lesions throughout the trunk and extremities. Malignant lesions were also seen along the pleural surface of the lungs, mediastinum and left hilum, mesentery, retroperitoneal and pelvic lymph nodes, spleen, liver and skeleton. Due to the patient's presentation, it was decided to urgently start the patient on a chemotherapy regime. She was given intrathecal methotrexate to prevent spread of the disease to the central nervous system. IV fluids and allopurinol was started because she had a high risk of developing tumor lysis syndrome due to extensive tumor burden. The patient was then started on a chemotherapy regime of etoposide, vincristine and doxorubicin with bolus cyclophosphamide and prednisone (EPOCH) plus daratumumab.

References

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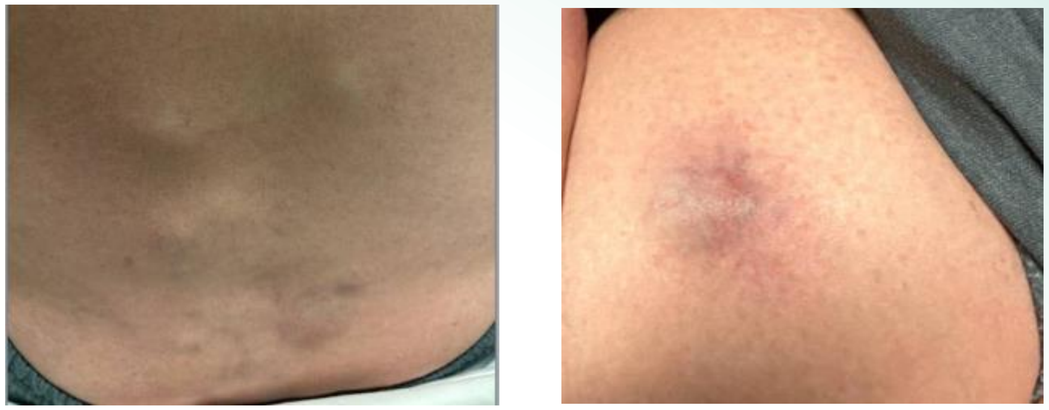
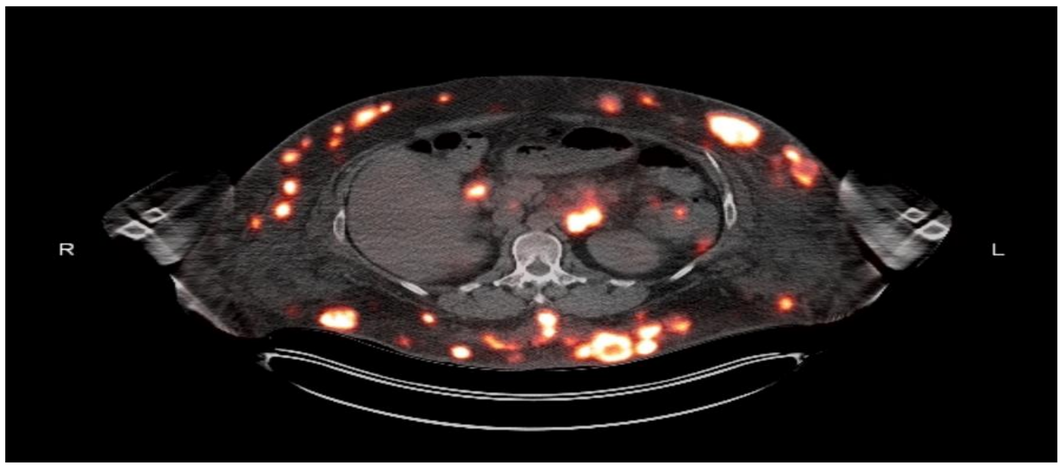
Imaging



Pictured above are the initial nodules the patient presented with.



PET/CT demonstrating multiple lesions.

Discussion and Conclusion

Only 6% of HIV positive patients present with skin lesions in PBL[1] 1. Additionally, this patient presented with multiple uncommon sites of PBL involvement. The most common areas of involvement in HIV patients are the oral cavity/jaw at 48% and gastrointestinal system at 12%. Only 5% of cases involve the bones, 2 % the liver, 2% the lungs, and 0.2% the breast[1].

Chemotherapy is the first line therapy to treat PBL. It is currently recommended to use more intense chemotherapy regimens like as CODOX-M/IVAC (cyclophosphamide, vincristine, doxorubicin, and high-dose methotrexate alternated with ifosfamide, etoposide, and high-dose cytarabine), dose-modified EPOCH (etoposide, prednisone, vincristine, cyclocarbonamide, and doxorubicin), or Hyper-CVAD (Cyclophosphamide, vincristine, doxorubicin, and dexamethasone) alternated with high-dose methotrexate and cytarabine [1,2,4]. However, some studies have demonstrated that this does not provide any survival benefit when compared to less intensive chemotherapy regimens[1,4].

In this patient daratumumab was used in addition to her primary chemotherapy regimen. PBL often expresses CD 38, and daratumumab can induce NK cells to produce antigen-dependent cell-mediated cytotoxicity attacking this antigen[2].

Bortezomib (A proteasome inhibitor) may also improve outcomes, but most data demonstrating this is based on retrospective studies. Other treatments, like immune checkpoint inhibitor nivolumb, are currently being investigated as they have shown promise in case reports[2].

The prognosis of PBL is poor despite current treatments. There have been cases of complete resolution of PBL, but these remain the exception[3]. Currently, the median survival time of patients with PBL is between 6-19 months[1,2,4] . Further treatments need to be investigated to improve survival in patients with PBL, but this remains difficult to do given the rarity of the disease.

The patient initially showed improvement after starting chemotherapy. There was a large reduction of tumors on repeat imaging. However, unfortunately her most recent PET/CT showed progressive disease. She continues to receive treatment at this times for her PBL and HIV.