Plasmablastic Lymphoma (PBL): A Diagnostic and Therapeutic Challenge

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INTRODUCTION

Plasmablastic lymphoma (PBL), a distinct subtype of diffuse large B-cell lymphoma, accounts for almost 2.6% of HIV-related non-Hodgkin lymphomas. It needs to be distinguished from multiple myeloma (MM) and other B-cell lymphomas as the treatment and prognosis are entirely different.

The mean age at presentation is 39 years with a male: female ratio of 7:1. Plasmablastic lymphoma presents on average 6 years after the initial diagnosis of HIV and the CD4 counts are usually <200 cells/cu.mm. Commonly involved sites include oral cavity, Epstein Barr virus is detected in 75% of cases and human herpesvirus-8 in 16% of cases. Initial response to therapy may be good, but long-term prognosis remains poor.

CASE DESCRIPTION

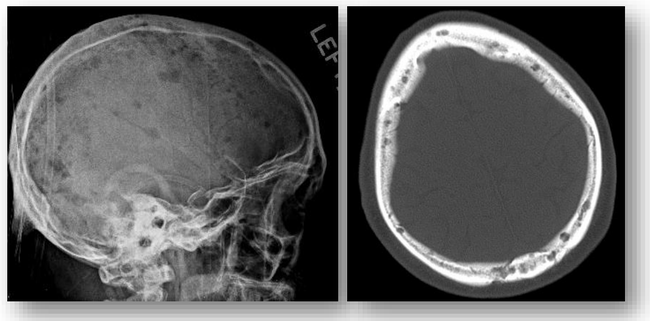
HISTORY:

- A 38 y/o Hispanic male, diagnosed with AIDS two months prior to presentation, was admitted for altered mental status. He lost 40 lbs but denied fevers or night sweats . Patient was also complaining of difficulty and pain on swallowing.

PHYSICAL EXAM:

- Examination revealed a cachectic and lethargic male, with mild splenomegaly but no palpable lymphadenopathy.
- Laboratory: Corrected calcium markedly elevated at 19.5 mg/dL, mild anemia and renal insufficiency; elevated total protein to 9.1 G/L. LDH elevated to 960 u/L.

IMAGING

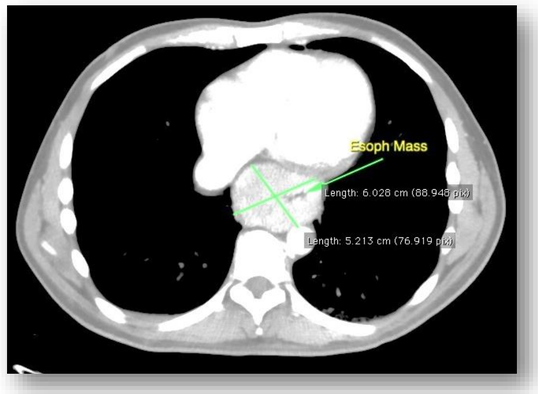


Images of skull showing multiple lytic lesions involving the whole skull. No intra cranial abnormality identified in MRI, CT brain.

HOSPITAL COURSE:

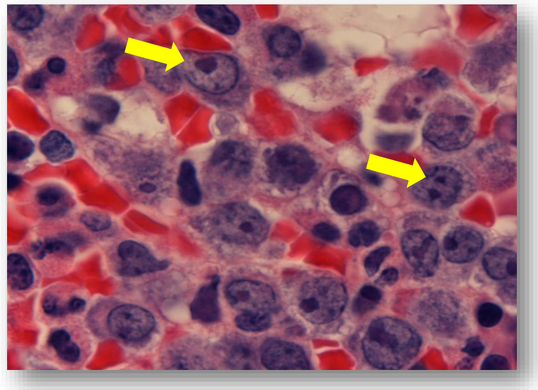
- IgM-kappa monoclonal protein by IFE, hypercalcemia and calvarial lytic lesions on CT scan raised concerns for MM and lymphoplasmacytic lymphoma.
- IgM levels increased to 2647 mg/dL but erum viscosity was not elevated at 2.5 cp.
- CD4 count decreased to <40 cell/mm^3, HIV viral load increased to 44,600 copies/mL
- EGD revealed CMV esophagitis; CMV PCR 1544 copies/mL.
- Bone marrow biopsy demonstrated monomorphic proliferation of large cells positive for CD 138 with kappa restriction but negative for CD20 corresponding to a plasma cell immunophenotype.
- In situ hybridization was positive for EBV encoded RNA (EBER) consistent with a diagnosis of PBL.
- Treatment for PBL held while Induction ganciclovir initiated for CMV disease
- Two weeks into ganciclovir therapy, esophageal mass developed. EGD biopsy confirmed PBL.
- Chemotherapy with DA-EPOCH started immediately.

CT CHEST



Development of new 6 x 5 cm distal esophageal mass in 3 weeks.

PATHOLOGY



- PBL typically larger than a mature RBCs with round eccentrically placed nucleus with less mature chromatin. Moderate amount of basophilic (blue) cytoplasm with prominent perinuclear hof (clearing) Nucleoli are Prominent.

DISCUSSION

- AIDS-related (PBL) rare, highly aggressive cancer that can present both a diagnostic and therapeutic dilemma.
- Expression of EBER differentiates it from plasmablastic MM.
- PBL has predilection to the oral cavity.
- Extra-oral presentation frequently occurs in HIV patients; our patient with esophageal and bone marrow involvement. HAART improves survival but rarely leads to spontaneous remission.
- PBL frequently relapses after chemotherapy, and is often rapidly fatal.
- Active CMV infection prohibited prompt chemotherapy.

A CLINICIAN'S PERSPECTIVE

- Awareness of plasmablastic cancers arising in advanced AIDS patients.
- Diagnostic challenge as it mimics other cancers such as MM, WM and other lymphomas
- Therapeutically challenging due to poor prognosis despite aggressive chemotherapy and accompanying opportunistic infections.

REFERENCES

REFERENCES: 1.. 2., 3. 4. Voice RA, Bradley SF, Sangeorzan JA, Kauffman CA. Clin Infect Dis.