Clinicopathological correlation in cases of Plasmablastic lymphoma

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BACKGROUND

Plasmablastic Lymphoma (PBL)

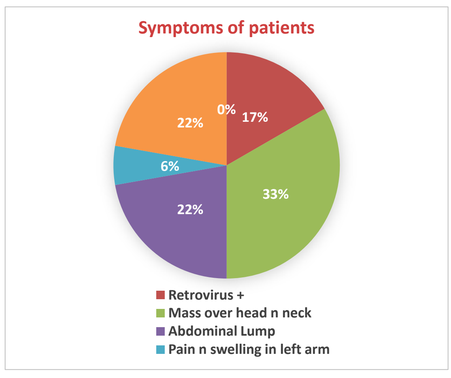
- Is an aggressive B- cell lymphoma
- Has post germinal center B cell phenotype and immunoblastic morphology
- Typically presents with extranodal involvement and could affect any organ
- The rate of extranodal disease is higher with PBL (53%) than with DLBCL (35%)
  - though the proportion of patients with advanced clinical stage was similar at 57% and 55%, respectively.
- Commonly affected extranodal sites:
  - Oral cavity, gastrointestinal tract, lymph nodes, paranasal sinuses, and skin
  - Less commonly affected sites:
    - skeletal bone, genitourinary tract, central nervous system, liver, and lung.
- Usually associated with immunocompromised settings
  - HIV or EBV infections
  - Post-transplant status
- Also reported in immunocompetent patients
- Morphologically: on microscopy we find
  - Medium to small sized cells
  - Moderate amount of cytoplasm
  - Round to oval nucleus
  - Prominent nucleoli
  - plasmablasts like differentiation admixed with mature plasma cells
  - They usually have diffuse infiltration.
- Immunophenotyping:
  - Cells are negative for B-cell markers
    - CD19, CD20 & CD22
  - Positive for plasma cell markers
    - CD138, CD38 & MUM1

MATERIAL AND METHODS

- A retrospective study of 11 cases of plasmablastic lymphoma
- Between the years 2008 and 2020
- Relevant demographic, clinical, pathological information was collected and analyzed

RESULTS

- Out of the 11 patients,
  - 7(63.6%) were male patients
  - 4(36.3%) were female
- Median age 40 years (IQR 20-48)
- Laboratory investigations at presentation (median values) were
  - Hemoglobin 12.1 mg/dl (IQR 1-12.7)
  - TLC 7750 cells/micro litre (IQR 5900-8500) and
  - Platelets count 270000 cells/micro litre (IQR 94000-430000)
- Symptoms
  - 3/11 (27.2%) patients were retrovirus positive, with one of them was also a chronic hepatitis B carrier.
  - 6/11 (54.5%) patients presented with mass over head and neck region.
  - 4/11 (36.3%) patients presented with abdominal lump.
  - 1/11 (%) patient presented with pain and swelling in left arm.
  - 4/11 (36.3%) patients had B symptoms.
  - * some patients presenting with 2 or more symptoms*



- On biochemical investigations median values were as follows:
  - Serum albumin 4 g/dl (IQR 3.3-4.1)
  - S. Creatinine 0.7 mg/dl (IQR 0.5-0.9)
  - S. calcium 9.2 mg/dl (8.5 - 10)
  - S. LDH 355 IU/L (IQR 271-566)
  - Only the LDH values were slightly elevated
- Bone marrow (BM) aspirate examination:
  - 4/11 (36.3%) had infiltration of bone marrow by plasma cells
  - All of them presented with swelling over head and neck
- BM biopsy:
  - Infiltration by large atypical lymphoid cells with plasmacytoid morphology was seen in all the cases
  - All samples were
    - Positive for CD138, MUM1
    - Negative for CD20 (except 1 sample)

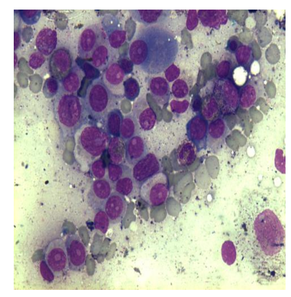


Figure 2: BMA showing sheet of plasmablasts, MGG stain, 100X OIL

Treatment:

- Dose adjusted EPOCH regimen,
  - 3/11 (27.2%) patients
- BFM 90 protocol
  - 2/11(18.1%) patients
- R- CHOP protocol
  - 2/11(18.1%) patients
- VPD and Bortezomib
  - 4/11 (36.3%) patients

- 1 patient was lost to follow up before starting therapy
- 2 patients had a relapse and they were given ICE regimen
- Only 1 patient had died in the duration of follow up
- The median follow up duration being 5 months (IQR 3-12)

DISCUSSION

- PBL is a rare, aggressive lymphoma
- First described in HIV-positive patients
- But increasingly seen in immunocompetent individuals (1,2)

In this study:

- most patients were immunocompetent (8/11 cases)
  - with the head/neck region as the predominant site (6/11 cases)
  - often linked to bone marrow infiltration
- Unlike other aggressive lymphomas,
  - generalized lymphadenopathy was absent,
  - supporting prior reports of extranodal predominance (Taddesse-Heath et al., 2010)[4].
- Immunophenotype (CD138+, MUM1+, CD20-) was consistent with classical PBL morphology (Colomo et al., 2004) [3]
- Our study shows Elevated LDH reinforced its aggressive biology.
- No standard treatment regimen has been established yet
  - While CHOP regimens are generally inadequate, DA-EPOCH is favored in HIV-positive cases
  - Combinations with Bortezomib show promise(Castillo et al.,2012; Morscio et al.,201) [2].
- Despite short follow-up, our data align with literature noting frequent relapse and poor outcomes (median OS: 6-19 months).

TAKE-HOME MESSAGES

- PBL is not restricted to immunodeficient patients
- Head/neck involvement is common; bone marrow disease may co-occur
- Presence of B symptoms is common
- Lymphadenopathy being rare
- Regimens that can be used for this highly aggressive lymphoma include
  - CHOP, DA-EPOCH, hyper-CVAD
- DA-EPOCH is considered to be superior to CHOP regimen in HIV positive patients
  - Is now considered as first line therapy
- The prognosis of patients with PBL is dismal
  - Median overall survival (OS) of patients being 6-19 months
  - Highlights the need for clinical trials and molecular studies

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