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EBV+ B-cell polymorphic lymphoproliferative disorder of the lip in a patient with advanced chronic lymphocytic leukemia

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History

- A 77 year-old white male with a history of advanced chronic lymphocytic leukemia (CLL) on rituximab and bendamustine presented with a 6-week history of a growing, tender nodule on his right lower lip.
- He had previously failed an empiric trial of acyclovir for suspected herpes labialis.

Examination

- Involving the right lower lip, there was an ~ 3 x 2 cm nodule with central hemorrhagic crust.



Figure 1. ~ 3 x 2 cm nodule with central ulcer

Differential and Clinical Course

- Differential diagnosis included EBV+ B-cell polymorphic lymphoproliferative disorder (B-PMLD), EBV+ mucocutaneous ulcer (MCU), EBV+ marginal zone lymphoma (MZL), and cutaneous CLL.
- Unfortunately, the patient expired a few weeks after presentation from complications of his CLL.

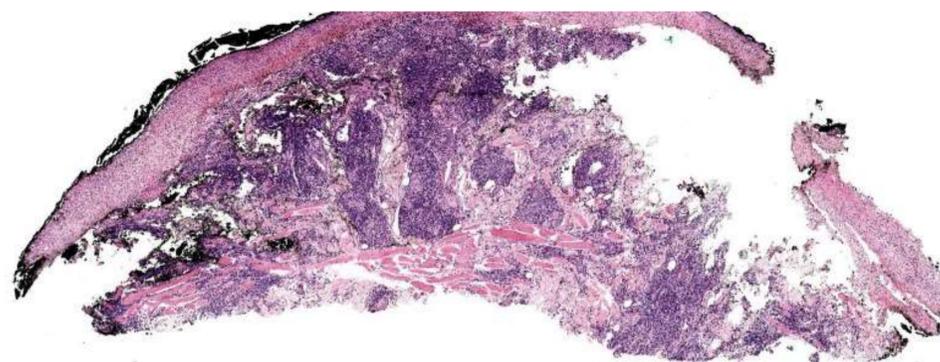


Figure 2. H&E, Original Magnification, 50x

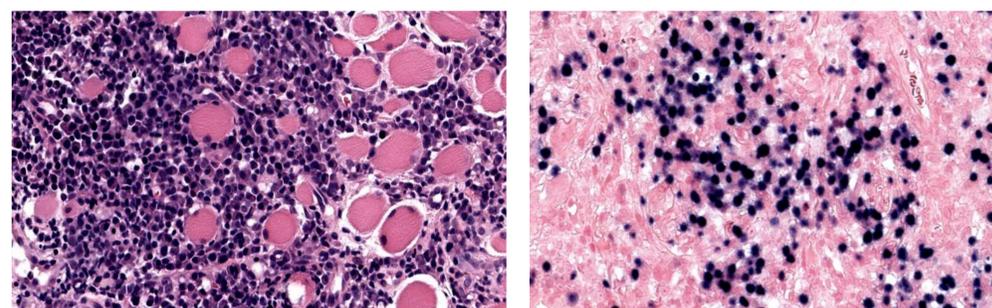


Figure 3. The polymorphous infiltrate is seen dissecting through skeletal muscle fibers (H&E, Original Magnification, 200x)

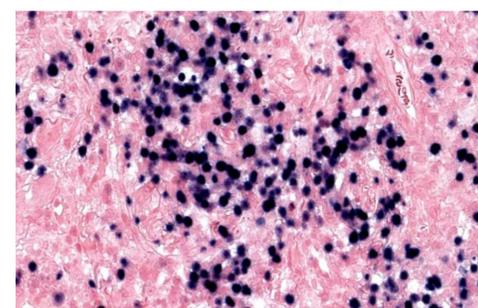


Figure 4. EBV positivity (EBER ISH, Original Magnification, 200x)

Discussion

- EBV+ B-PMLD is a rare lymphoid neoplasm that typically occurs in post-transplant and other immunodeficiency settings. Lesions are morphologically heterogenous and can result in destructive masses that may mimic large B-cell or classical Hodgkin lymphomas.^{1,2,3}
- Histologically, B-PLMDs exhibit a full range of B-cell maturation stages including small- to medium-sized lymphocytes, plasma cells, and immunoblasts. Clonality is usually demonstrable via IGH gene rearrangement studies and/or evaluation of lambda & kappa for light chain restriction.^{1,2}
- First line treatment is reduction of immunosuppression, but more aggressive immunotherapy, chemotherapy, or radiation therapy may be required for cases that do not regress or when reversal of immunosuppression is not feasible. Fatal dissemination of disease may still occur despite intervention.^{1,2}
- EBV+ B-PMLD falls on a spectrum of B-cell lymphoproliferative disorders that are often challenging to definitively distinguish given overlapping morphologic and immunophenotypic findings.
- EBV+ MCU represents a localized form of immunodeficiency that most commonly presents as a sharply demarcated ulcer on the oropharyngeal mucosa. Unlike EBV+ B-PMLD, EBV+ MCU typically affects immunocompetent individuals, has more conspicuous CD30+ immunoblasts than plasmacytoid cells, and often lacks significant nodularity.^{1,2}
- Cutaneous EBV+ MZL is a rarely reported immunodeficiency-associated lymphoma.⁴ While entity cannot be entirely excluded in this case, cutaneous lesions of EBV+ MZL would be expected to show some evidence of pre-existing lymphoid follicles.⁴
- Cutaneous CLL was not favored given the distinctly plasmacytoid differentiation of the tumor. Furthermore, the patient's CLL had demonstrated kappa light chain restriction as opposed to the lambda restriction evident in this lesion.⁵

Histopathology, Immunohistochemistry, and Molecular Studies

A biopsy via deep saucerization was performed revealing a dense perivascular and interstitial infiltrate of lymphocytes and plasmacytoid cells extending from the dermis through the subcutis and skeletal muscle plane. Plasmacytoid cells comprised the majority of the infiltrate. Rare immunoblast-like cells and atypical plasmacytoid cells were observed.

- CD79a- Diffusely positive
- CD3- Highlighted smaller population of cells than CD79a
- CD5- Similar staining pattern to CD3
- CD20- Rare clusters of positive cells that highlighted the immunoblastic cells
- CD30- Rare positive cells
- Pax5- Similar staining pattern to CD20
- BCL6 and CD56- Negative

- CD23- Rare clusters of positive cells; no follicular dendritic cell meshwork appreciated
- Kappa-to-lambda ratio of ~1:10, consistent with lambda light chain restriction
- IgA, IgG, IgM- Predominantly IgA positive cells with fewer IgG and rare IgM positive cells.
- EBV-encoded RNA in situ hybridization (EBER ISH)- Diffusely positive
- IGH B-cell gene rearrangement: Failed to detect monoclonal population

References

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