“What makes it a lymphoma?”

Disclosure

- No conflict of interest to disclose
Clinical History

89 year old male with history of Crohn’s disease was found to have multiple necrotic and ulcerating lesions in oral cavity.
• A lip biopsy was performed
Diagnosis:

EBV-associated lymphoproliferative disorder

Differential Diagnosis

1. EBV+ mucocutaneous ulcer

2. EBV+ diffuse large B-cell lymphoma, NOS

<table>
<thead>
<tr>
<th>Location</th>
<th>Systemic nodal/extranodal disease</th>
<th>Skin or Mucosa (oral, GI)</th>
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</thead>
<tbody>
<tr>
<td>Risk Factors</td>
<td>Older age</td>
<td>Immunosuppression</td>
</tr>
<tr>
<td>Microscopy</td>
<td>Large transformed immunoblasts</td>
<td>Large transformed immunoblasts</td>
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<tr>
<td></td>
<td>Hodgkin/Reed-Sternberg-like cells</td>
<td>Hodgkin/Reed-Sternberg-like cells</td>
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<tr>
<td></td>
<td>Inflammatory background</td>
<td>Inflammatory background</td>
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<tr>
<td>Immunophenotype</td>
<td>CD19, CD20, PAX5, CD79a</td>
<td>CD20, PAX5, OCT2</td>
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<tr>
<td></td>
<td>CD34, CD38, MUM1</td>
<td>MUM1</td>
</tr>
<tr>
<td></td>
<td>CD31 and PDL2</td>
<td>CD30 and CD15*</td>
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<tr>
<td>Molecular</td>
<td>Clonal</td>
<td>Clonal or non-clonal</td>
</tr>
<tr>
<td>Treatment</td>
<td>Chemo-immunotherapy</td>
<td>Reduction of Immunosuppression</td>
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<tr>
<td></td>
<td></td>
<td>Rituximab</td>
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<tr>
<td></td>
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<td>Local radiation</td>
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<tr>
<td>Prognosis</td>
<td>Poor</td>
<td>Good</td>
</tr>
</tbody>
</table>
Clinical Information!

**EBV+ MCU**
- No systemic symptoms
  - i.e. fever, night sweats, weight loss
- Localized lesion

**EBV+ DLBCL**
- B-symptoms
- Systemic disease

**EBV+ MUCOCUTANEOUS ULCER**
- Age or immunosuppressant-related immunodeficiency
- Cutaneous and mucosal ulcers, no systemic disease
- Classical Hodgkin lymphoma, diffuse large B-cell lymphoma or monomorphic PTLD-like morphology and immunophenotype
- Angioinvasion and necrosis can be present
- EBER+; serum EBV titers not elevated (unlike PTLD)
- Good prognosis, often spontaneous regression with reduced immunosuppressants
**Take Home..**

- **Diagnostic line:** “EBV+ lymphoproliferative disorder, see comment”

- Discuss likelihood of EBV+ MCU; BUT, include large B-cell lymphoma in the differential

- Patients need to be evaluated for systemic disease

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**THANK YOU**

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