Intravascular Lymphoma: When Tissue is the Issue

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Introduction

- Intravascular large B-cell lymphoma (IVLBCL) is a malignancy characterized by proliferation of abnormal B-cells within blood vessels.
- IVLBCL has in incidence of one case per million people per year.
- Heterogeneous organ involvement leads to diverse clinical manifestations.

Case: A 66 year old man with leg weakness

HPI:
- A 66-year-old man was transferred to the MICU for encephalopathy and respiratory failure in the setting of worsening lower extremity weakness.
- Hospitalized 3 times in preceding 4 months for bilaterally lower extremity weakness.
- He recovered with steroid therapy with shortening duration of remission.

Objective data:
- Exam: ill-appearing, somnolent, tachypneic, with bibasilar crackles, right upper respiratory failure in the setting of worsening lower extremity weakness.
- Vitals: afebrile HR 130 BP 95/63 RR 40 SpO2 90% on HHFNC 40/40

Management:
- Treated with broad spectrum antibiotics. Neurology consulted, recommended brain biopsy which was not within his goals of care.

Objective data:
- FLAIR hyperintensity in the periventricular white matter, subcortical white matter and brainstem. No enhancing lesions or definitive hyperperfusion. (B) Serum and CSF laboratory data, with abnormal values highlighted in bold font.

Management:
- Found to have widely disseminated IVLBCL on autopsy (Figure 2).
- Aspiration event led to cardiac arrest in setting of DNI status.
- CT chest/abdomen/pelvis without malignancy or infection.
- Exam: ill-appearing, somnolent, tachypneic, with bibasilar crackles, right upper

Figure 1 Laboratory and imaging findings on admission to the ICU

Table 1 Sensitivity and specificity of random skin biopsy

<table>
<thead>
<tr>
<th>Authors</th>
<th>Study site</th>
<th>Population</th>
<th>Biopsy method</th>
<th>Sensitivity</th>
<th>Specificity</th>
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<tbody>
<tr>
<td>Matsue et al</td>
<td>Kameda General Hospital, Japan</td>
<td>113 patients with suspected IVLBCL</td>
<td>Incisional</td>
<td>79%</td>
<td>99%</td>
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<tr>
<td>Rosenbaum et al</td>
<td>Massachusetts General Hospital, USA</td>
<td>56 patients with suspected IVLBCL</td>
<td>Incisional or Punch</td>
<td>50%</td>
<td>100%</td>
</tr>
</tbody>
</table>

Conclusions

- The clinical features of Intravascular large B-cell lymphoma vary widely among patients.
- The diagnosis should be considered in patients presenting with fever and neurological deficits of unknown cause, especially in the presence of cytopenias and elevated serum LDH.
- Random skin biopsy, specifically incisional biopsy of 3 or more sites, can offer an alternative to more invasive biopsy testing but should not be used to rule out IVLBCL.

References