Clinical History

• 45-year-old woman presented to outside hospital with non-specific neurological symptoms.

• MRI revealed a 4.6 cm mass within the right frontal horn of the lateral ventricle which extended into the right frontal lobe with surrounding vasogenic edema, which enhanced post-contrast.
Imaging
Thoughts?
Differential

- Reactive
- Leukemic infiltrate (e.g. CLL)
- Diffuse Large B-Cell Lymphoma
- Lymphoplasmacytic Lymphoma/Waldenström's
- Low-Grade B-Cell Lymphoma
- T-Cell Lymphoma
Microscopic Findings

Neuro:
• H&E stained sections show a tumefactive response of mixed inflammatory cells with cholesterol clefts and fibrosis, consistent with xanthogranuloma of the choroid plexus.
• Monomorphic lymphocytic infiltrate.
• Abundant extracellular proteinaceous debris.

Heme:
• H&E stained sections demonstrate an atypical lymphoplasmacytic infiltrate.
• CD20(+) / Pax-5(+) B-cells.
• Kappa and lambda in-situ hybridization show apparent lambda light chain restriction.
IHC of Lymphomatous Cells

- **Positive**
  - CD20
  - Pax-5
  - BCL-2

- **Negative**
  - BCL-6
  - CD30
  - CD10
  - C-kit/CD117
  - CD56
  - Cyclin-D1
  - S-100
  - CD1a
  - CD3
  - CD5
Molecular

- Monoclonal B-Cell Population demonstrating an IgH-FR1-3 rearrangement, likely representing marginal zone lymphoma.
- MYD88 mutation not detected.

Mass Spec

- Extracellular deposits of:
  - Lambda Ig light chains.
  - Alpha Ig heavy chains.
Diagnosis

- Atypical lymphoplasmacytic infiltrate of the choroid plexus with background xanthogranulomatous reaction, most consistent with marginal zone B-cell lymphoma.
Discussion

- Marginal Zone B-cell lymphoma:
  - Significant female predominance.
  - Most often dural-based (80%).
  - 3 previously described in the choroid plexus.
  - Good prognosis.
    - Complete response 77%.
Discussion

• Xanthogranuloma of choroid plexus:
  • Usually an incidental finding (found in 1.6-7.0% of autopsies).
  • Could lead to hydrocephalus.
  • Commonly bilateral, within the lateral ventricles.
References


