

Diagnostic Slide Session Case #11

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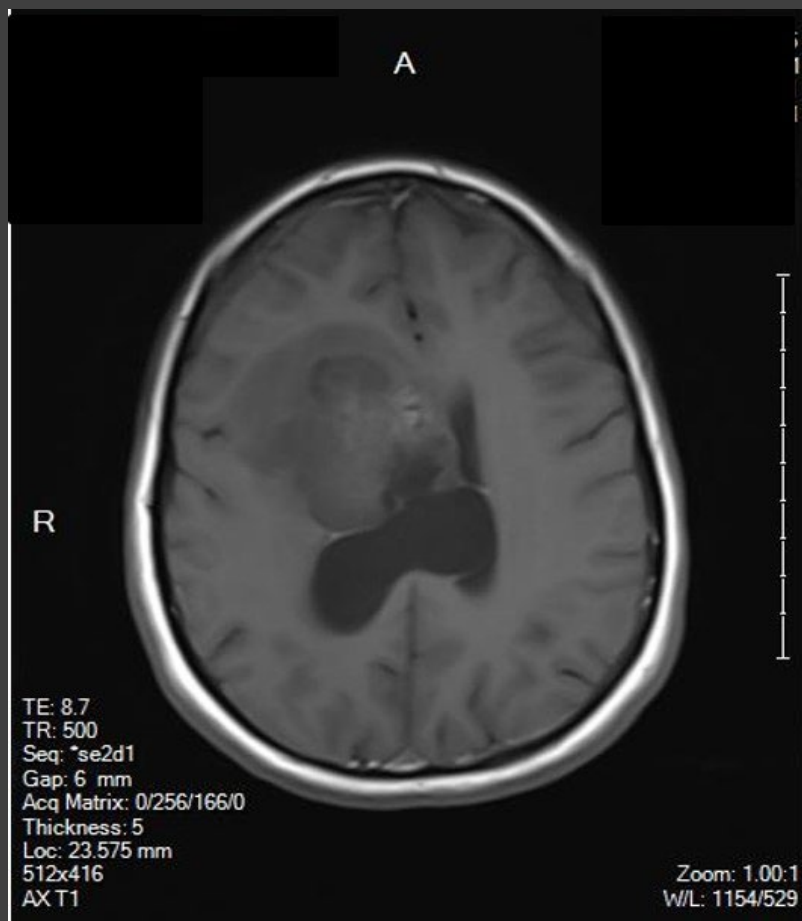
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Clinical History

- 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

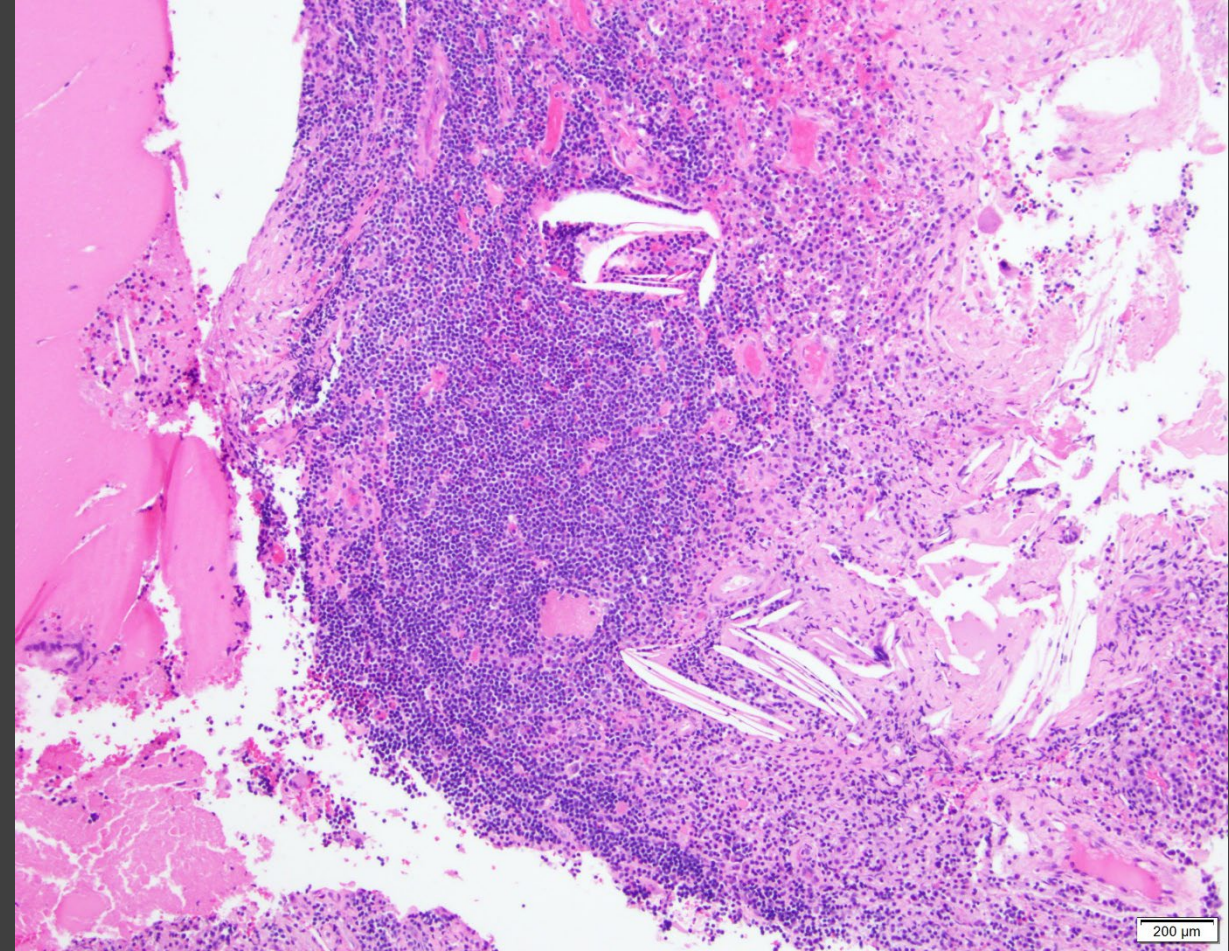
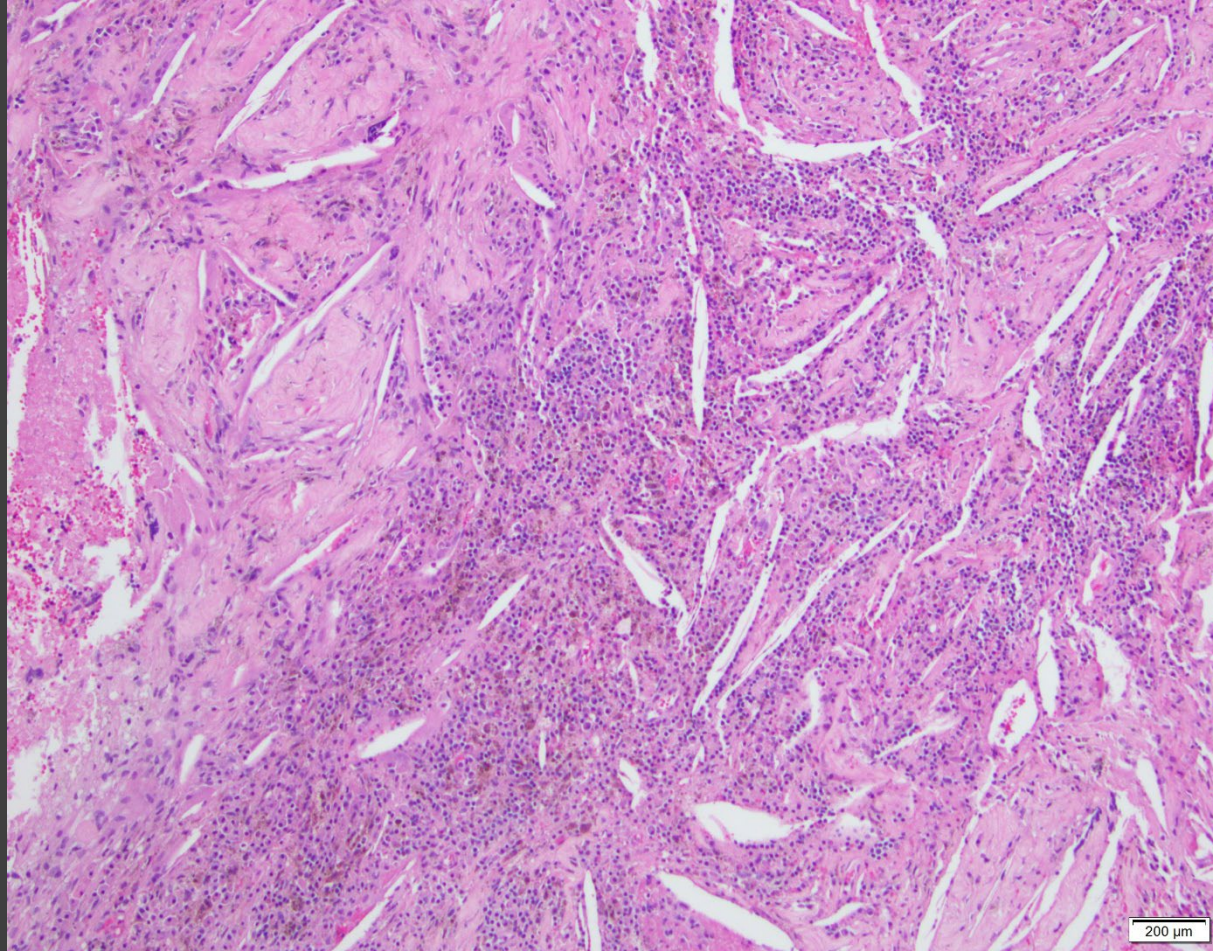
T₁



T₂



H&E



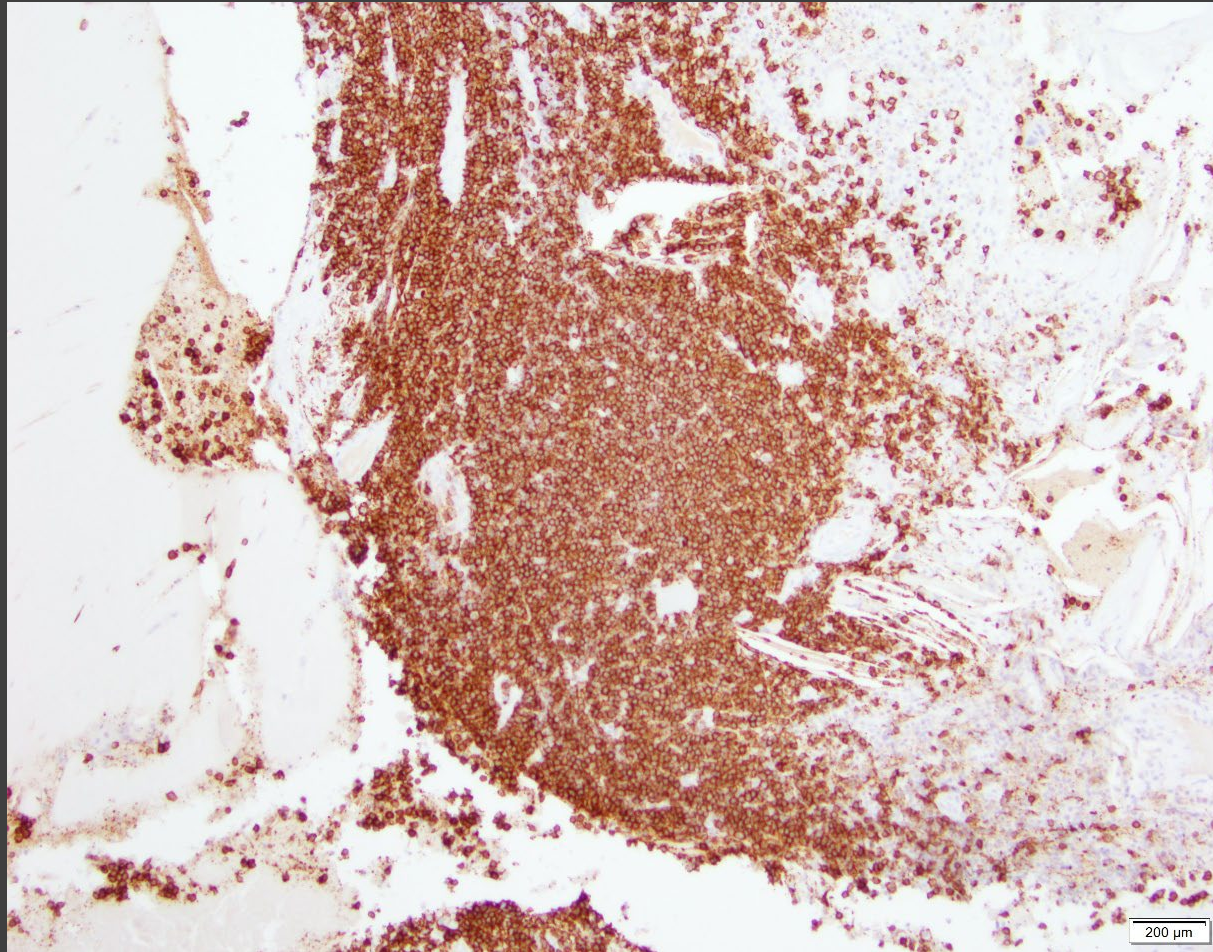
Thoughts?



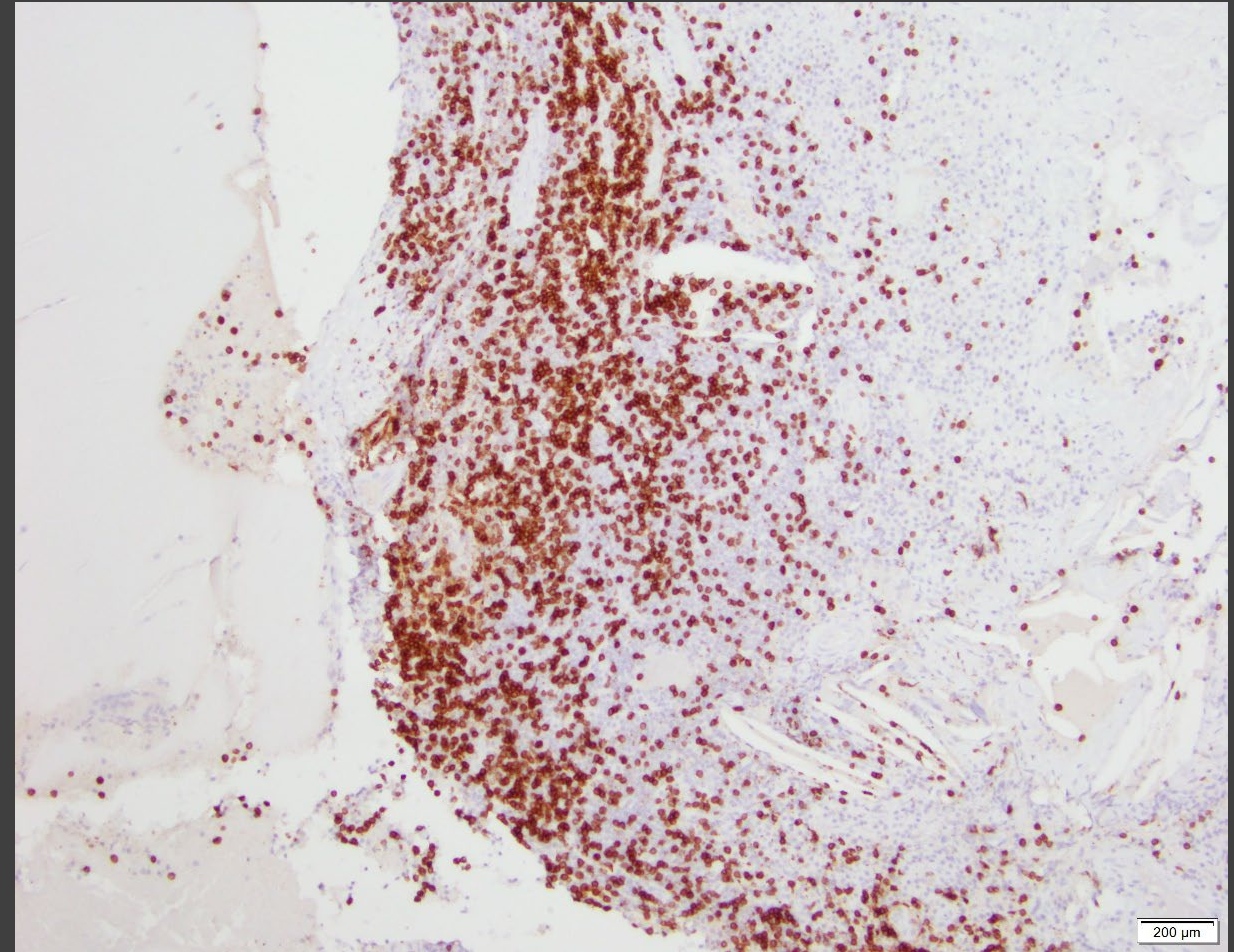
Differential

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

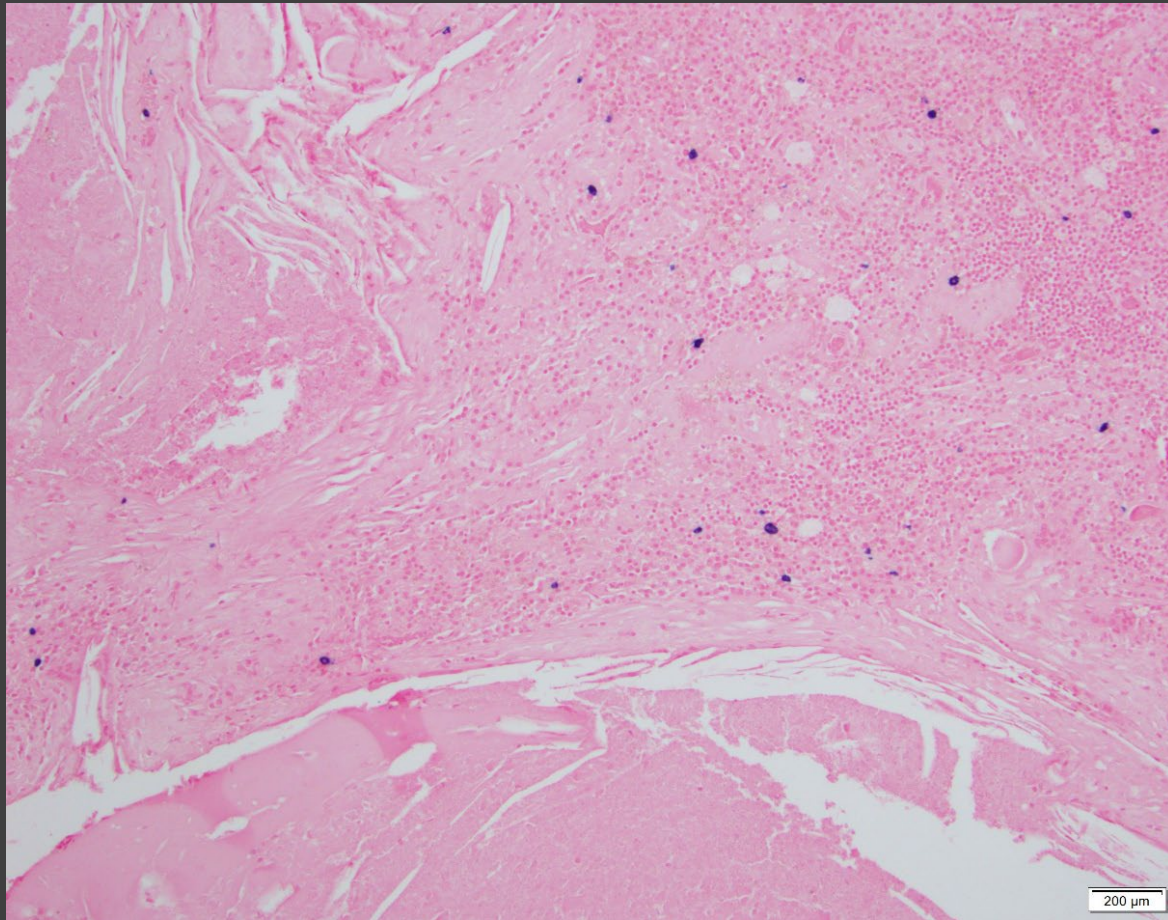
CD20



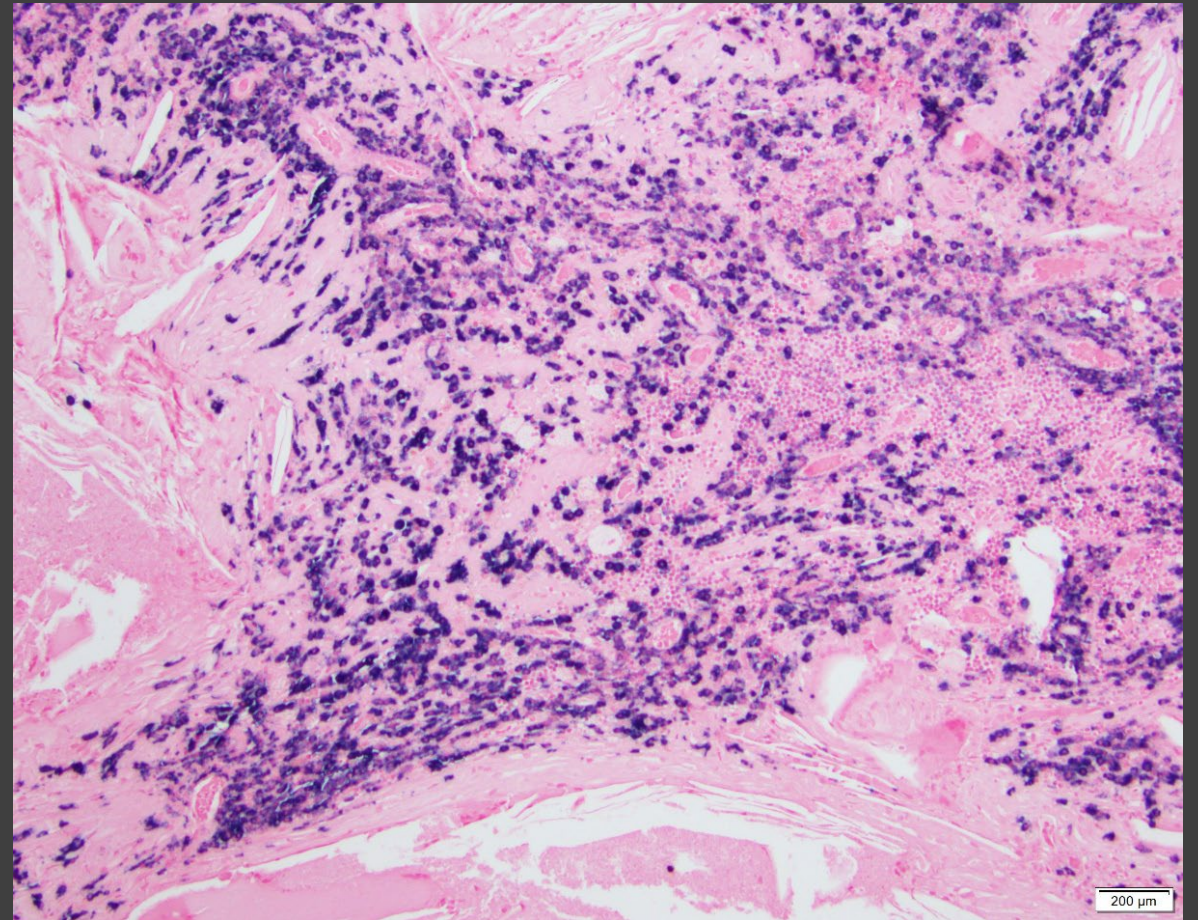
CD3



Kappa ISH



Lambda ISH



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

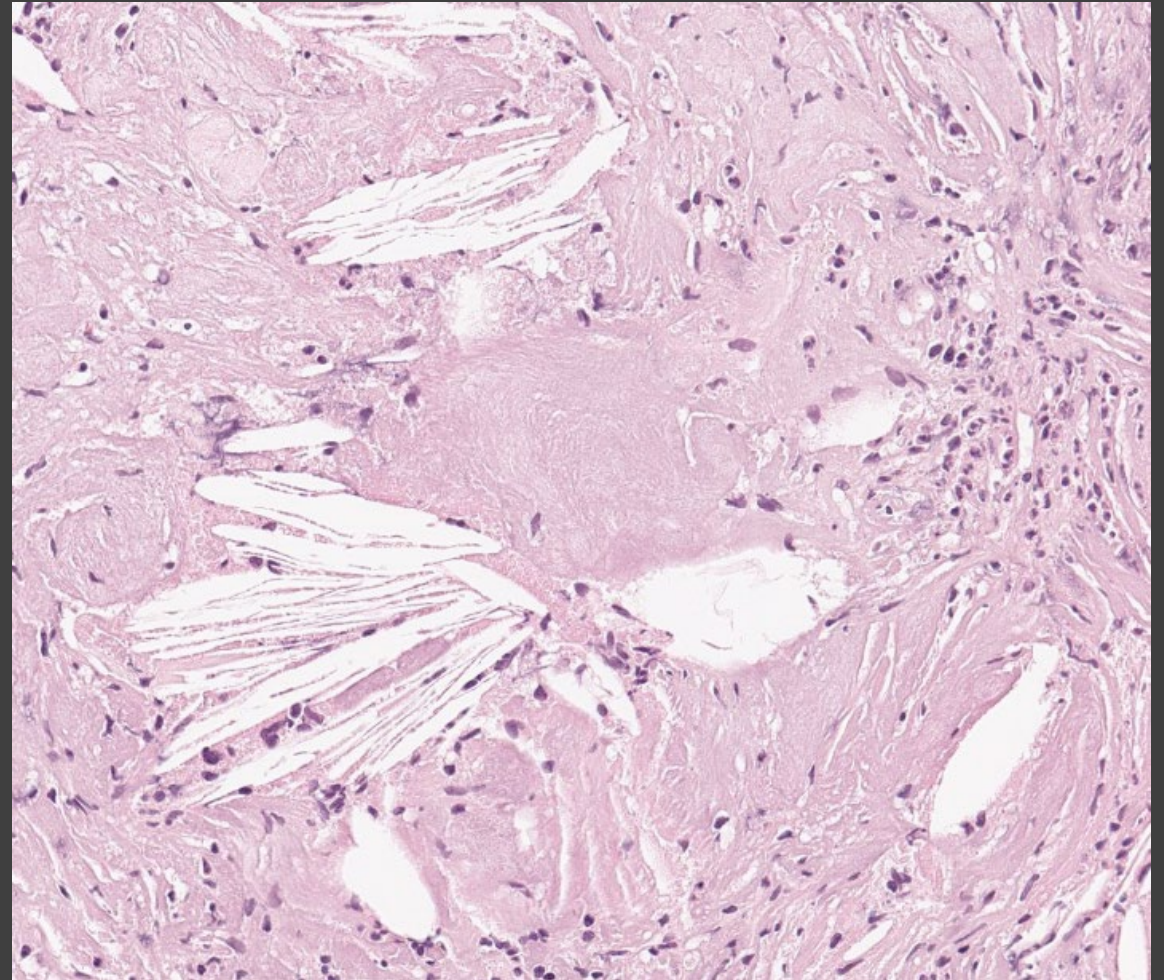
- Atypical lymphoplasmacytic infiltrate of the choroid plexus with background xanthogranulomatous reaction, most consistent with marginal zone B-cell lymphoma.

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

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- Kelley, T. W., Prayson, R. A., Barnett, G. H., Stevens, G. H., Cook, J. R., & Hsi, E. D. (2005). Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue arising in the lateral ventricle. *Leukemia & lymphoma*, 46(10), 1423–1427. <https://doi.org/10.1080/10428190500205895>
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- S. Kumar, D. Kumar, E.P. Kaldjian, S. Bauserman, M. Raffeld, E.S. Jaffe **Primary low-grade B-cell lymphoma of the dura: a mucosa associated lymphoid tissue-type lymphoma** Am J Surg Pathol, 21 (1997), pp. 81-87