A case of rapidly progressive dementia

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Disclosure statement

- I have no relevant financial relationship(s) or nonfinancial relationship(s)

Case presentation

- A middle-aged man from rural Utah
- Progressive wheezing, hyperventilation, muscle spasms and parasthesias in right arm, back and neck
- Acutely delirious on admission
- Transferred to higher acuity care center where he showed florid psychosis, dysphasia, dystonic posturing, akathisia and orofacial dyskinesia
- PMH: Poland syndrome, NSTEMI, GERD, HTN and hyperlipidemia

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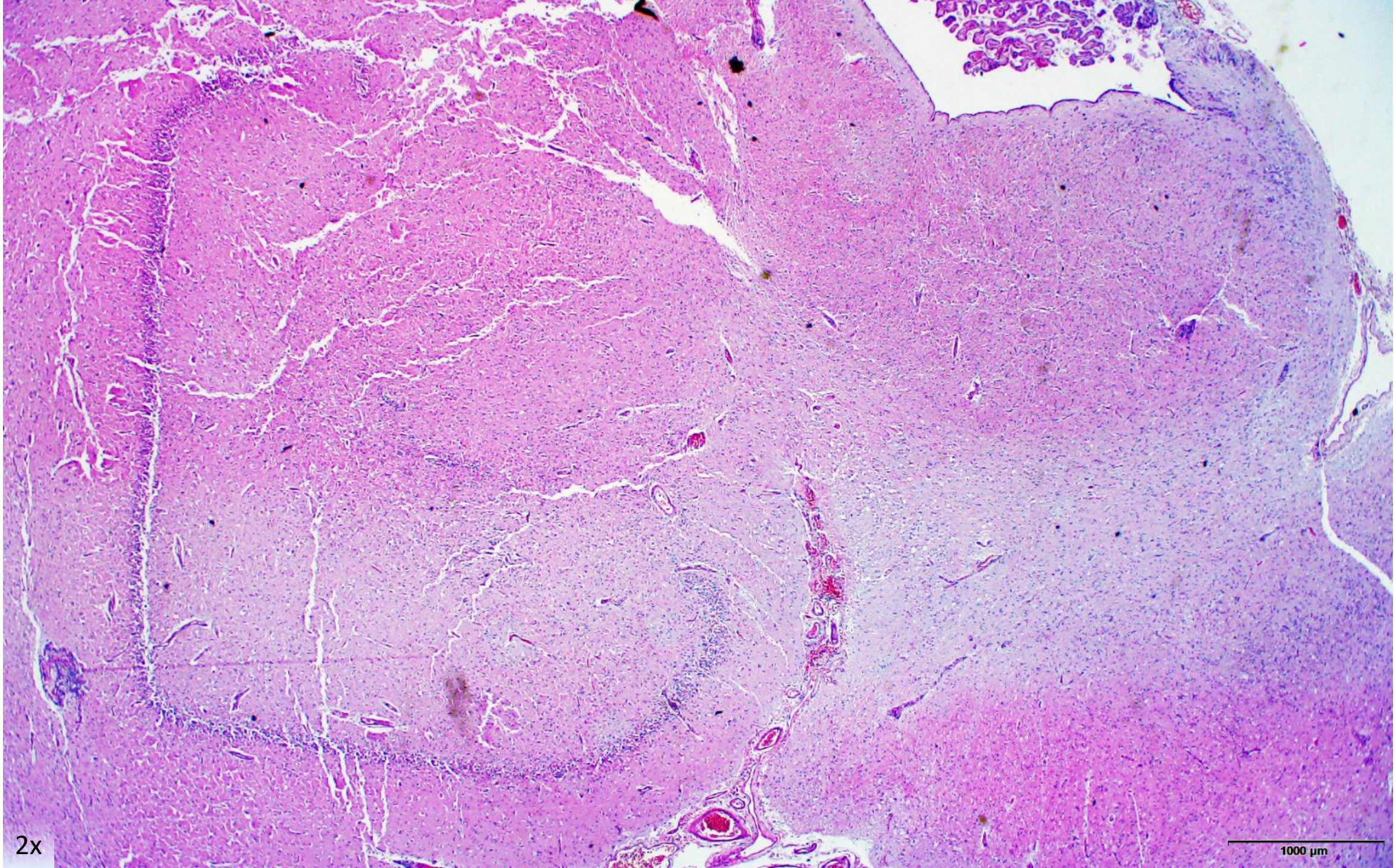
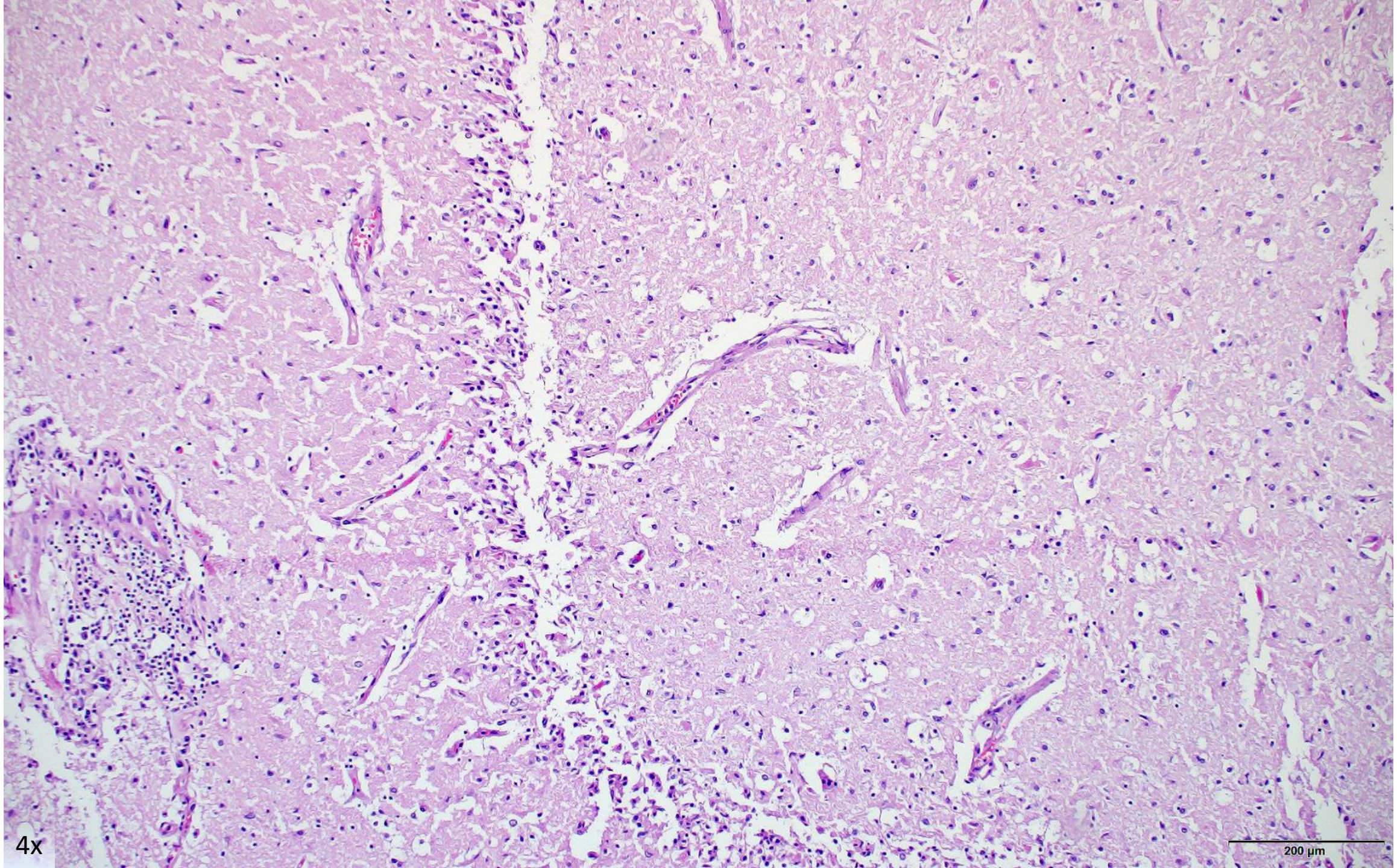
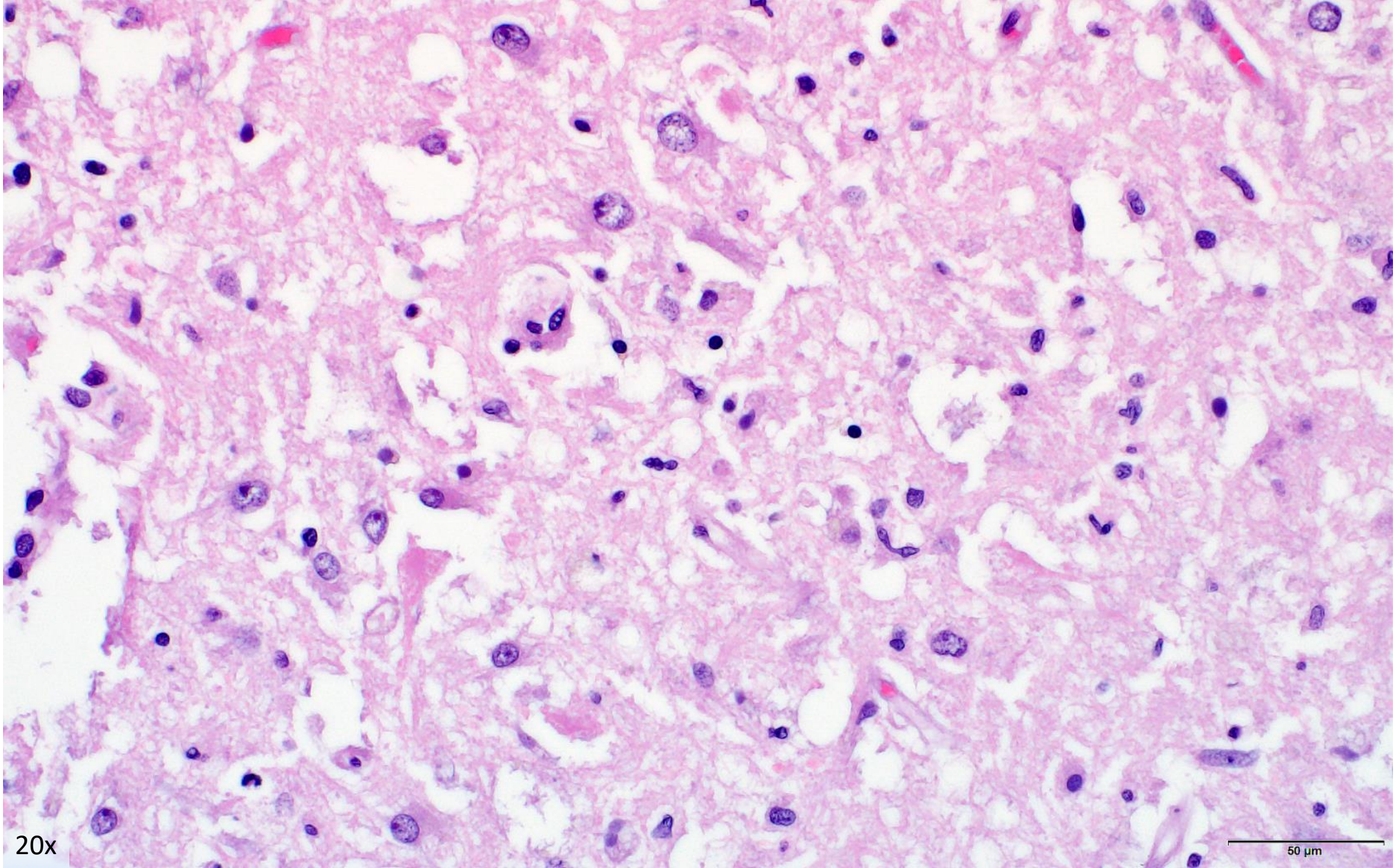
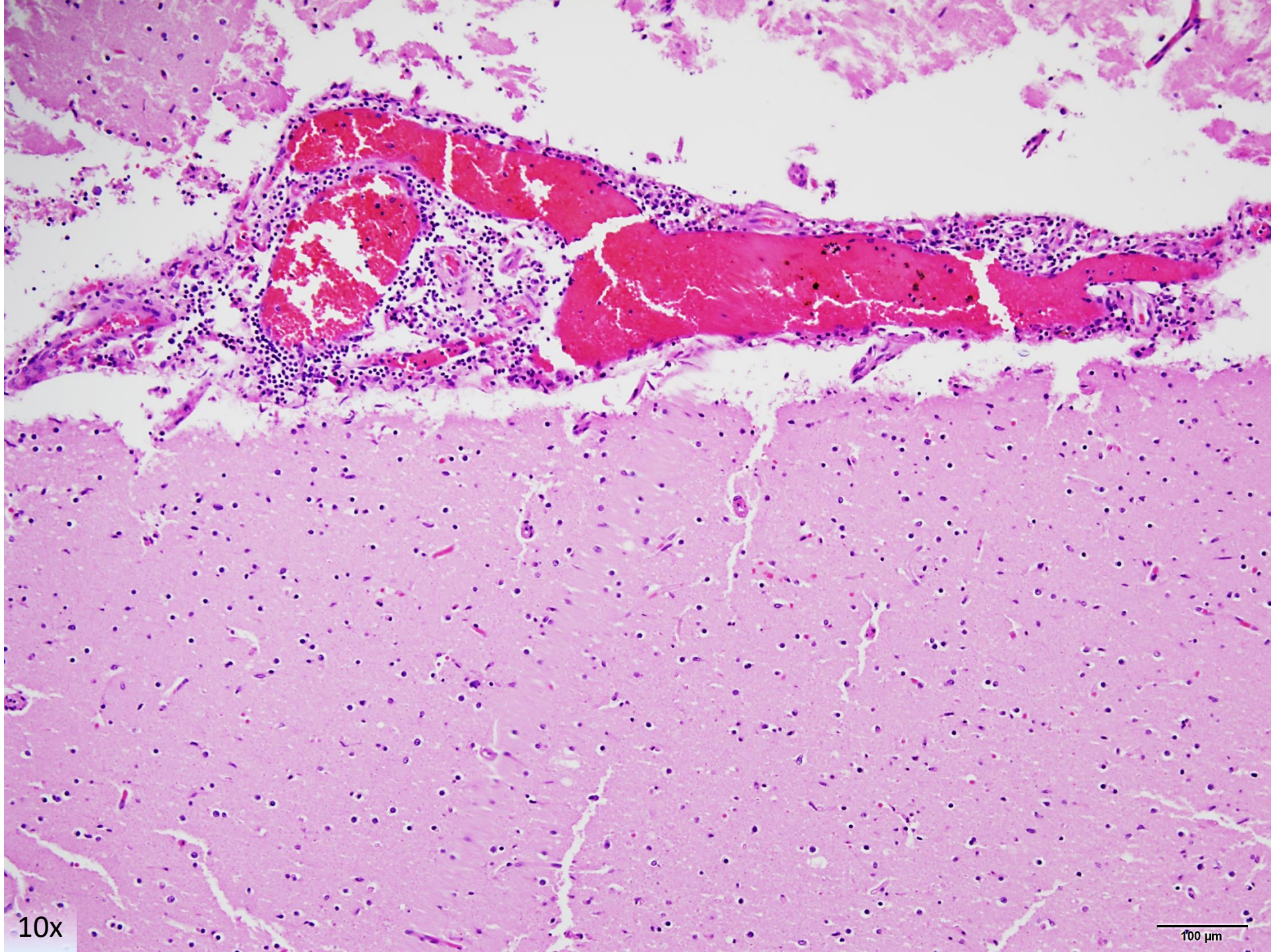
A. SPECIMEN LABELED “HIPPOCAMPUS”

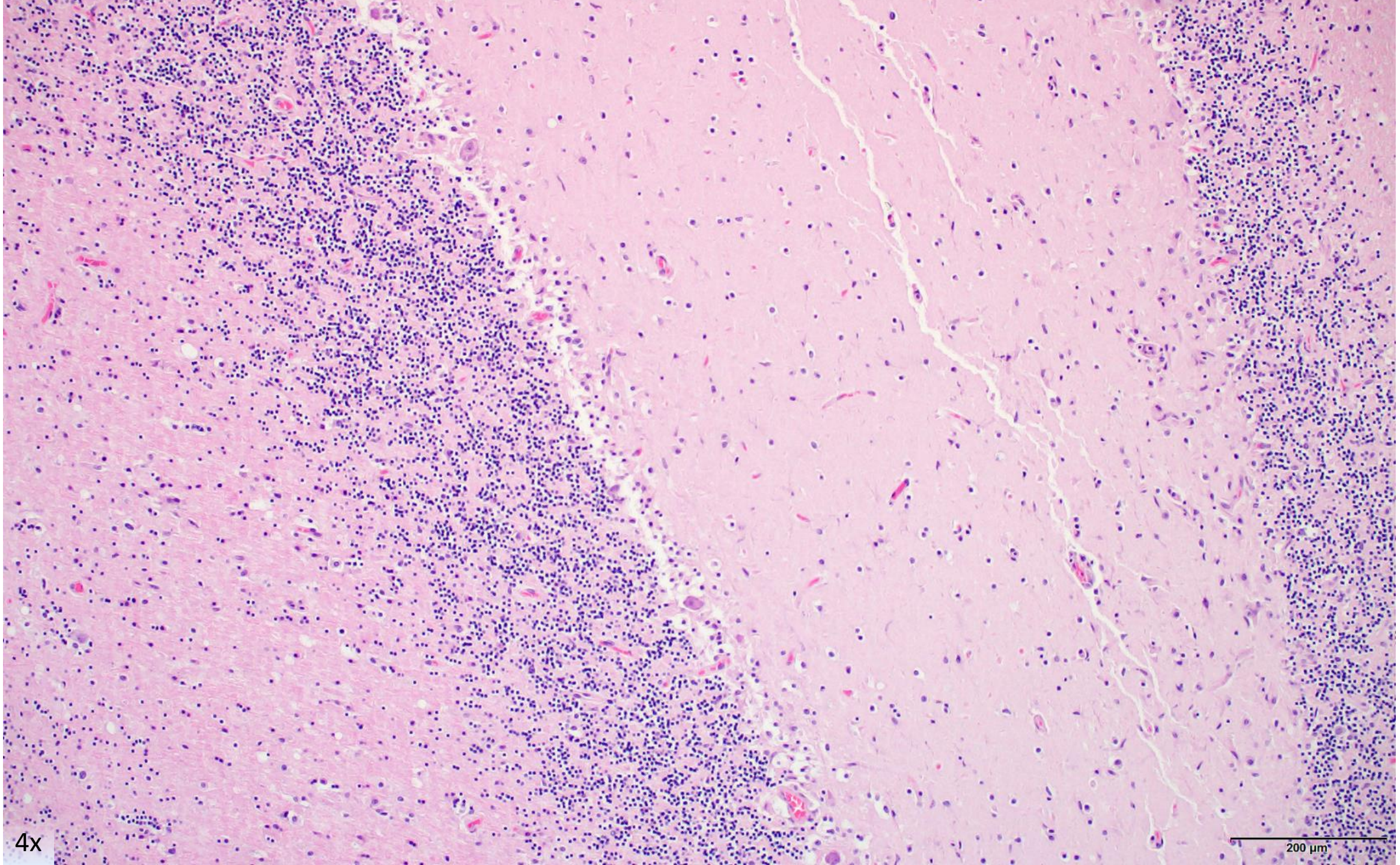
B. SPECIMEN LABELED “CEREBELLUM”

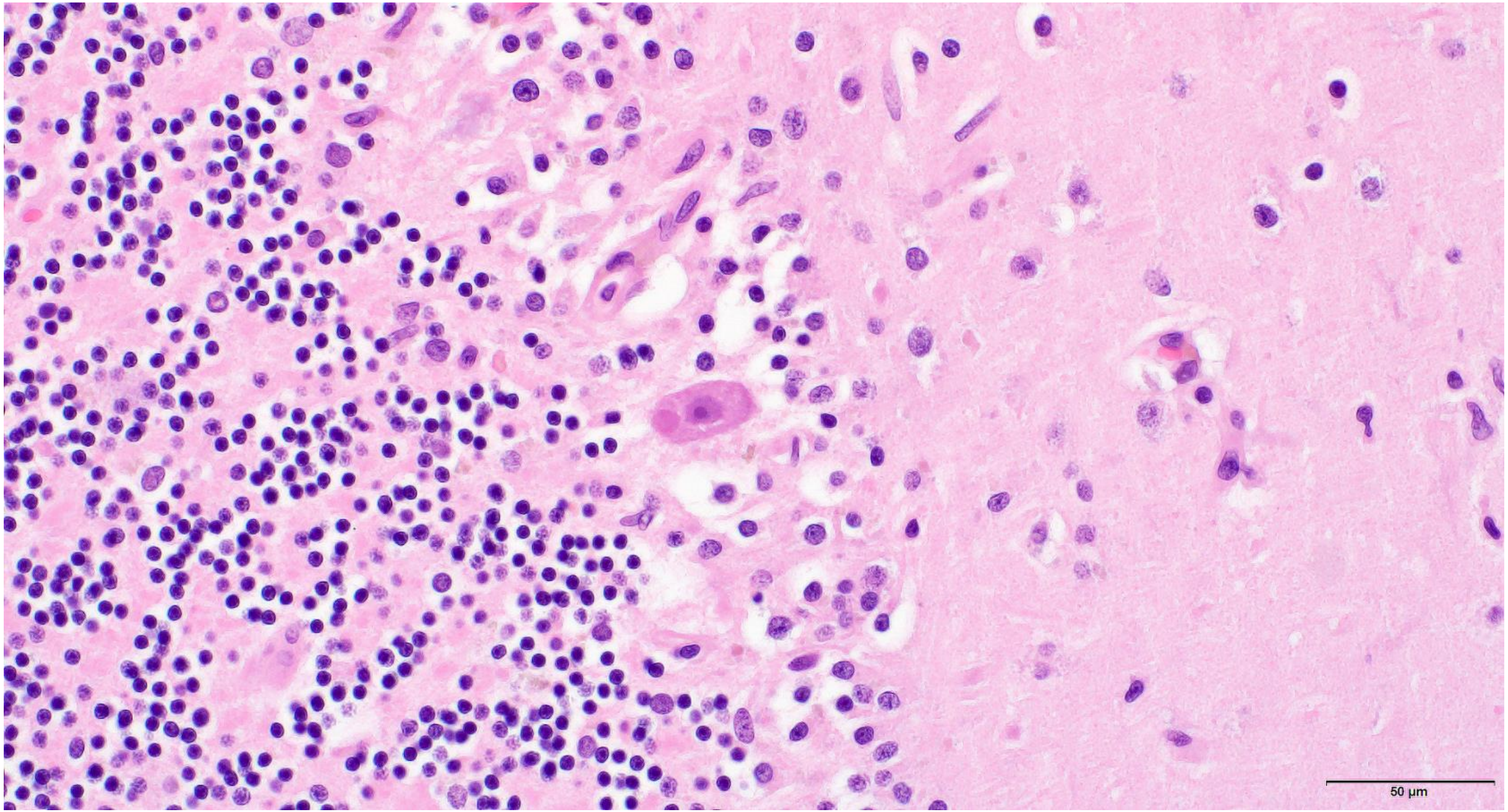

2x
1000 µm

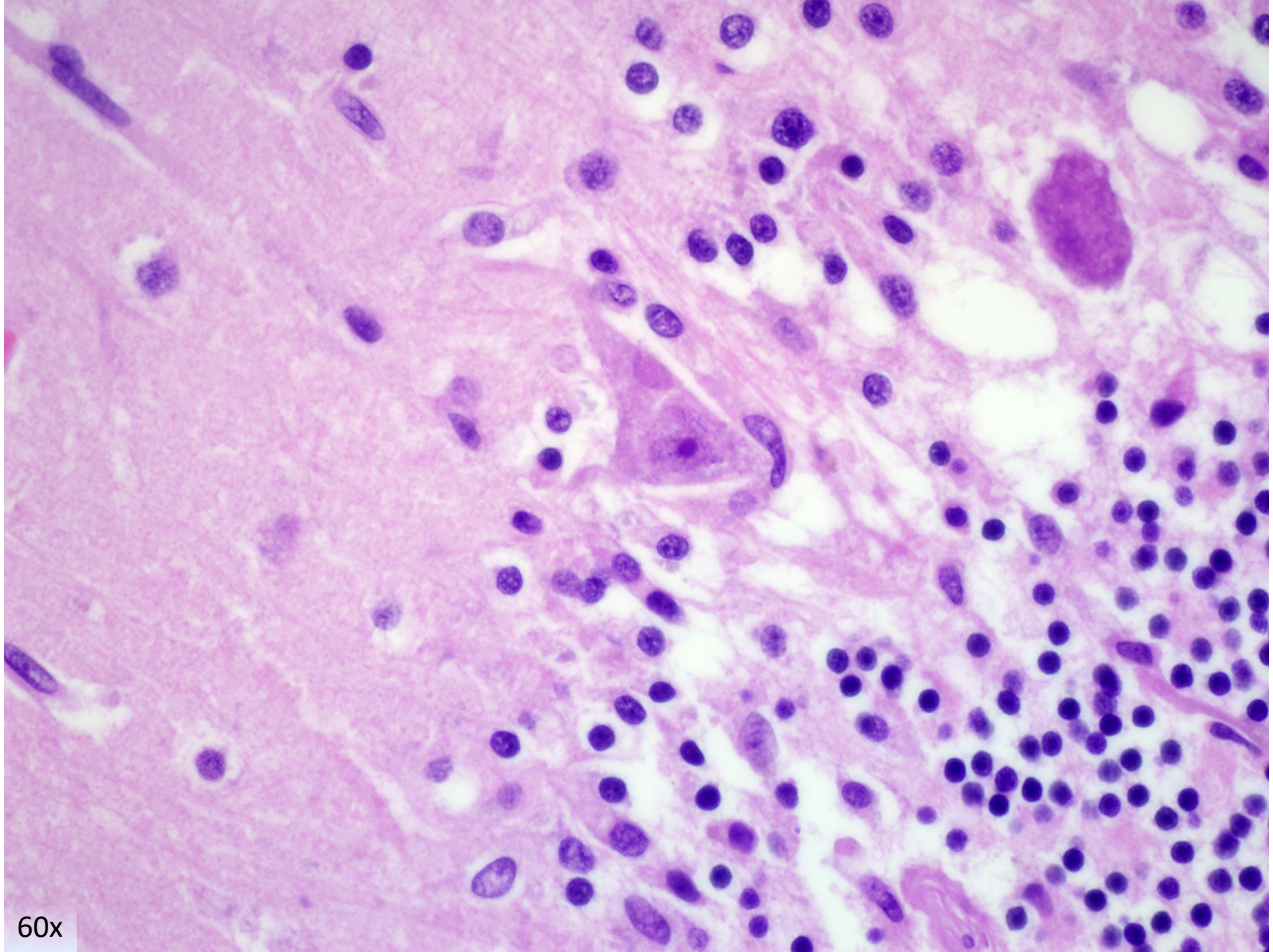

4x
200 µm


20x
50 µm


10x
100 µm


4x
200 µm


50 µm


60x

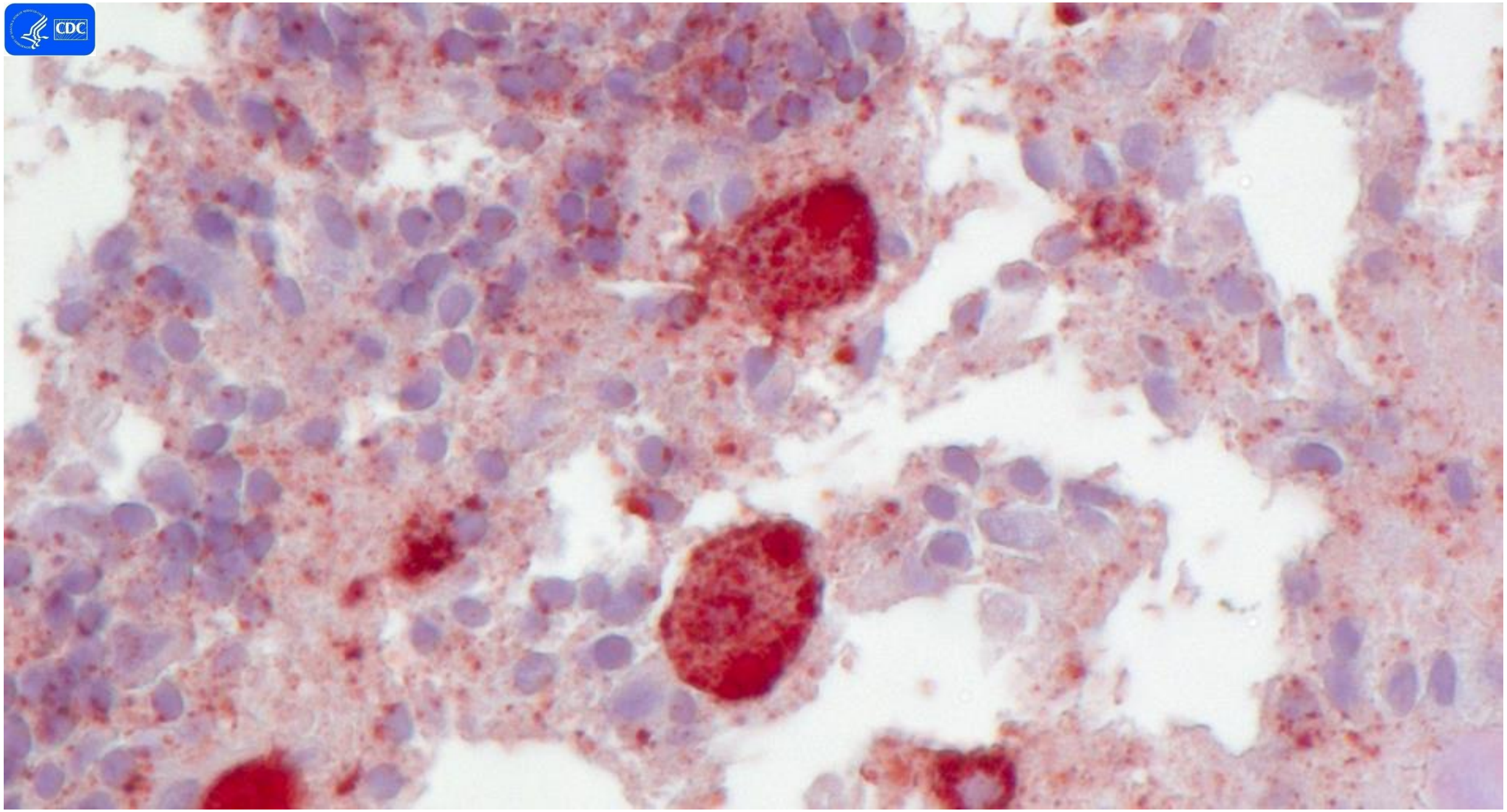
Discussion

- Histologic characteristics
- Differential diagnosis (imaging, histology)?

Pathologic Diagnosis

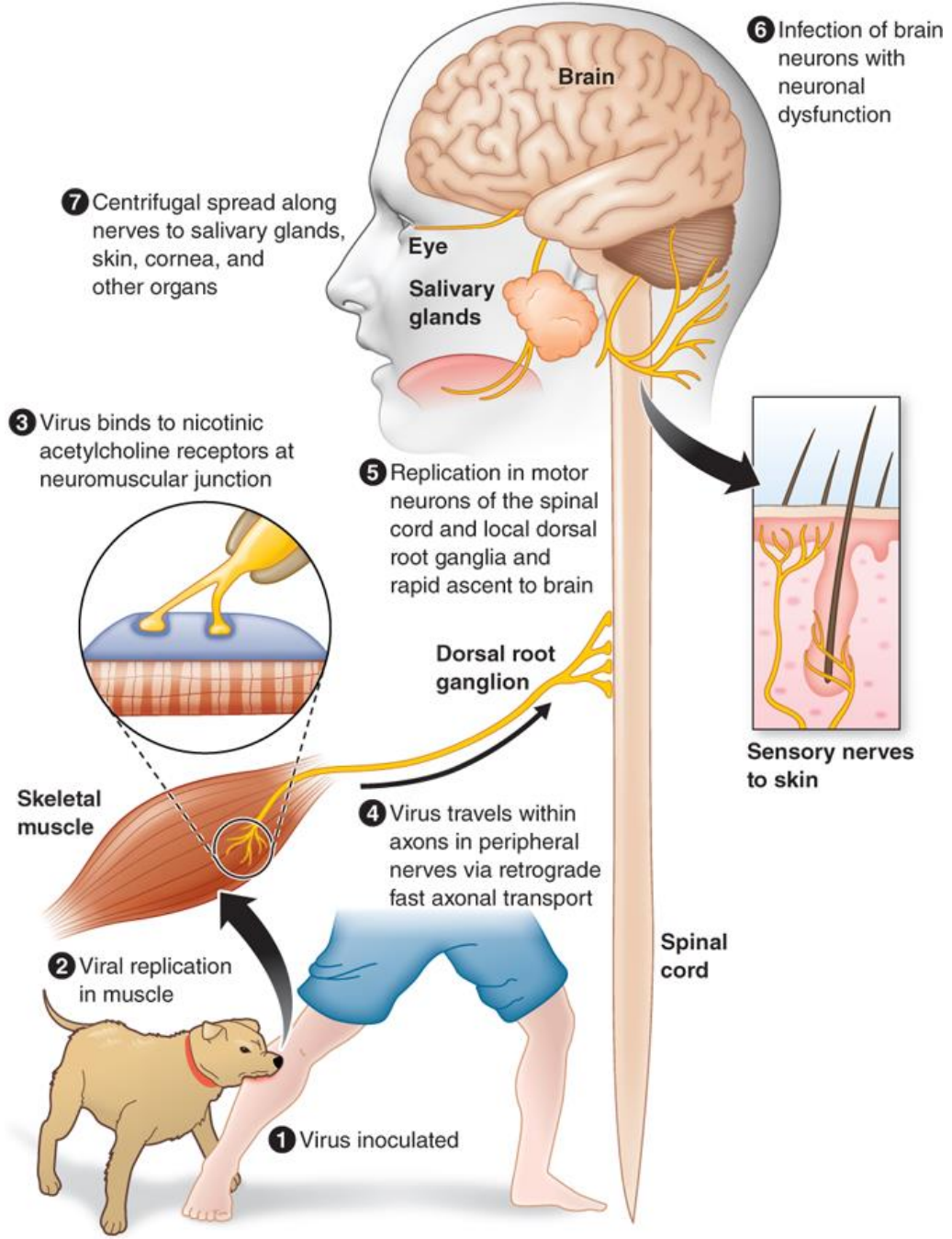
1, 2. HIPPOCAMPUS AND CEREBELLUM:

RABIES MENINGOENCEPHALITIS



Rabies - infection

- Negative sense ssRNA unsegmented virus
- NCAM (CD56) and nAChR at NMJ
- Incubations 30 – 90 days
- Centripetal spread early
- Meningoencephalomyelitis
- Centrifugal spread late – cornea, nuchal skin, heart, salivary/lacrimal, lungs, GI, adrenals, kidneys and bladder

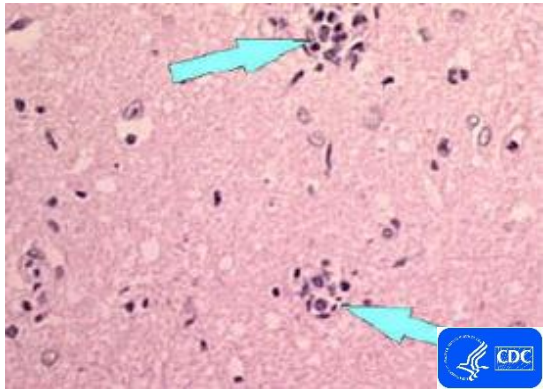


Source: J.L. Jameson, A.S. Fauci, D.L. Kasper, S.L. Hauser, D.L. Longo, J. Loscalzo: Harrison's Principles of Internal Medicine, 20th Edition

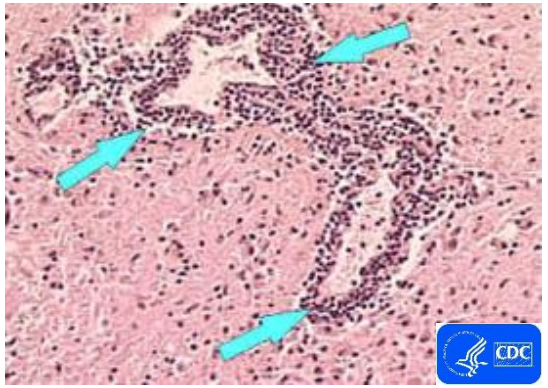
Rabies

Meningoencephalomyelitis

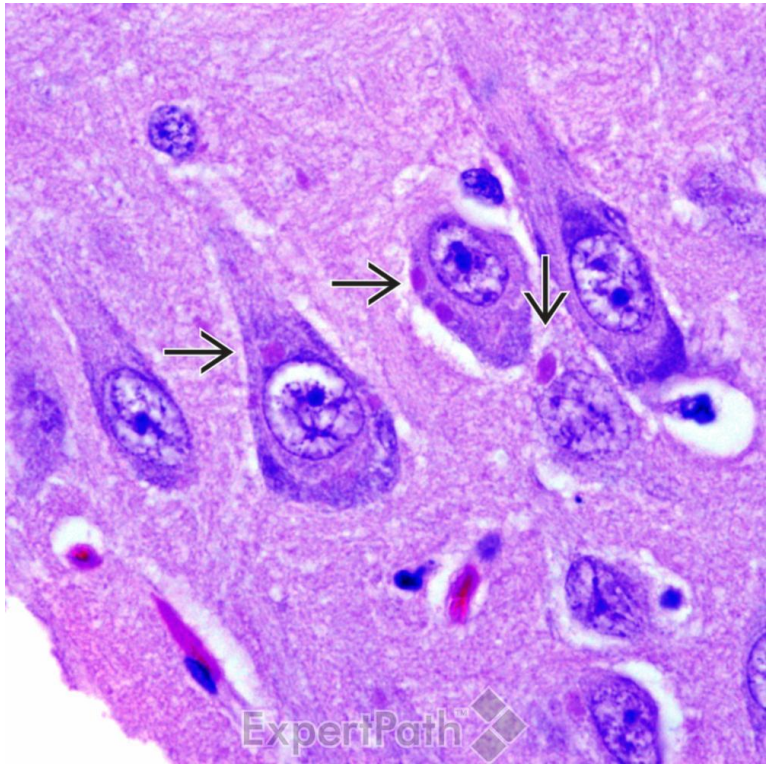
- Brain/spinal cord edema or normal
- Negri bodies- nucleocapsid material
  - Purkinje cells, hippocampal pyramidal cells and brainstem nuclei
  - rarely astrocytes/oligodendrocytes
  - not required for diagnosis
- Inflammatory disparity
- CSF pleocytosis



Babes nodules



Perivascular lymphocytic cuffing



Rabies

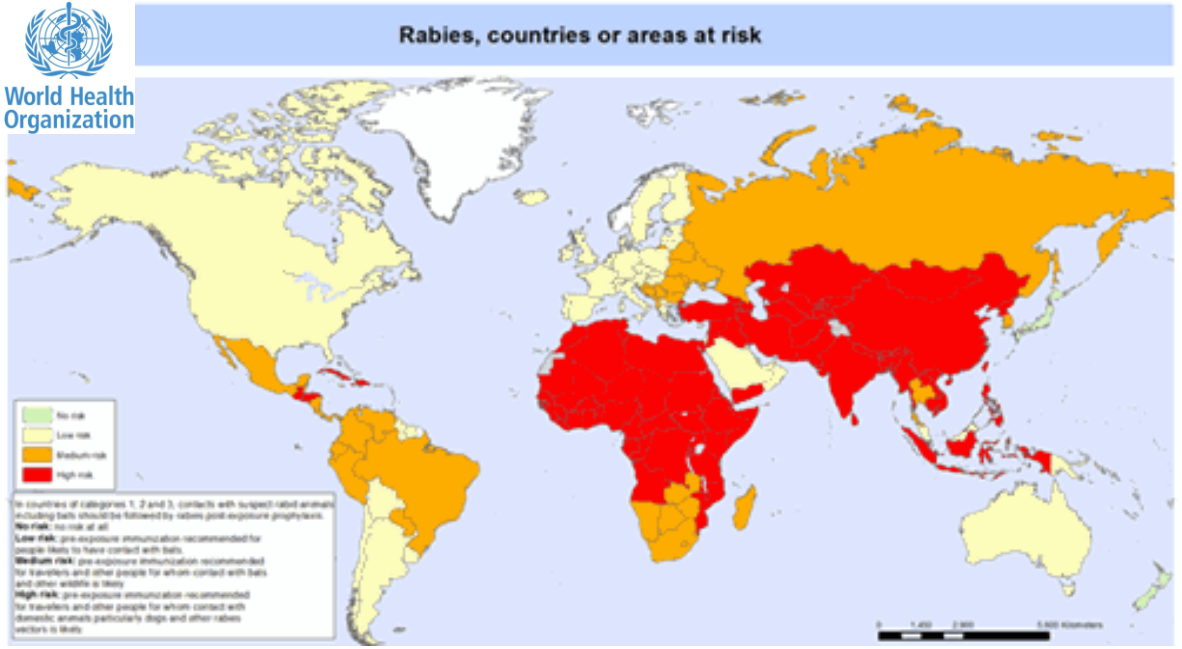
Meningoencephalomyelitis

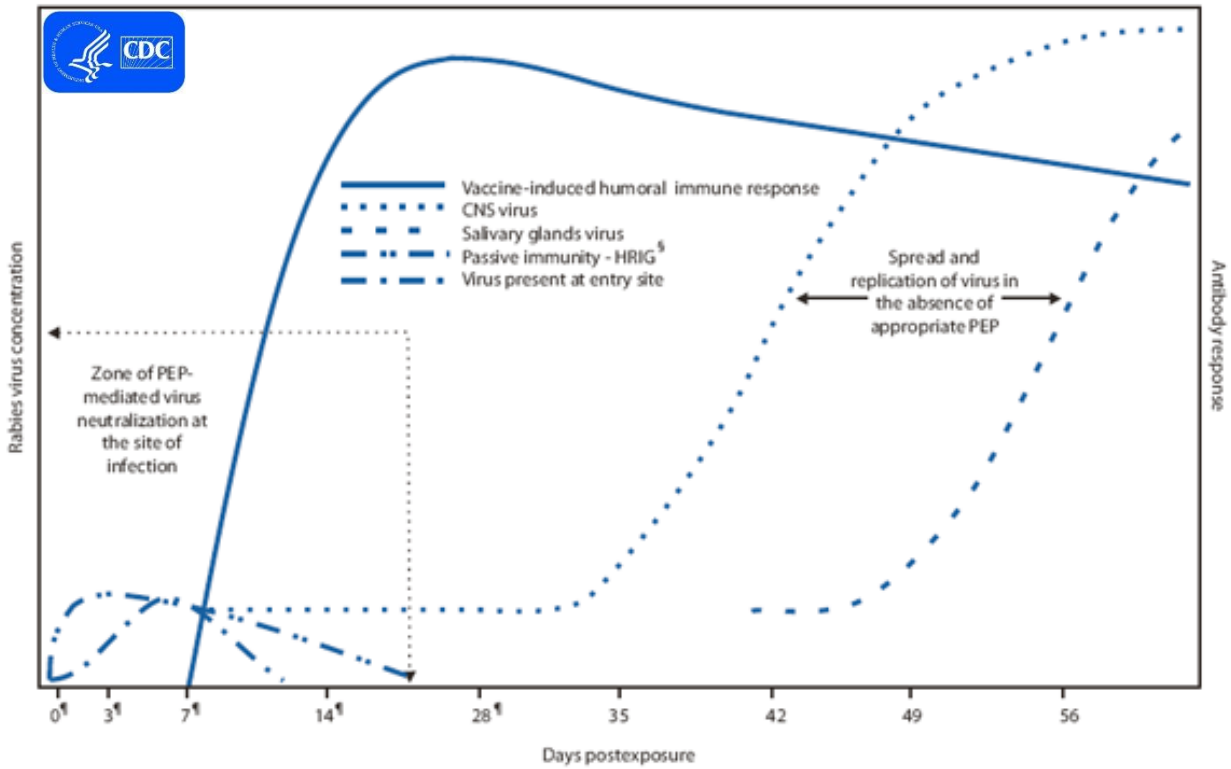
- 59,000 annual human deaths worldwide
- 95% of human deaths in Asia and Africa
- Dogs most common vector worldwide
- In the US:
  - Raccoons (41%)
  - Skunks (29%)
  - Bats (14%)
  - Foxes (5%)
- Treatment:

  - pre/post exposure prophylaxis – passive RIG and vaccine

  - symptomatic – analgesia, sedation and seizure control

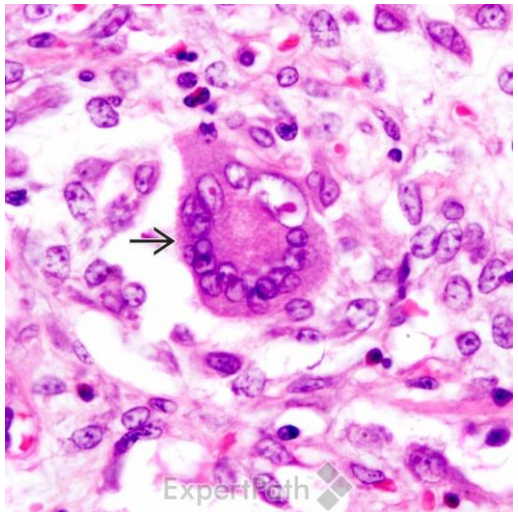
- DDX: VITAMINS, other viruses (Measles), prion disease





Other Viral infections

- Measles – rubeola
  - perivascular inflammation
  - gliosis
  - loss of neurons
  - giant cells with eosinophilic cytoplasmic inclusions
  - ground glass intranuclear inclusions



Acknowledgements

- Patient, family and clinical teams
- Utah OME
- Lily Marsden, MD
- CDC

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Thanks for your attention

Questions?