

DSS Case #7

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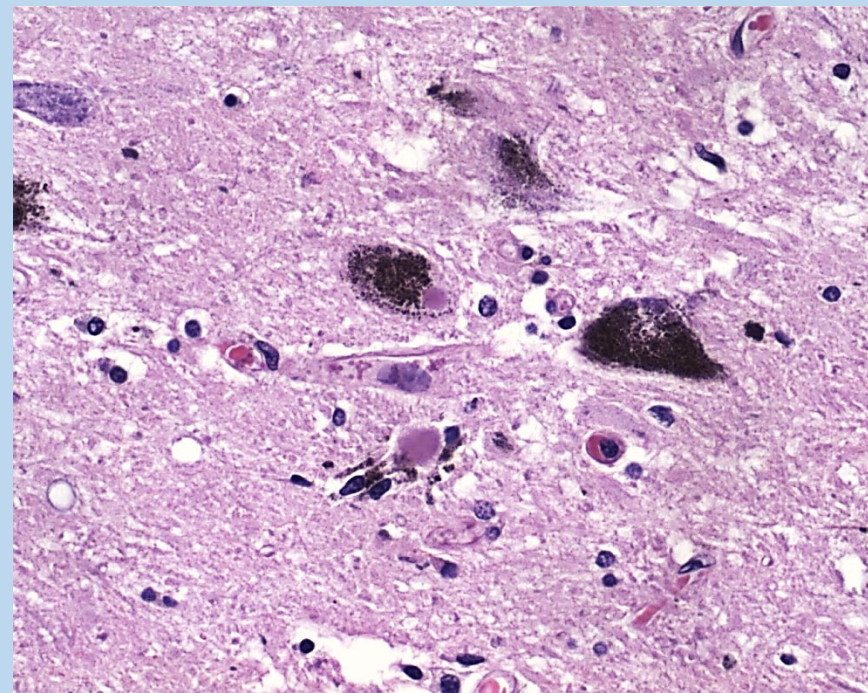
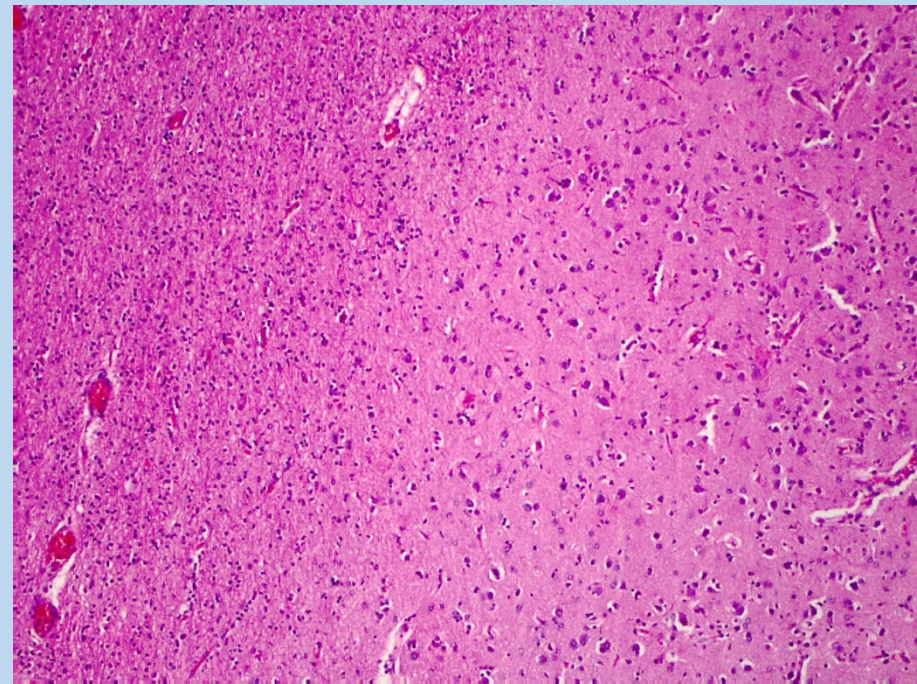
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Disclosures

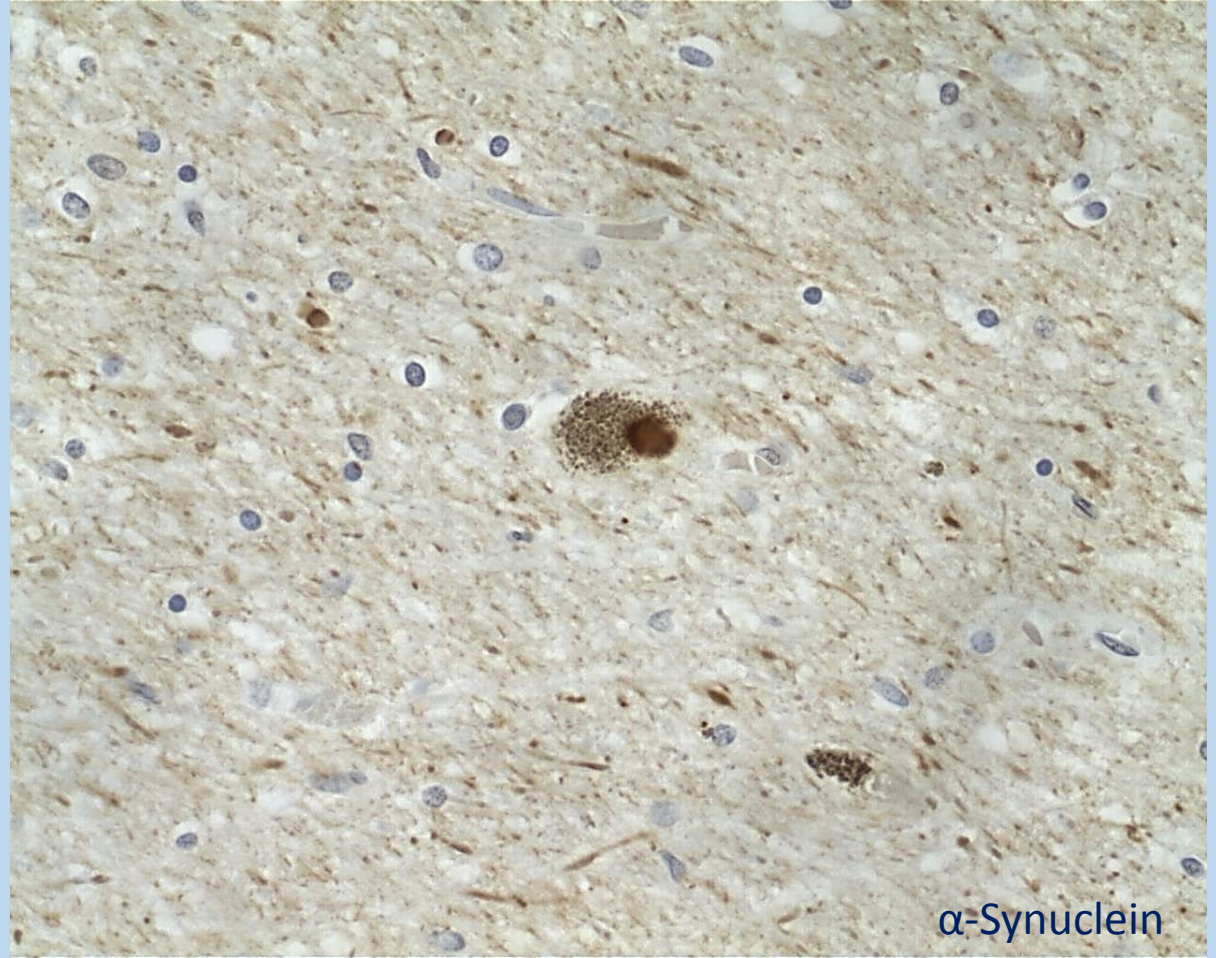
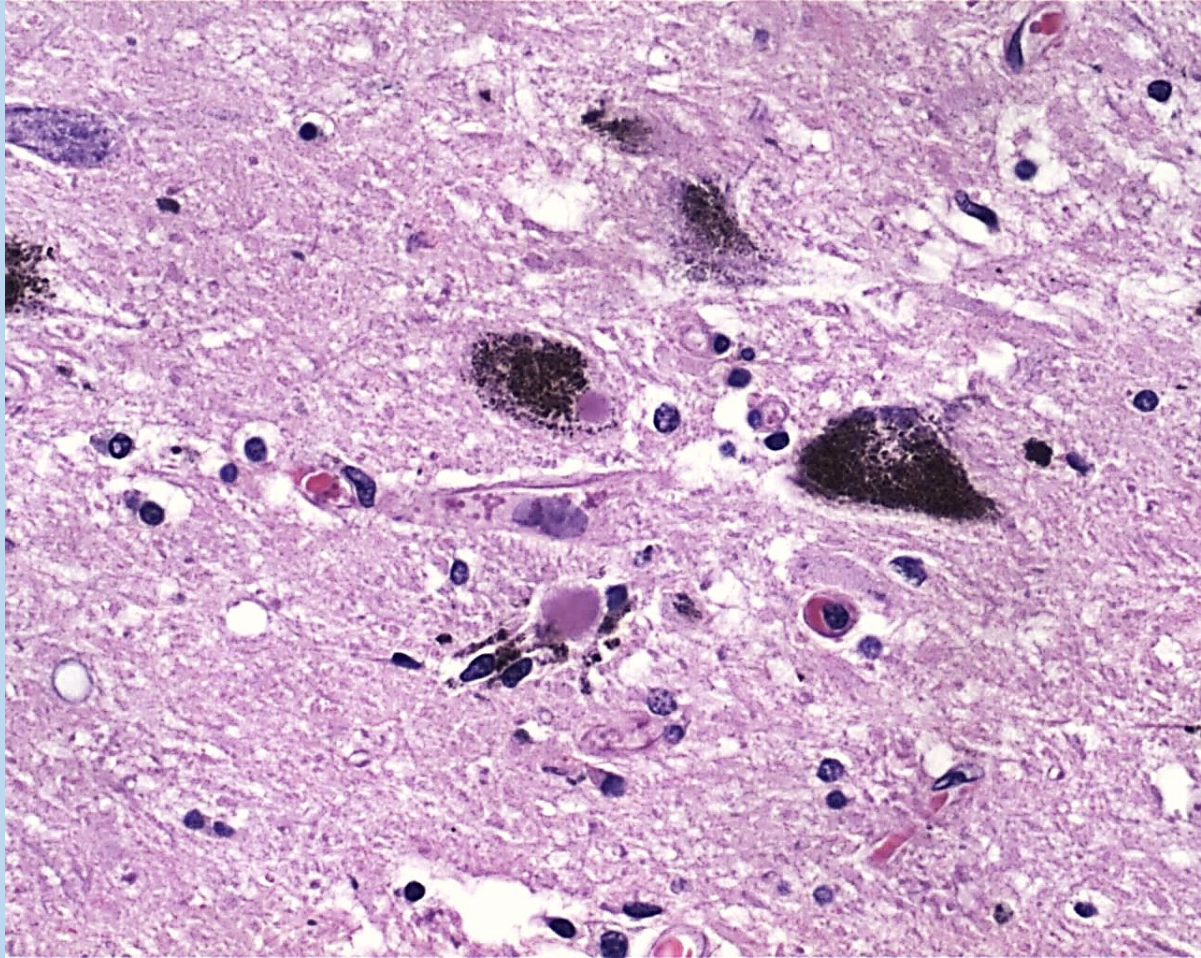
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We have no relevant financial disclosures.

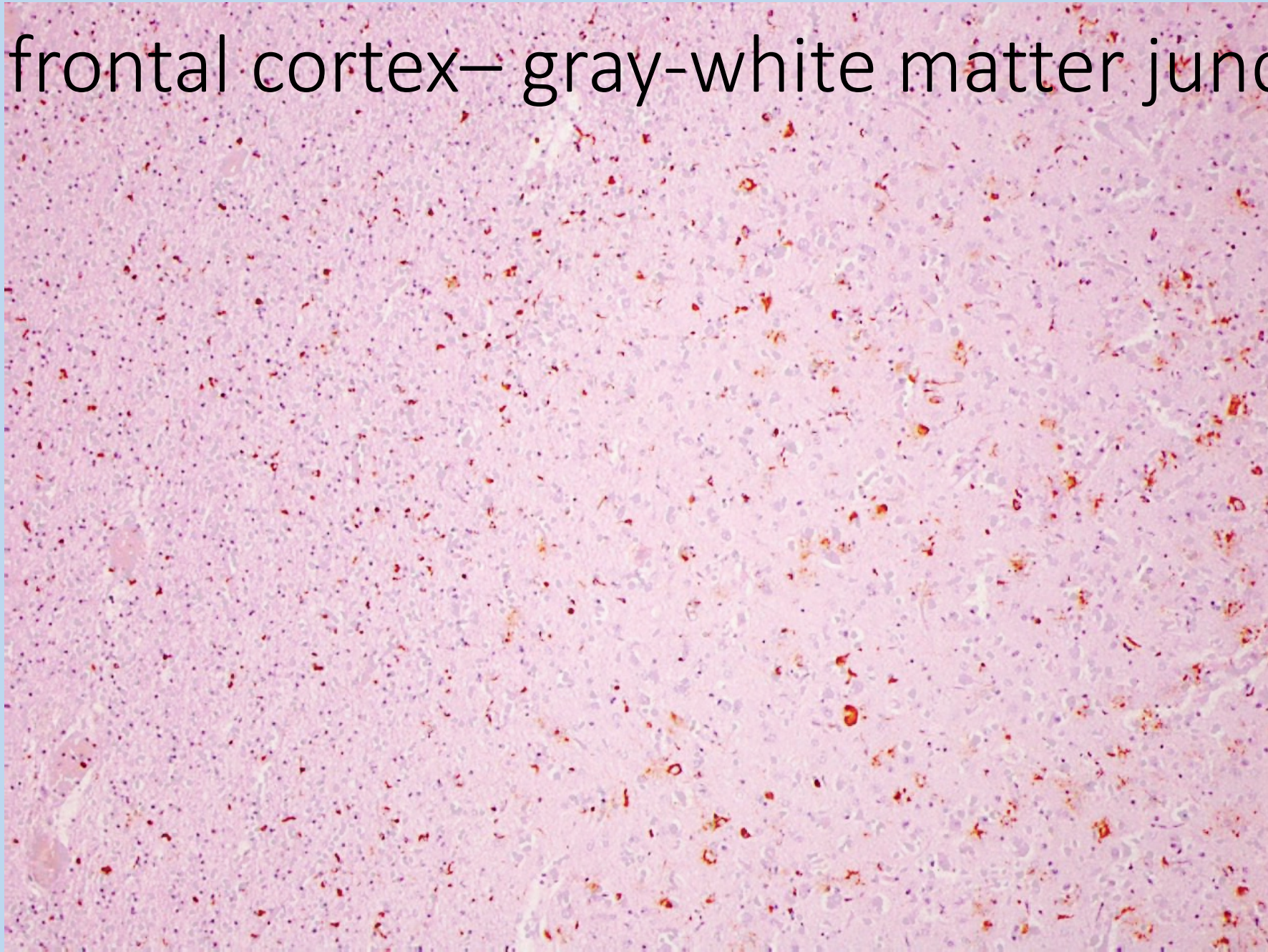


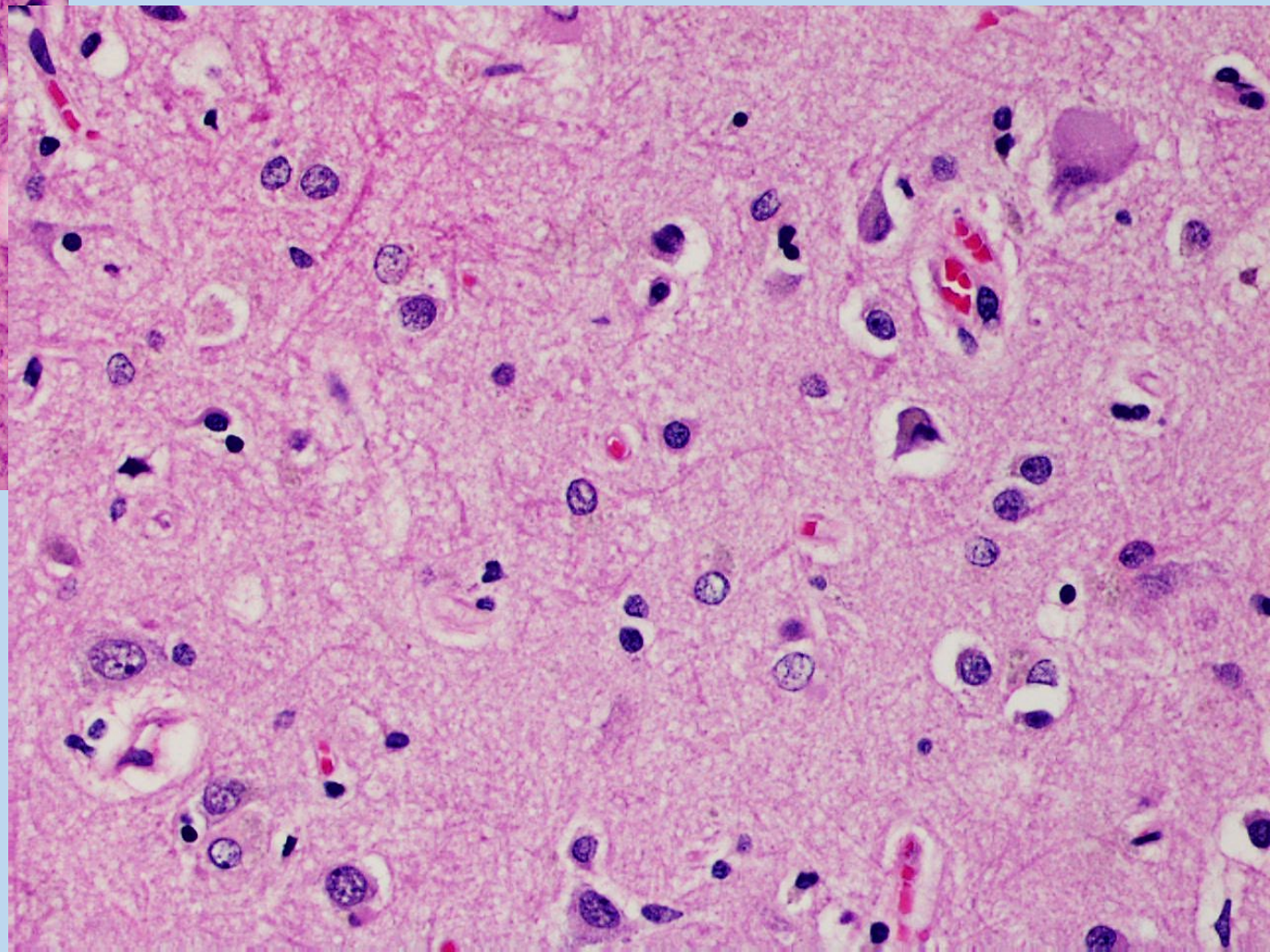
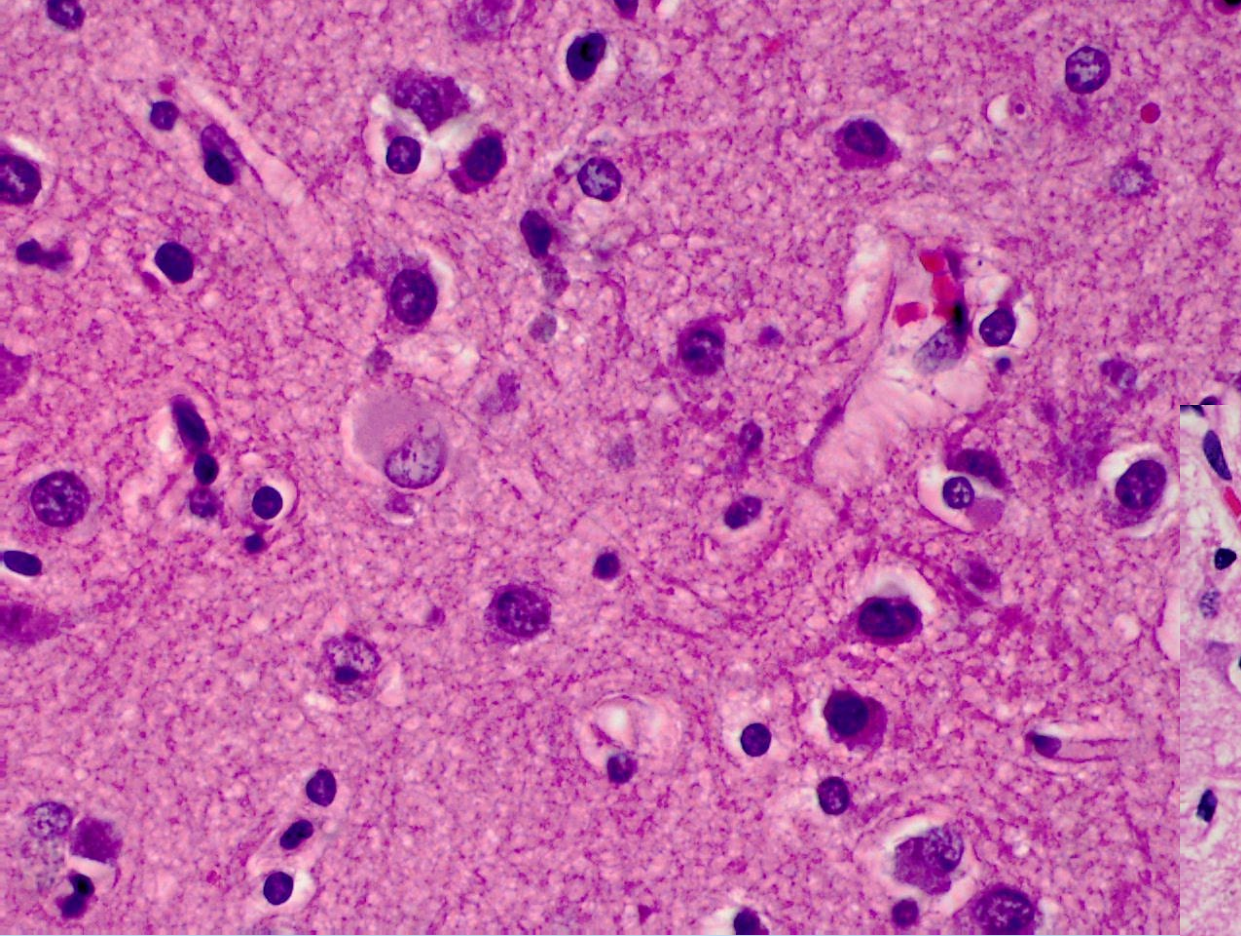
Discussion

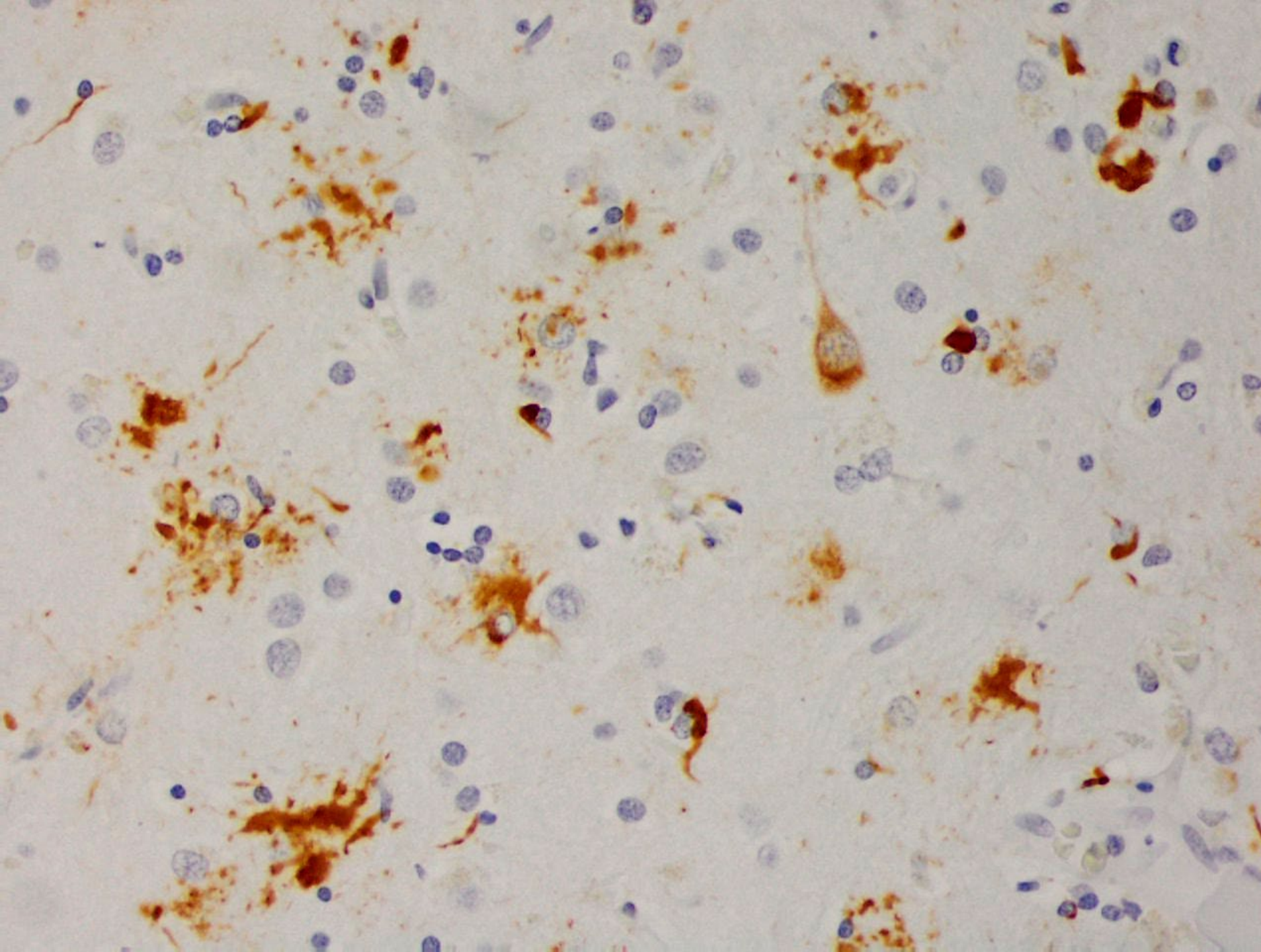




Mid frontal cortex— gray-white matter junction



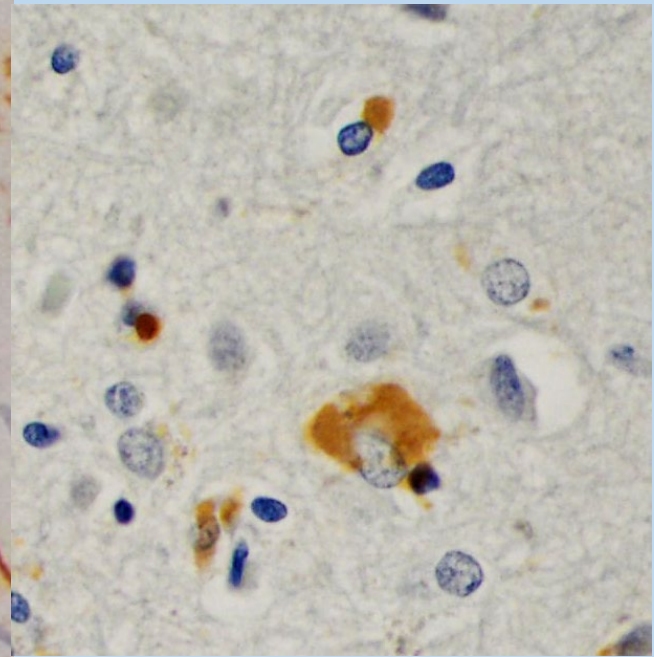




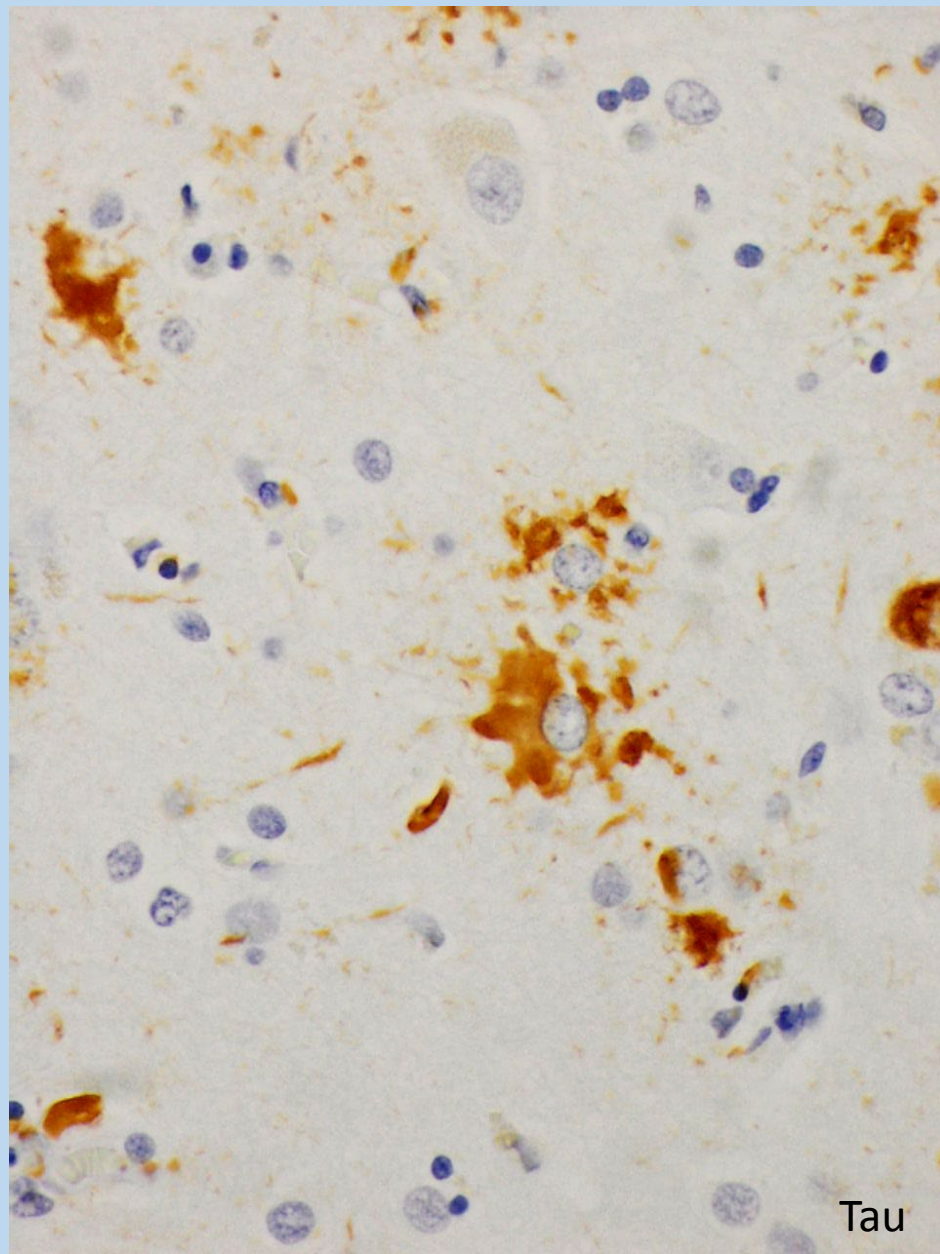
Mid-frontal cortex
deep layers

← Tau – 7F2

Tau – 4R (81A11)

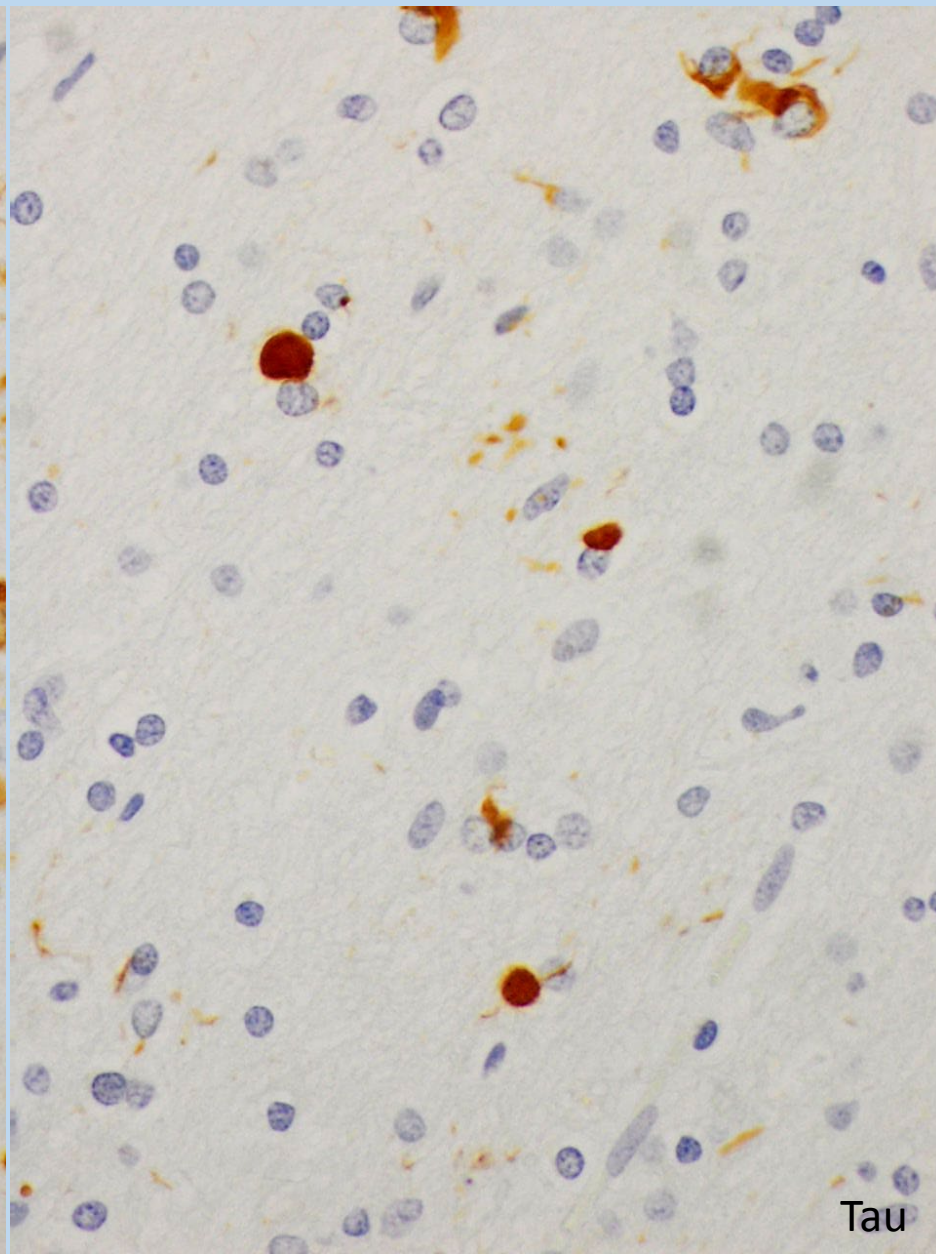


GAI



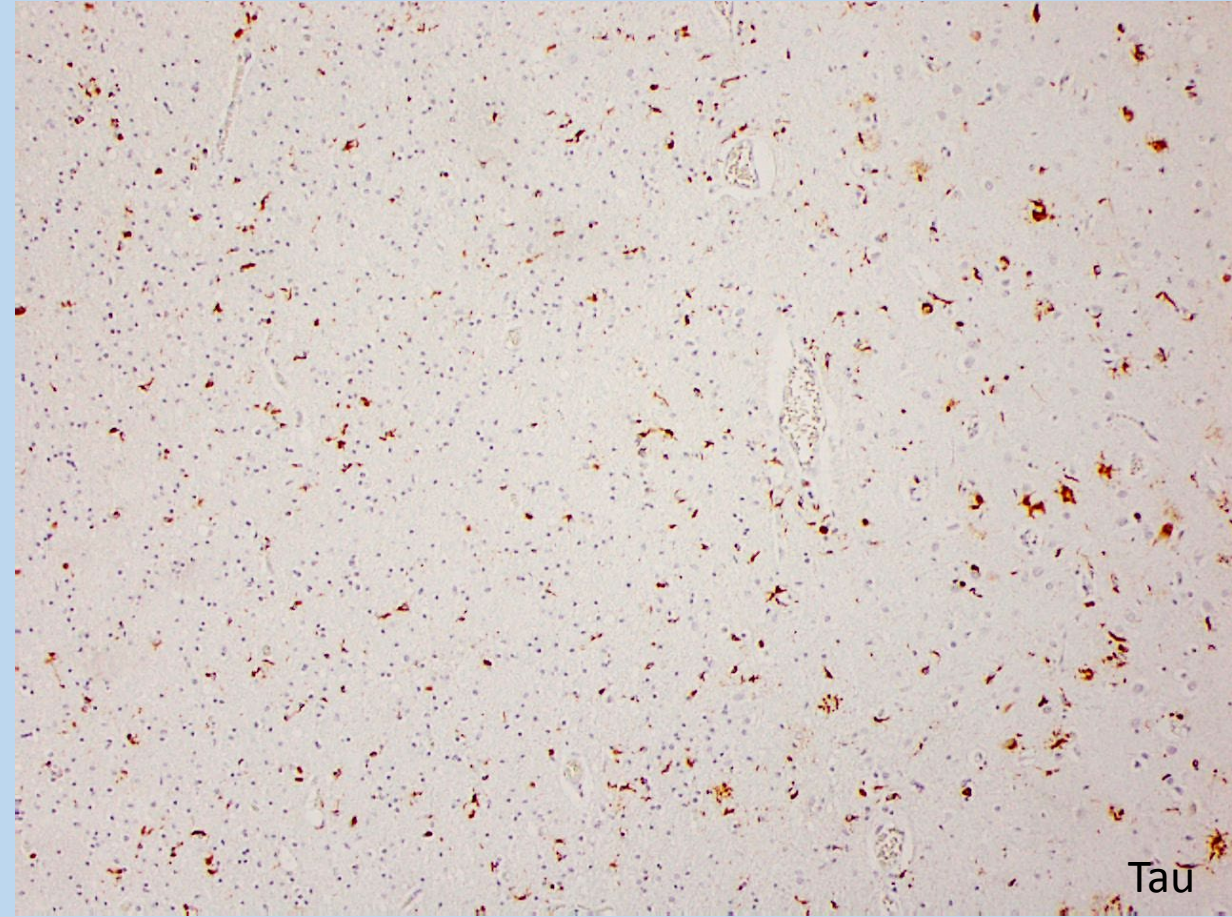
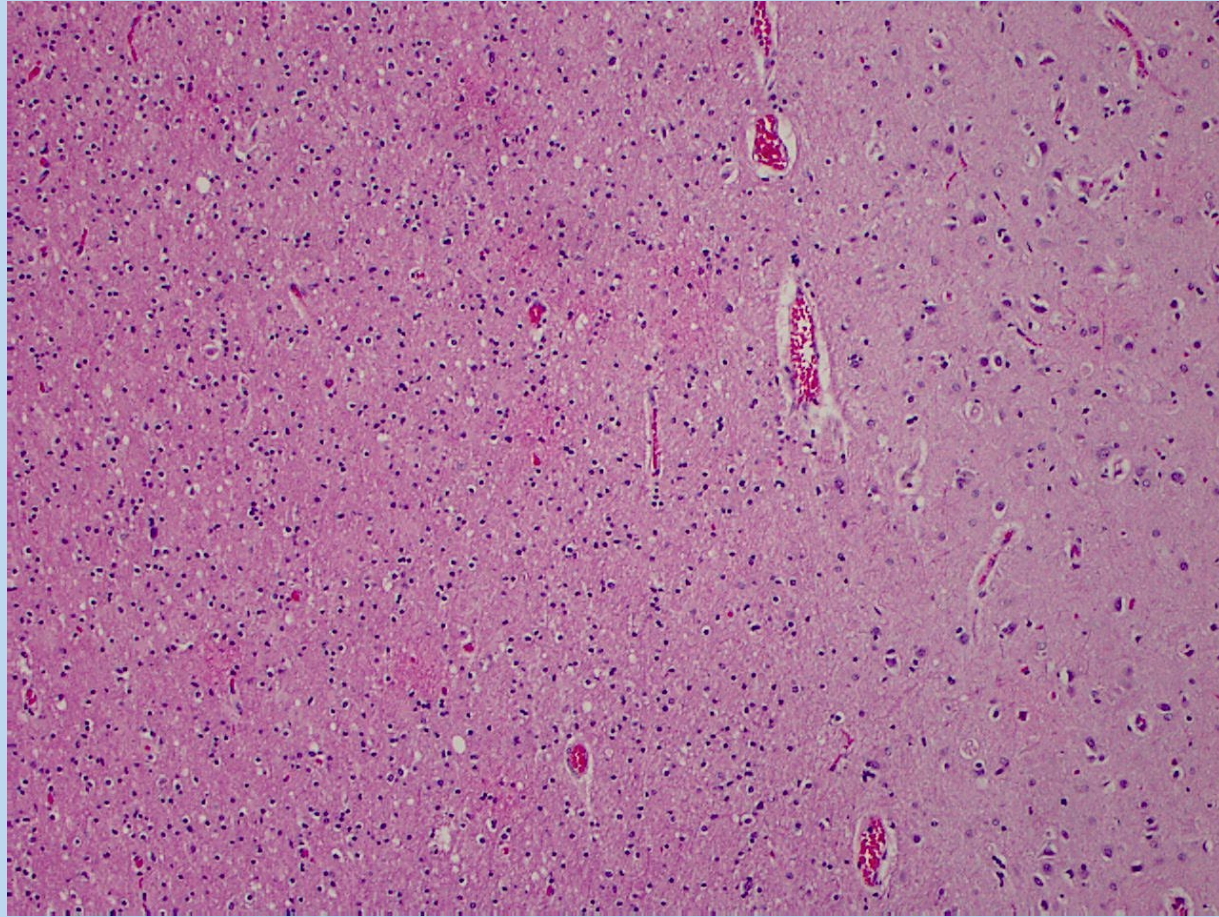
Tau

GOI

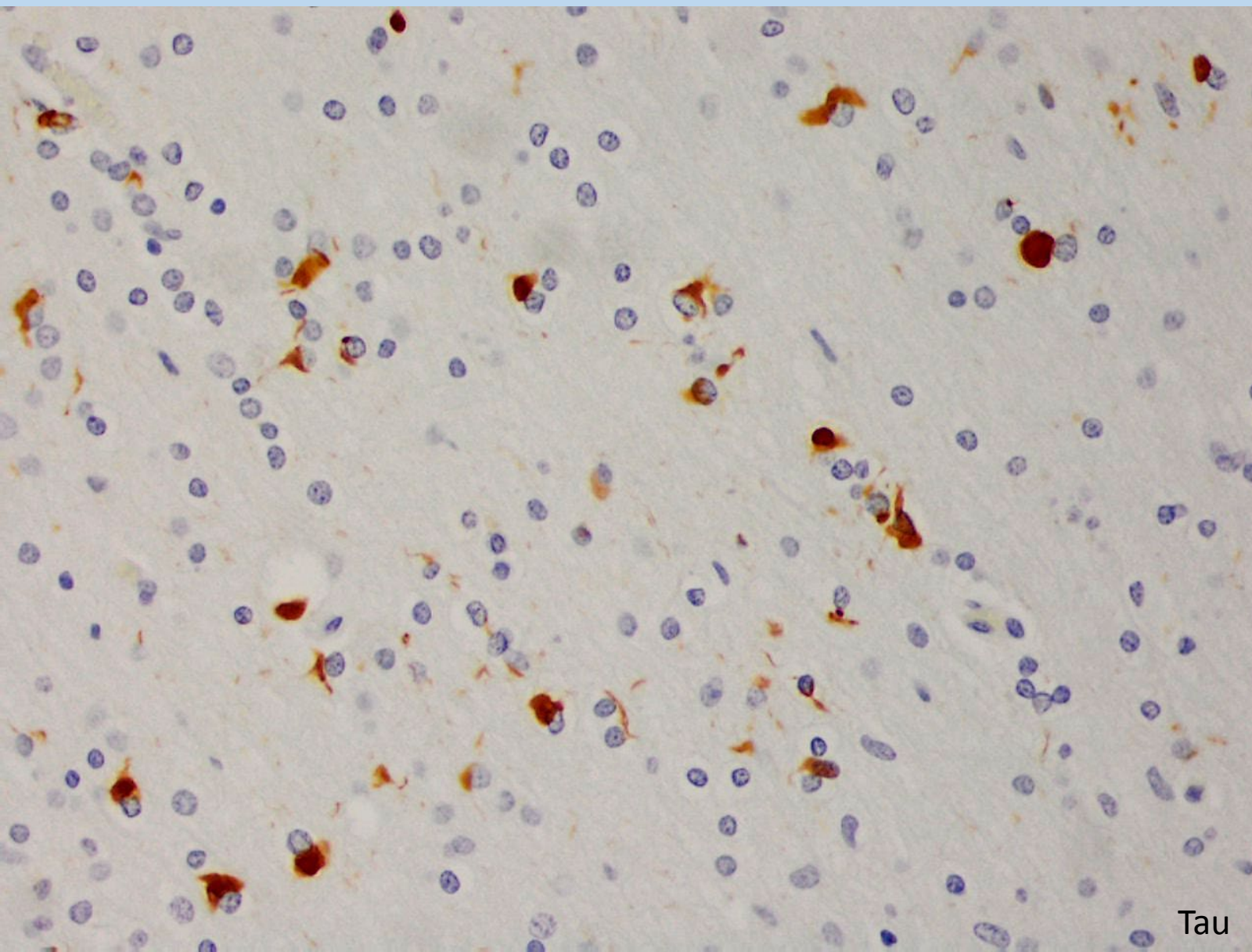


Tau

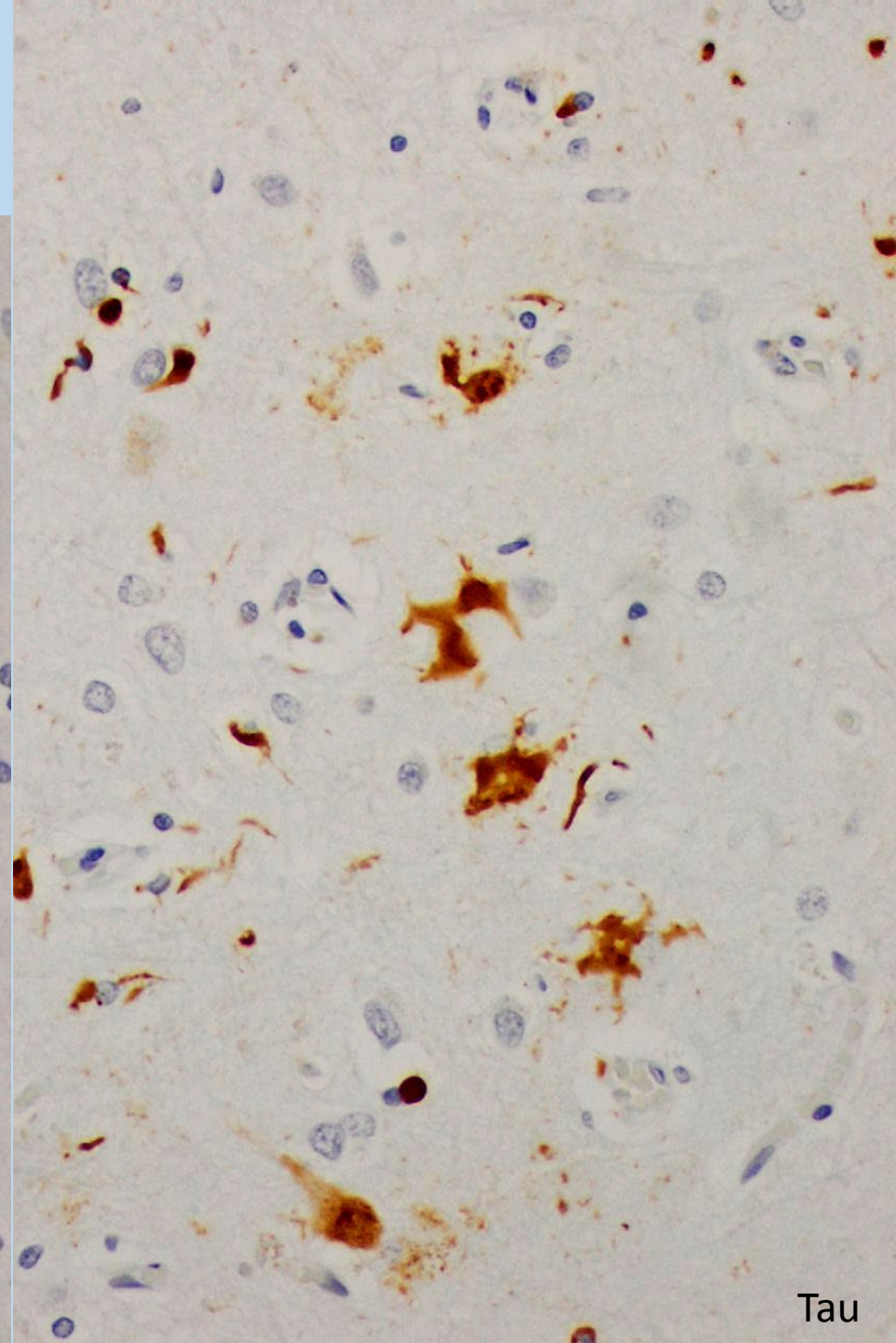
Motor cortex – gray-white matter junction



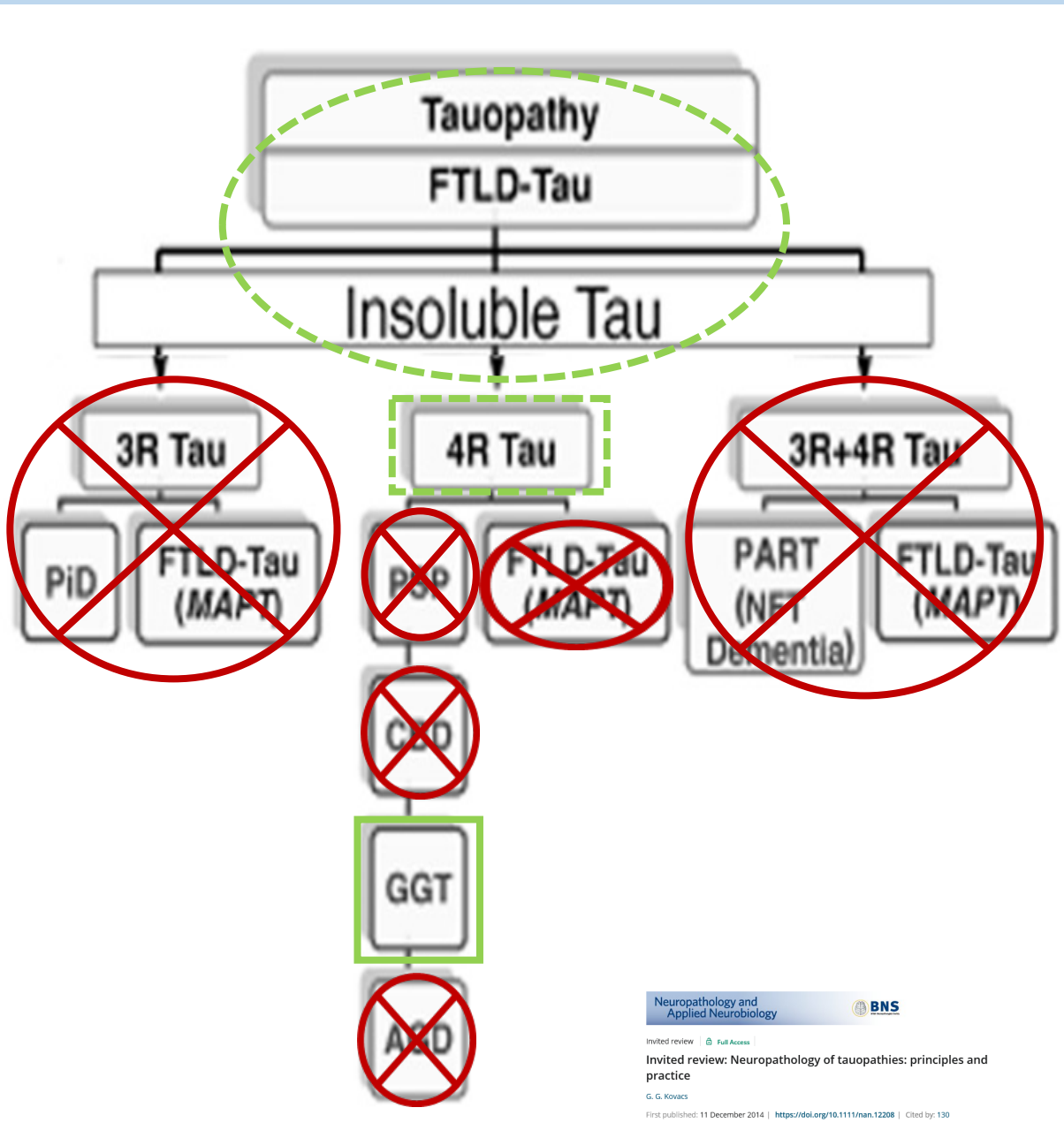
GOI – white matter



Tau



Tau



Differential:

- Sparse A β diffuse plaques and rare neuritic plaques
- No globose neurofibrillary tangles
- No balloon neurons
- No agyrophilic grains or ambient gyrus atrophy
- No known pathogenic MAPT mutation was identified
 - MAPT gene exons 1-5,7,9-13 sequenced
 - (Thanks to Dr. Crary)

Globular glial tauopathies (GGT): consensus recommendations

Zeshan Ahmed · Eileen H. Bigio · Herbert Budka · Dennis W. Dickson · Isidro Ferrer · Bernardino Ghetti · Giorgio Giaccone · Kimmo J. Hatanpaa · Janice L. Holton · Keith A. Josephs · James Powers · Salvatore Spina · Hitoshi Takahashi · Charles L. White III · Tamas Revesz · Gabor G. Kovacs

Subtyping GGT based on globular glial inclusions:

- Type I: Frontotemporal
- Type II: Motor cortex and/or corticospinal tracts
- Type III: Frontotemporal, motor cortex, and/or corticospinal tracts

Globular astrocytic inclusions tend to be more prominent in Type III GGT

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Frontal Lobe Dementia With Novel Tauopathy: Sporadic Multiple System Tauopathy With Dementia

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MATT BAKER, BSC, PARIMALA NACHARAJU, PHD, CHARLES L. WHITE III, MD, PETER DAVIES, PHD,
WENLANG LIN, PHD, AND DENNIS W. DICKSON, MD

Sporadic four-repeat tauopathy with frontotemporal lobar degeneration, Parkinsonism, and motor neuron disease: a distinct clinicopathological and biochemical disease entity

Yong-Juan Fu · Yasushi Nishihira · Shigetoshi Kuroda · Yasuko Toyoshima · Tomohiko Ishihara · Makoto Shinozaki · Akinori Miyashita · Yue-Shan Piao · Chun-Feng Tan · Takashi Tani · Ryoko Koike · Keisuke Iwanaga · Mitsuhiro Tsujihata · Osamu Onodera · Ryozo Kuwano · Masatoyo Nishizawa · Akiyoshi Kakita · Takeshi Ikeuchi · Hitoshi Takahashi

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- Another early paper describing GGT
- Discusses Parkinsonian symptoms; however, all cases in the series were negative for α -synuclein/Lewy bodies

Summary

- Diagnosis:
 - **Lewy body disease, neocortical type**
 - **Globular glial tauopathy**
- 4R tau mutation
- Spectrum of neurodegenerative diseases
- Combination of neurodegenerative features with minimal-to-no published reports

Special Thanks

- Dr. Dennis Dickson for reviewing this case