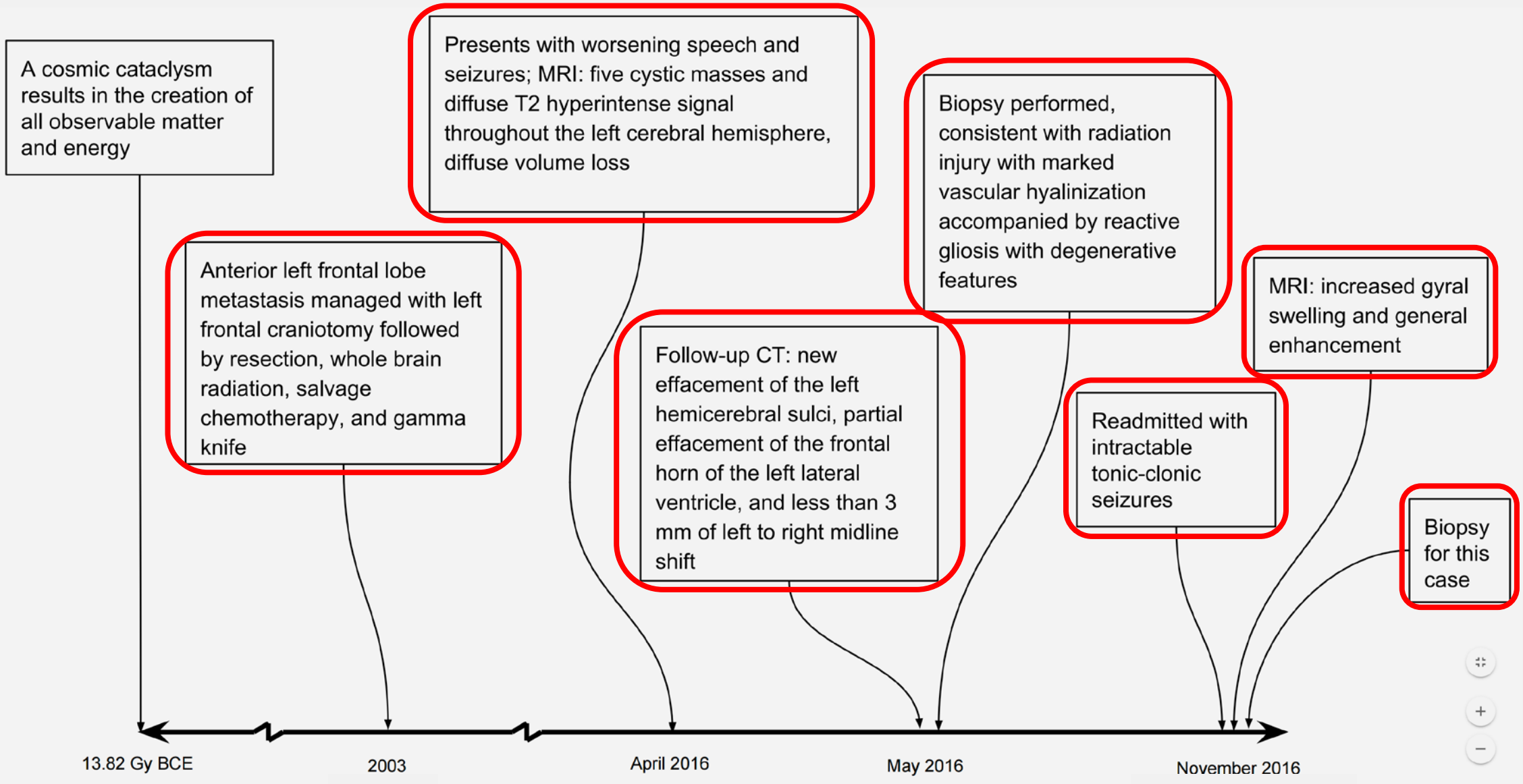
CASE 2017-2

Williamson D, Badve C, Hdeib A, Rogers L, Couce M, Cohen ML

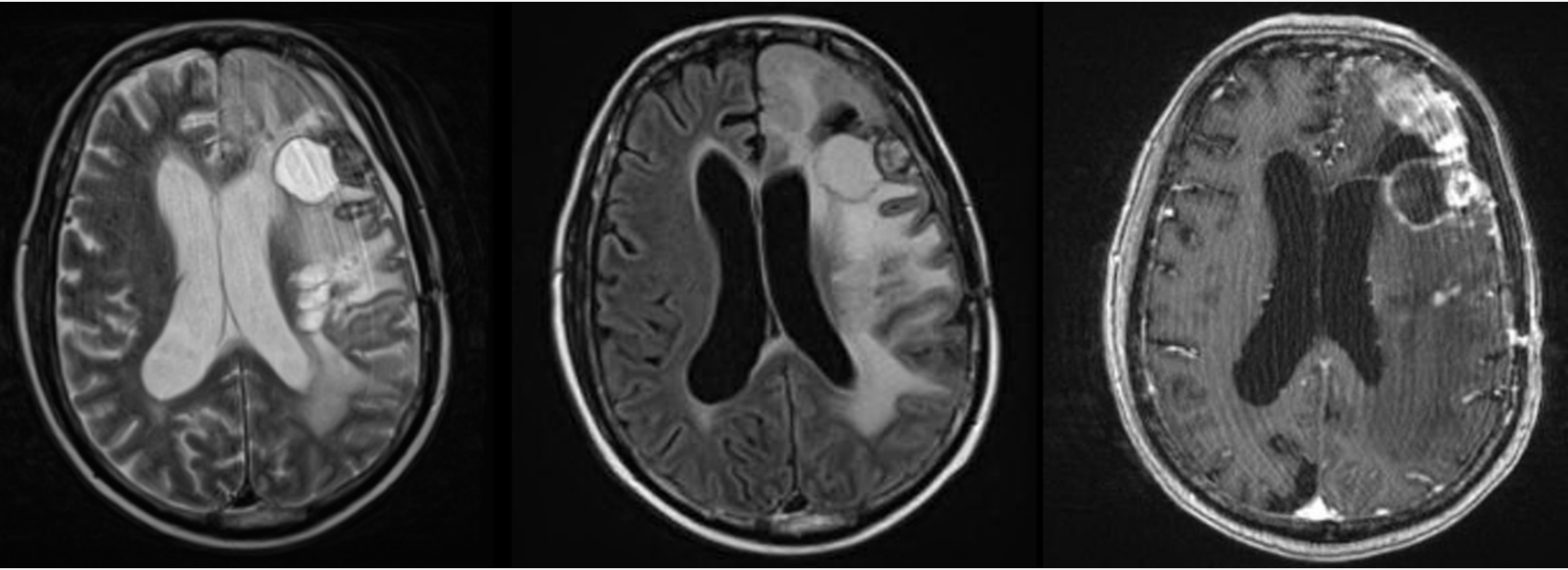
Disclosures

The presenter has no financial relationships to disclose.

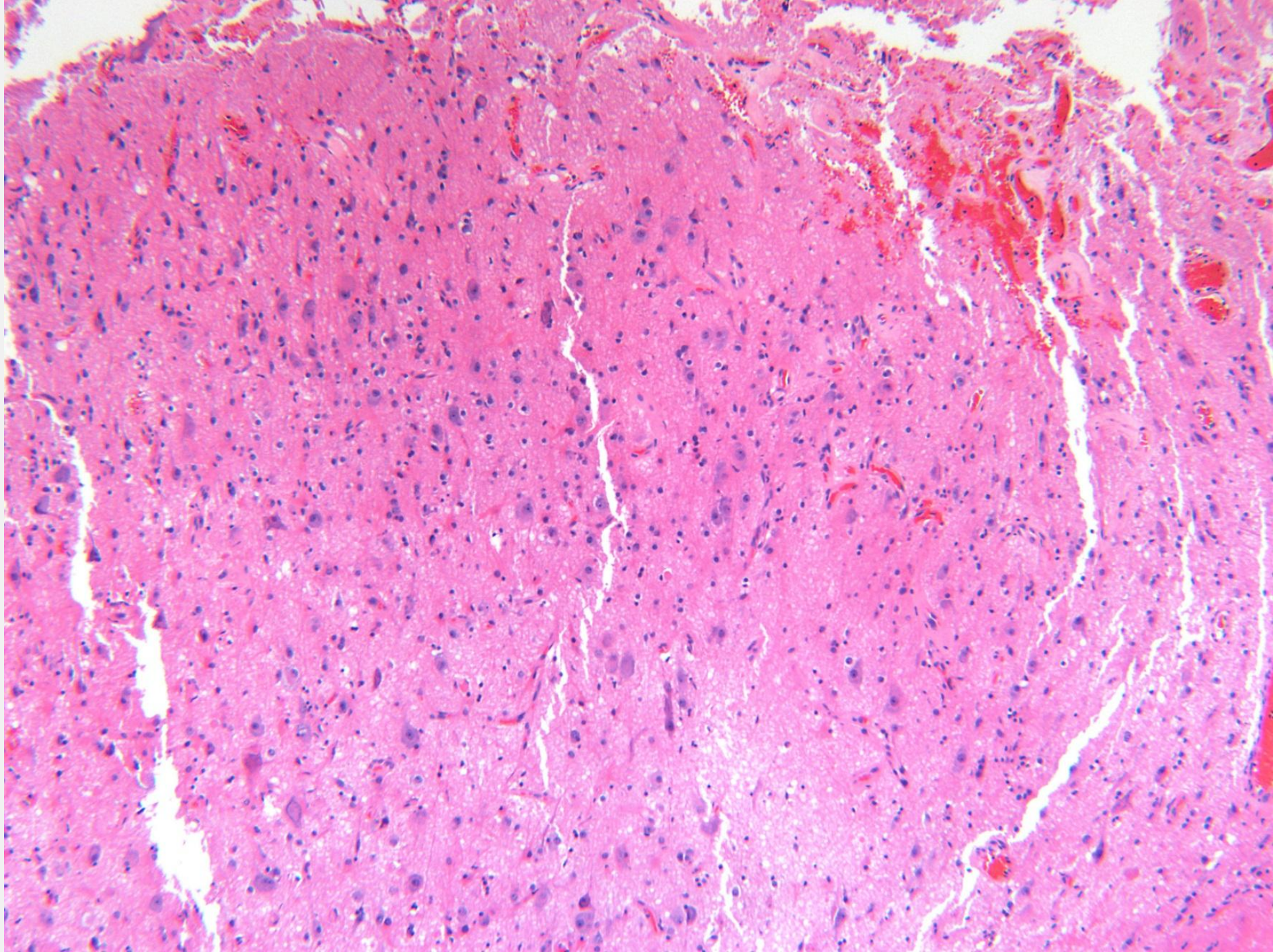
Timeline

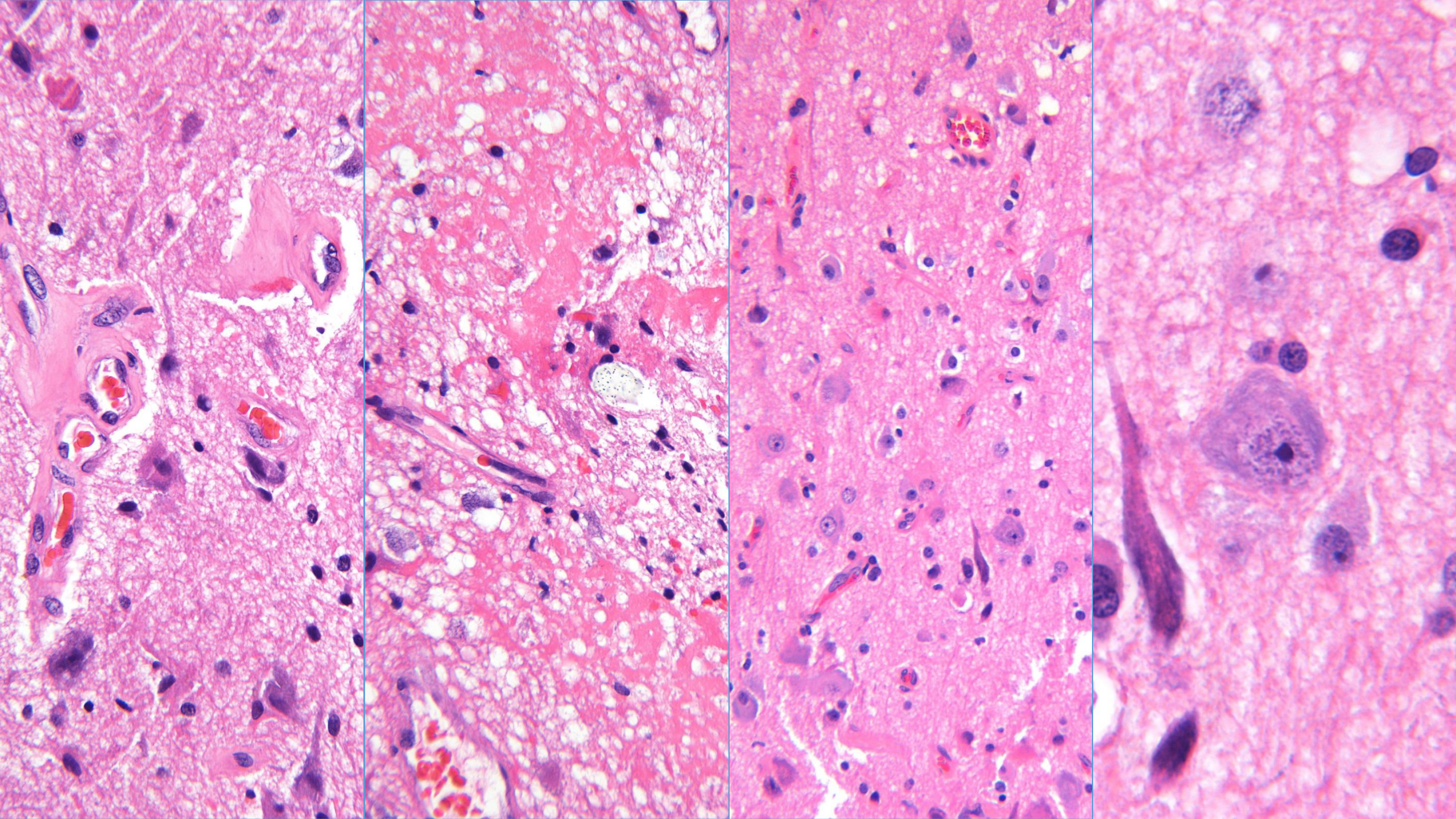

A cosmic cataclysm results in the creation of all observable matter and energy
Anterior left frontal lobe metastasis managed with left frontal craniotomy followed by resection, whole brain radiation, salvage chemotherapy, and gamma knife
Presents with worsening speech and seizures; MRI: five cystic masses and diffuse T2 hyperintense signal throughout the left cerebral hemisphere, diffuse volume loss
Follow-up CT: new effacement of the left hemicerebral sulci, partial effacement of the frontal horn of the left lateral ventricle, and less than 3 mm of left to right midline shift
Biopsy performed, consistent with radiation injury with marked vascular hyalinization accompanied by reactive gliosis with degenerative features
Readmitted with intractable tonic-clonic seizures
MRI: increased gyral swelling and general enhancement
Biopsy for this case
13.82 Gy BCE
2003
April 2016
May 2016
November 2016
Age: 36
Age: 50

Imaging



H&E section from November 2016 biopsy

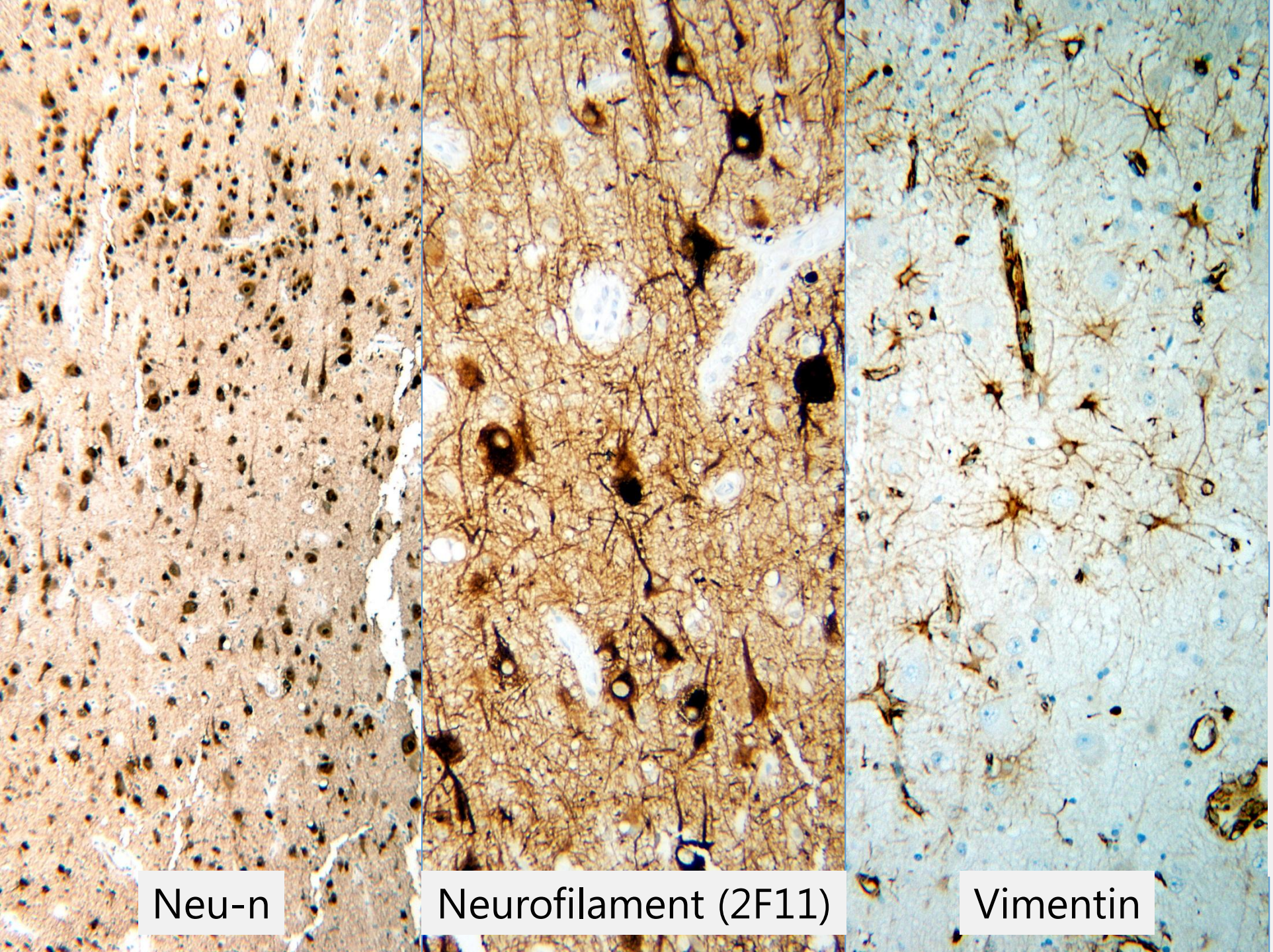




1. Provide 3 possible diagnoses in order of probability.

2. Postulate 2 possible pathogenic mechanisms responsible for the cytological abnormalities seen within the lesion.

3. Name 2 eminent California neuropathologists on whose shoulders we are now standing.

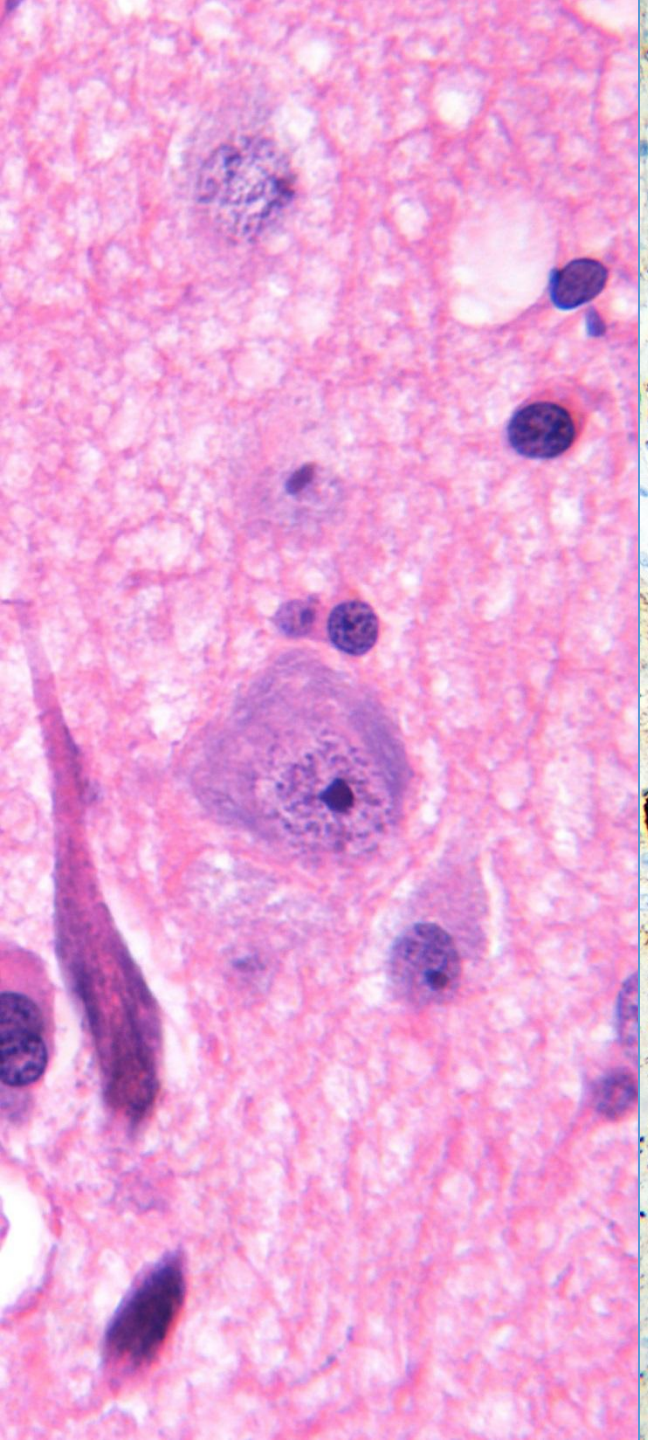


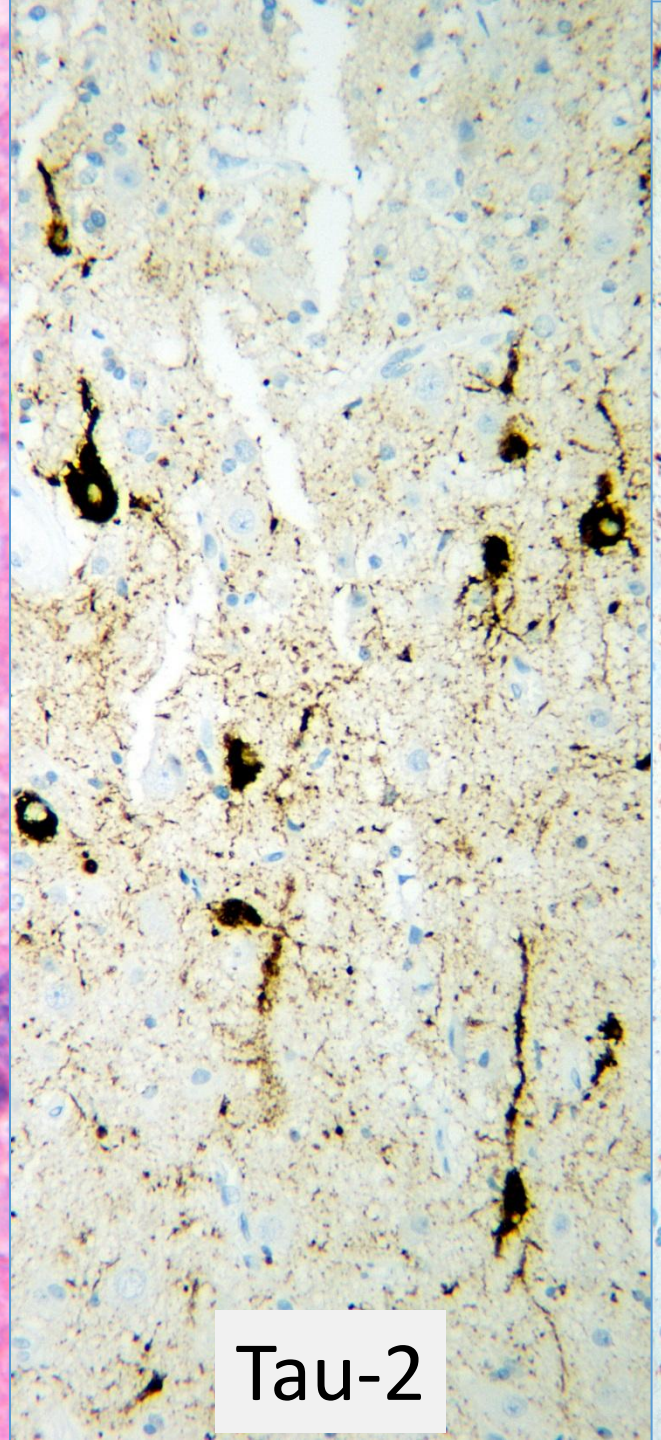
Provide 3 possible diagnoses in order of probability.

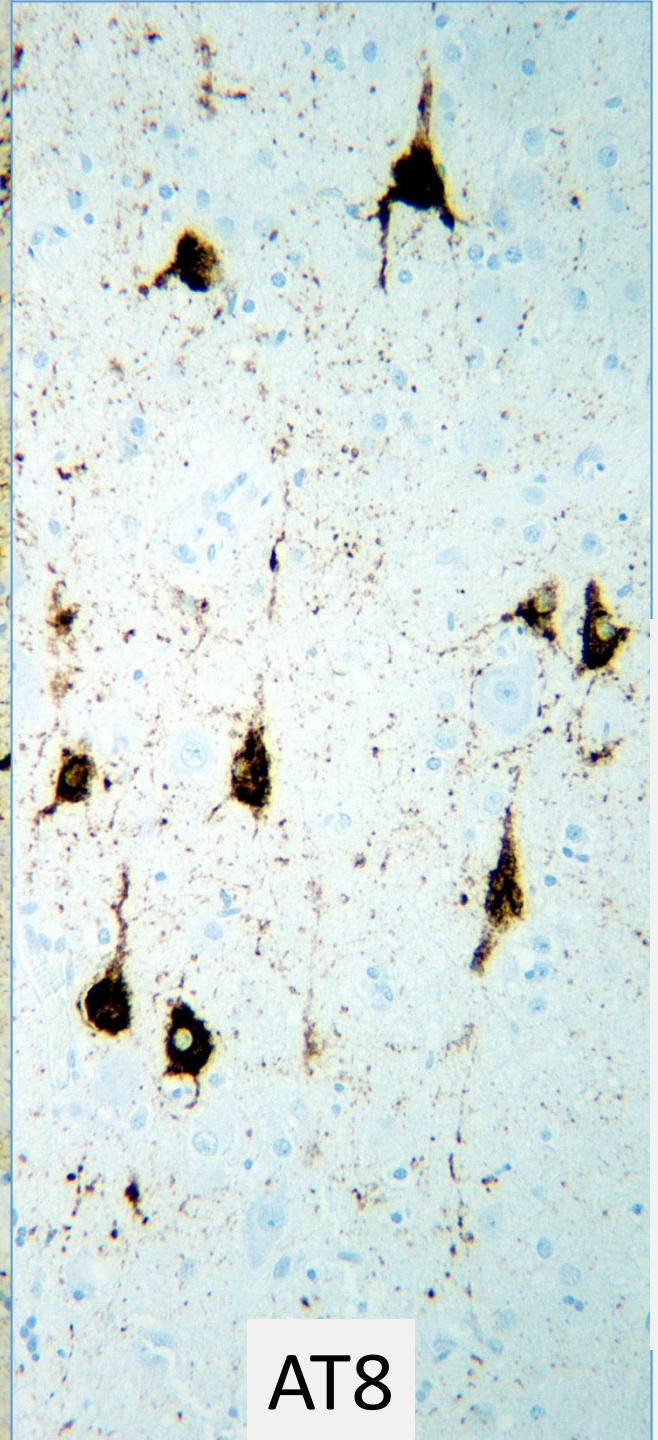
1. Glioneuronal tumor (?radiation-induced)

2. Cortical dysplasia (?radiation-induced)

3. Recurrent or residual germ cell tumor (mature glioneuronal component of teratoma only)







Additional Immunostaining

p62 – positive
Ki-67 – rare glial nuclei
β-amyloid – negative
α-synuclein – negative
TDP-43 – negative

Diagnosis

CORTICAL DYSPLASIA WITH NEUROFIBRILLARY DEGENERATION (13 years after radiotherapy for metastatic germ cell tumor)

Postulate 2 possible pathogenetic mechanisms responsible for the cytological abnormalities seen within the lesion.

doi:10.1093/brain/aww187 BRAIN 2016: 139; 2441–2455 | 2441

BRAIN
A JOURNAL OF NEUROLOGY

Hyperphosphorylated tau in patients with refractory epilepsy correlates with cognitive decline: a study of temporal lobe resections

Xin You Tai,[1,2] Matthias Koepp,[2] John S. Duncan,[2] Nick Fox,[3] Pamela Thompson,[2] Sallie Baxendale,[2] Joan Y. W. Liu,[1] Cheryl Reeves,[1] Zuzanna Michalak[1] and Maria Thom[1]

Cranial irradiation compromises neuronal architecture in the hippocampus

PNAS, July 2013

Vipan Kumar Parihar and Charles L. Limoli[1]

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ORIGINAL ARTICLE

Ionizing radiation causes increased tau phosphorylation in primary neurons

Li Li,*'† Wenzhang Wang,* Scott Welford,‡ Teng Zhang,† Xinglong Wang* and Xiongwei Zhu*

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†Yueyang Hospital and Clinical Research Institute of Integrative Medicine, Shanghai University of Traditional Chinese Medicine, Shanghai, China
‡Department of Oncology, Case Western Reserve University, Cleveland, Ohio, USA

nature neuroscience

Neuronal activity enhances tau propagation and tau pathology *in vivo*

Jessica W Wu[1], S Abid Hussaini[1,2], Isle M Bastille[1], Gustavo A Rodriguez[1], Ana Mrejeru[3], Kelly Rilett[1], David W Sanders[4], Casey Cook[5], Hongjun Fu[1], Rick A C M Boonen[1], Mathieu Herman[1], Eden Nahmani[1], Sheina Emrani[1], Y Helen Figueroa[1], Marc I Diamond[4], Catherine L Clelland[1], Selina Wray[6] & Karen E Duff[1,2,7]

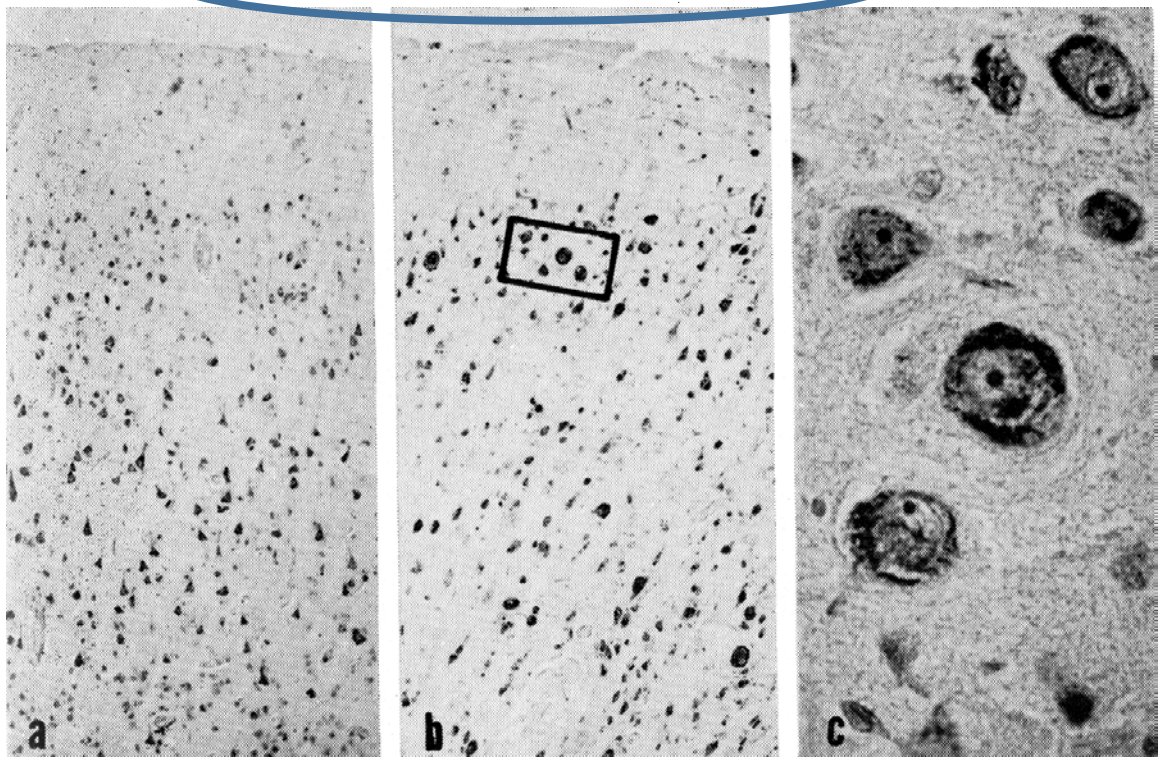
Name 2 eminent California neuropathologists on whose shoulders we are now standing.

Delayed effects of radiation on the human central nervous system

"Early" and "late" delayed reactions

Neurology, 1964

Peter W. Lampert, M.D., and
Lt. Cmdr. Richard L. Davis, M.C., U.S.N.

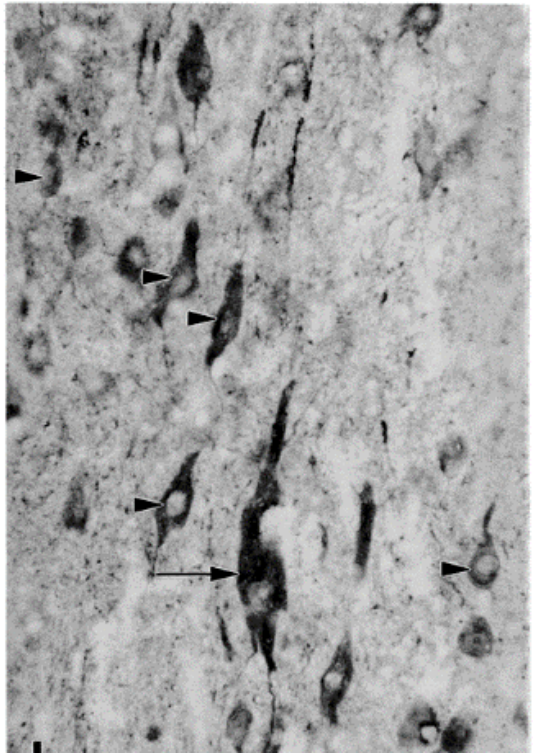


Acta Neuropathol (1994) 87 : 493–503

REGULAR PAPER

T. Duong · M.J. De Rosa · V. Poukens · H.V. Vinters
R.S. Fisher

Neuronal cytoskeletal abnormalities in human cerebral cortical dysplasia



Case	Age at Initial Diagnosis	Original Diagnosis	Interval (years)
Lampert, 1964	19	“Astroblastoma”	12
Caccamo, 1989	25	Pitutary adenoma	6
Gaughen, 2009	54	Anaplastic oligodendriglioma	7
Shaikh, 2017 (AAN abstract)	25	Glioblastoma	2
DSS 2017-2	36	Metastatic GCT	13

Lampert PW, Davis RL. Delayed Effects of Radiation on the Human Central Nervous System; “Early” and “Late” Delayed Reactions. Neurology. 1964 Oct;14:912-7.

Duong T, De Rosa MJ, Poukens V, Vinters HV, Fisher RS. Neuronal cytoskeletal abnormalities in human cerebral cortical dysplasia. Acta Neuropathol. 1994;87:493-503.

Caccamo D, Herman MM, Urich H, Rubinstein LJ. Focal neuronal gigantism and cerebral cortical thickening after therapeutic irradiation of the central nervous system. Arch Pathol Lab Med. 1989 Aug;113:880-5.

Gaughen JR, Bourne TD, Aregawi D, et al. Focal neuronal gigantism: a rare complication of therapeutic radiation. AJNR Am J Neuroradiol. 2009 Nov;30:1933-5. (AKA Diagnostic Slide Session Case 2009–5)

Tai XY, Koepp M, Duncan JS, et al. Hyperphosphorylated tau in patients with refractory epilepsy correlates with cognitive decline: a study of temporal lobe resections. Brain. 2016 Sep;139:2441-55.

Wu JW, Hussaini SA, Bastille IM, et al. Neuronal activity enhances tau propagation and tau pathology in vivo. Nat Neurosci. 2016 Aug;19:1085-92.

Parihar VK, Limoli CL. Cranial irradiation compromises neuronal architecture in the hippocampus. Proc Natl Acad Sci U S A. 2013 Jul 30;110:12822-7.

Li L, Wang W, Welford S, et al. Ionizing radiation causes increased tau phosphorylation in primary neurons. J Neurochem. 2014 Oct;131:86-93.