RUSH UNIVERSITY MEDICAL CENTER

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X. Robert Zhang, MD., PhD., Lubov Romantseva, MD.

(Rush University Medical Center)

Harry V. Vinters, MD.

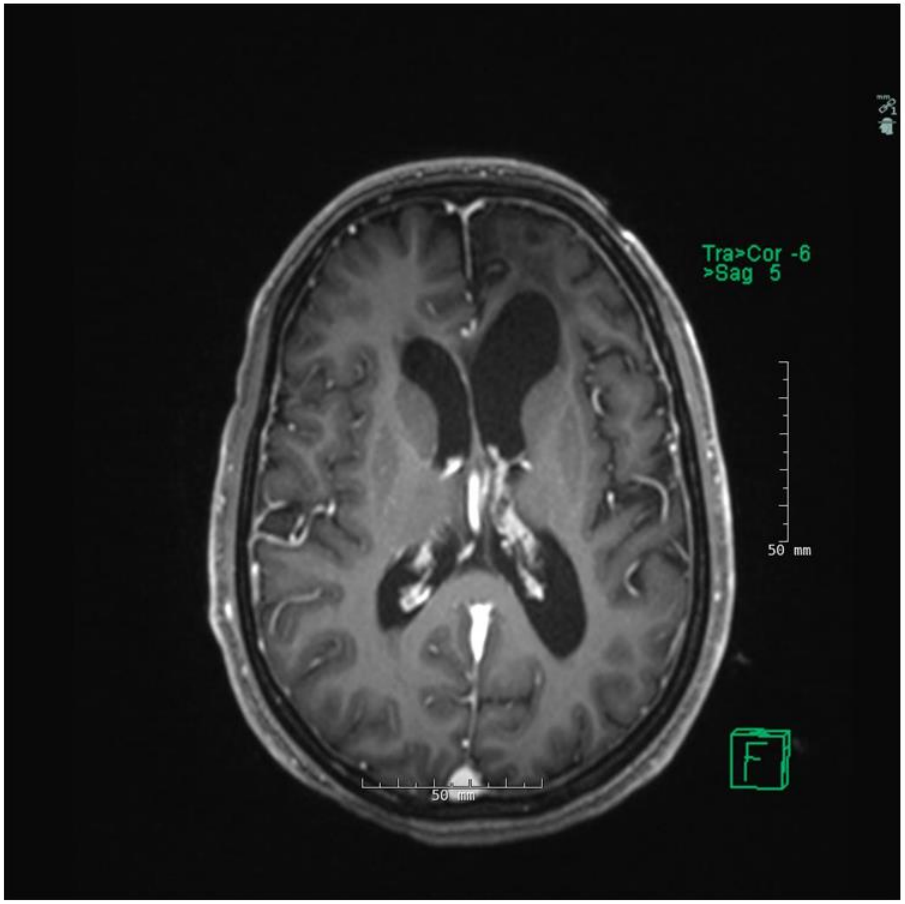
(UCLA Medical Center)

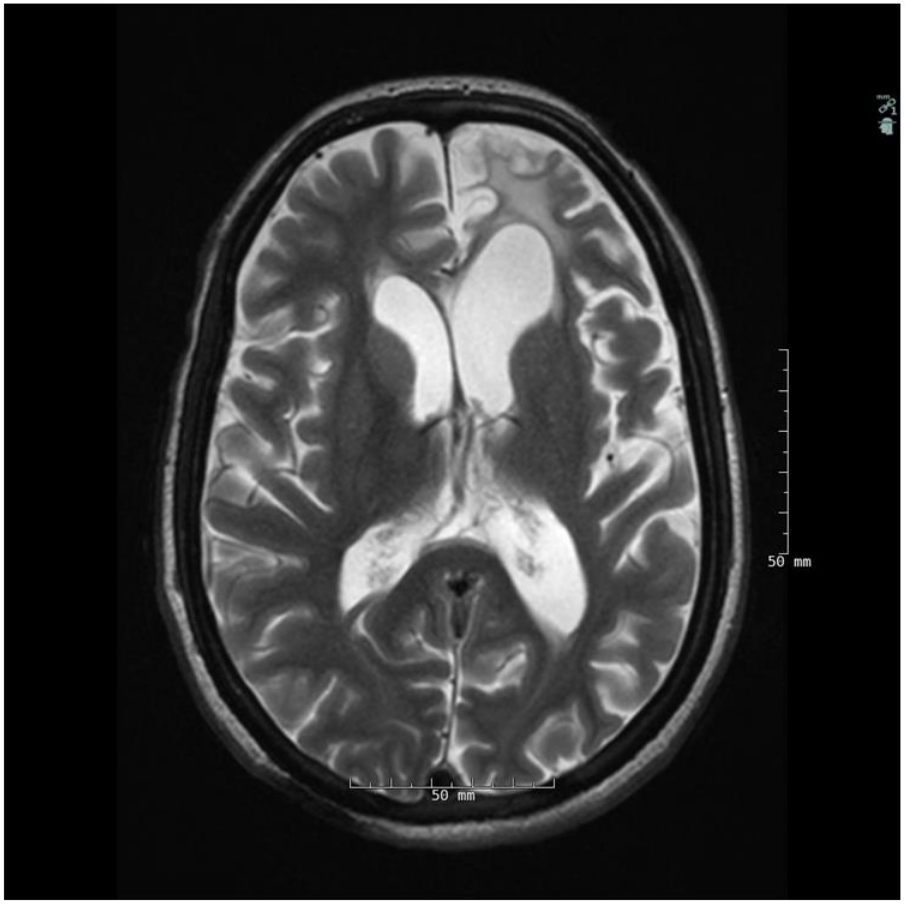
No disclosures or conflicts of interest

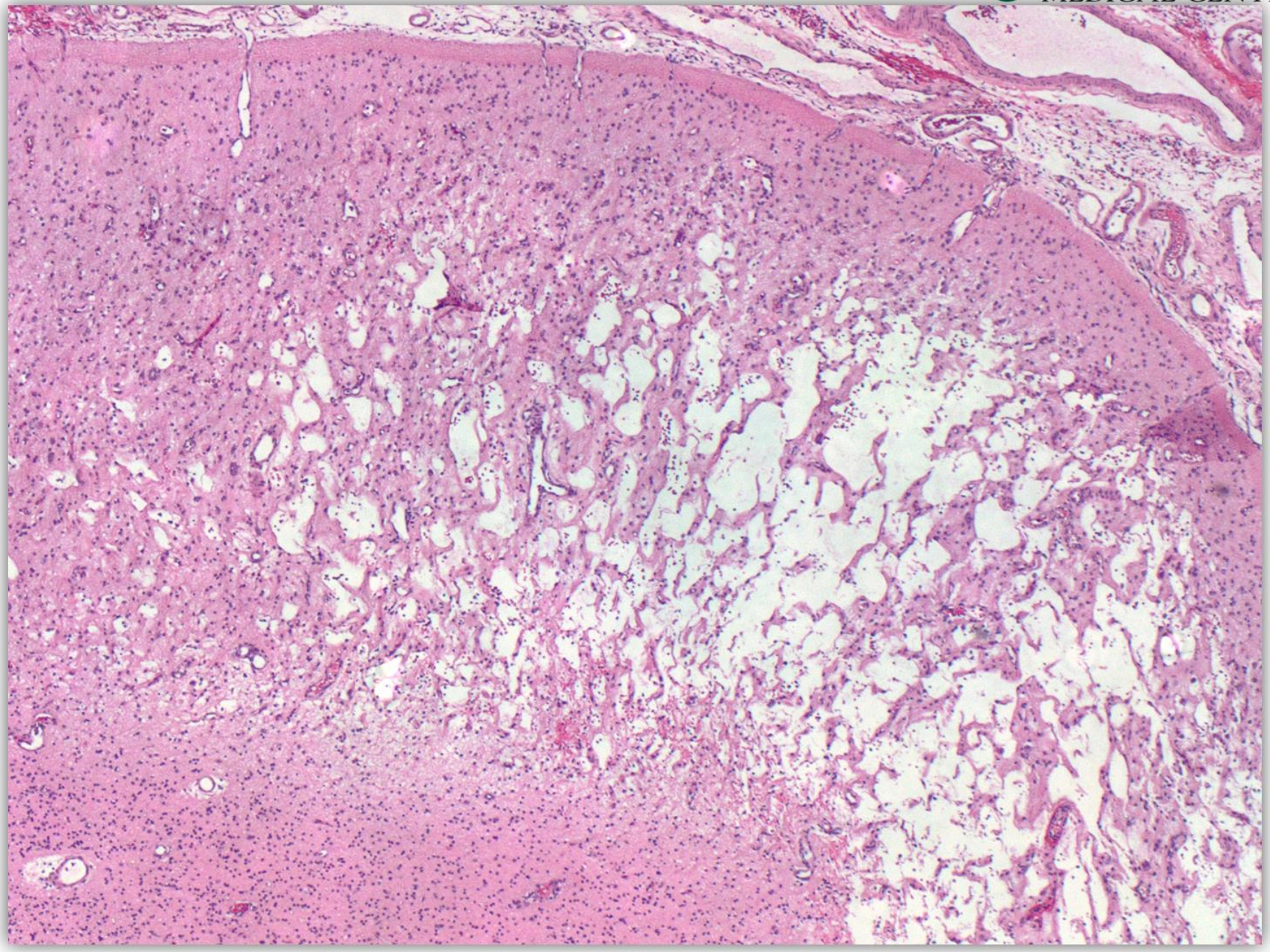
Clinical Summary

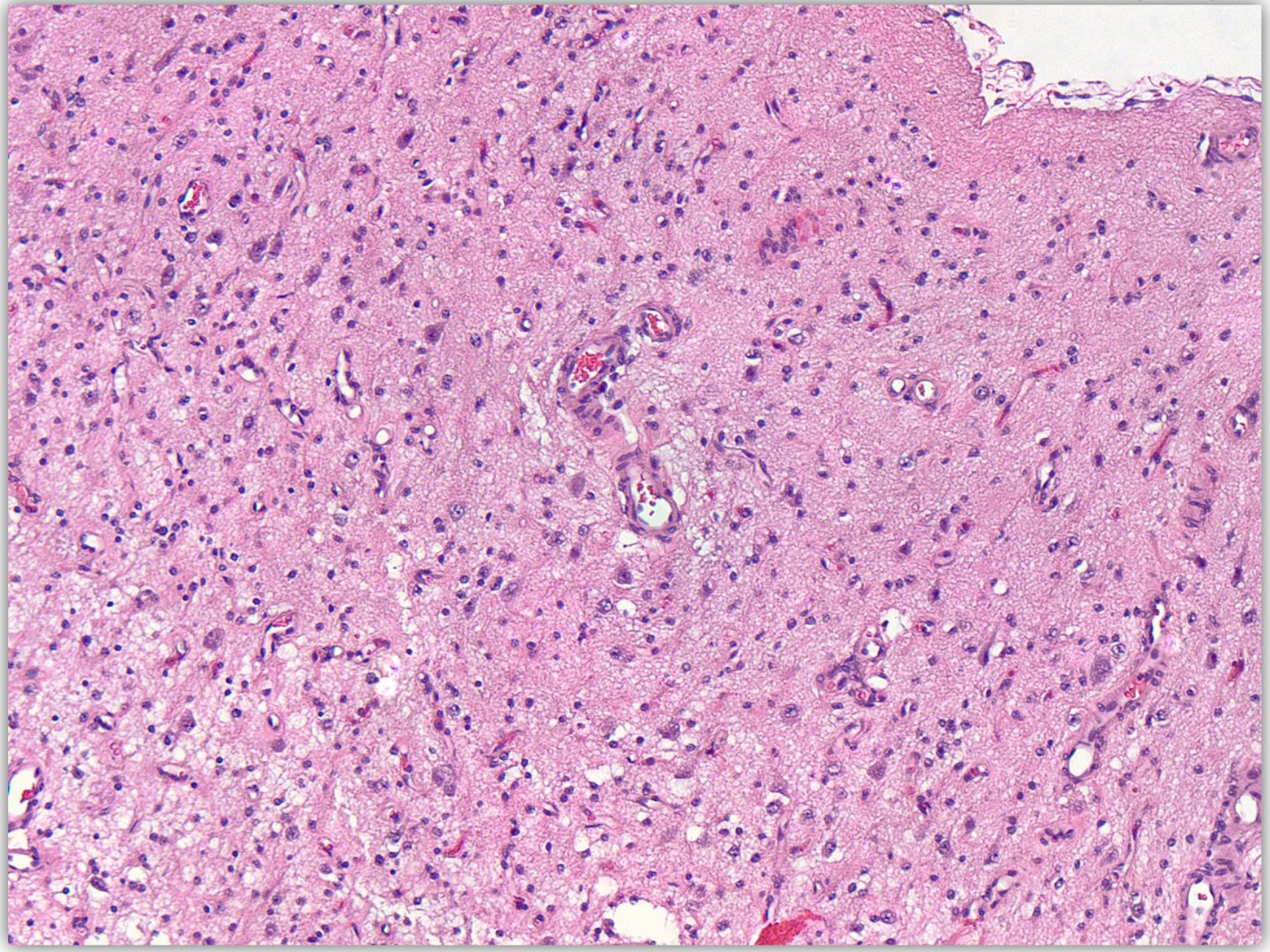
- 13-year-old girl with intractable localization-related epilepsy since the age of five
- Born full term with normal early development and milestones
- Developed right hemiparesis and developmental delays after seizures had started
- Significant work-up with allergy and immunology, rheumatology, neurology and infectious disease with unrevealing results
- EEG showed multifocal seizures, left fronto-temporal region and central mesial region
- Underwent left frontal lobectomy and anterior corpus callosotomy
- Post-operatively developed cerebral venous thrombosis and central diabetes insipidus
- Prothrombin G20210A heterozygote mutation
- Mostly seizure free, on 5 anti-seizure medications

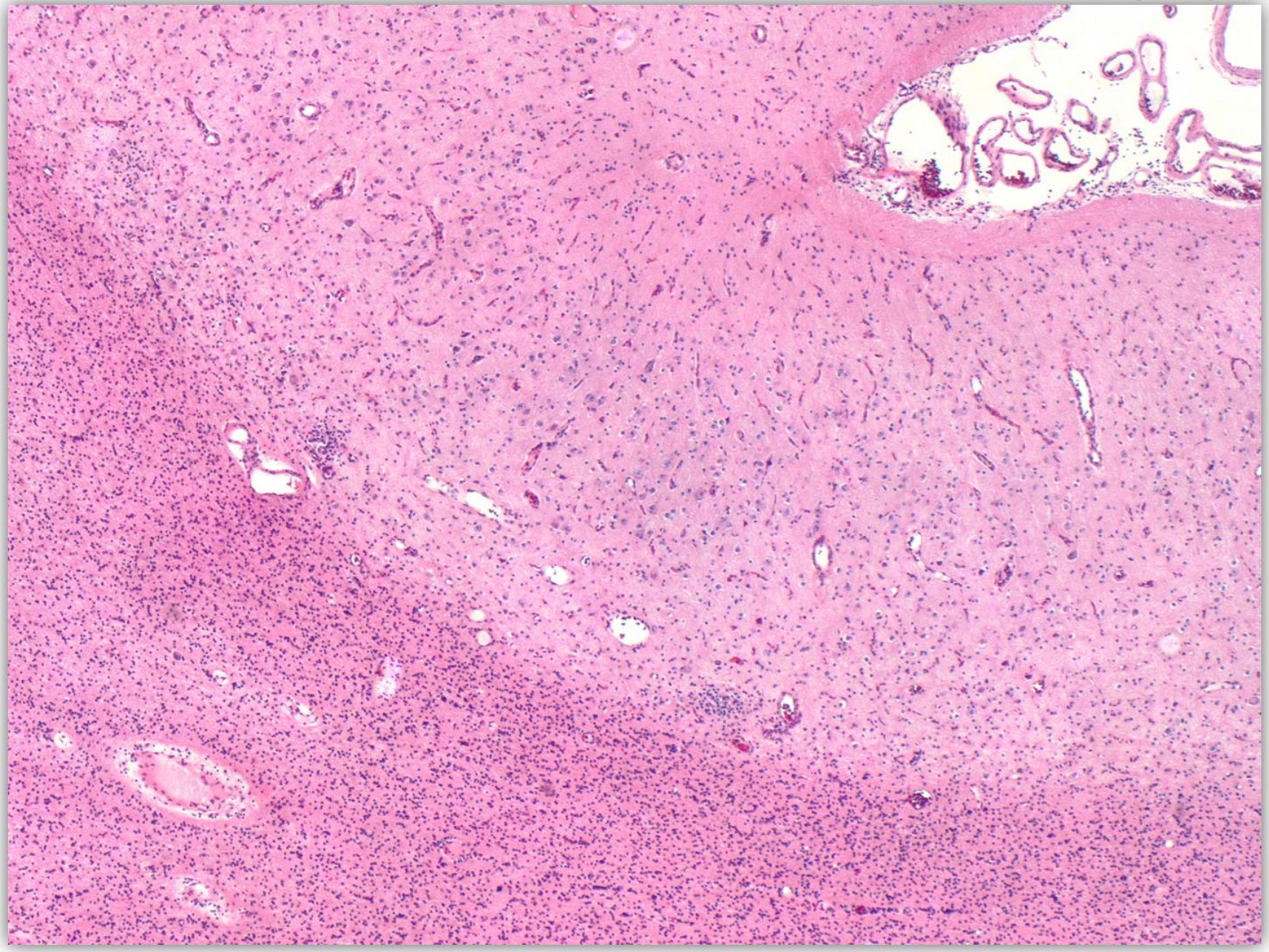
Neuroradiology







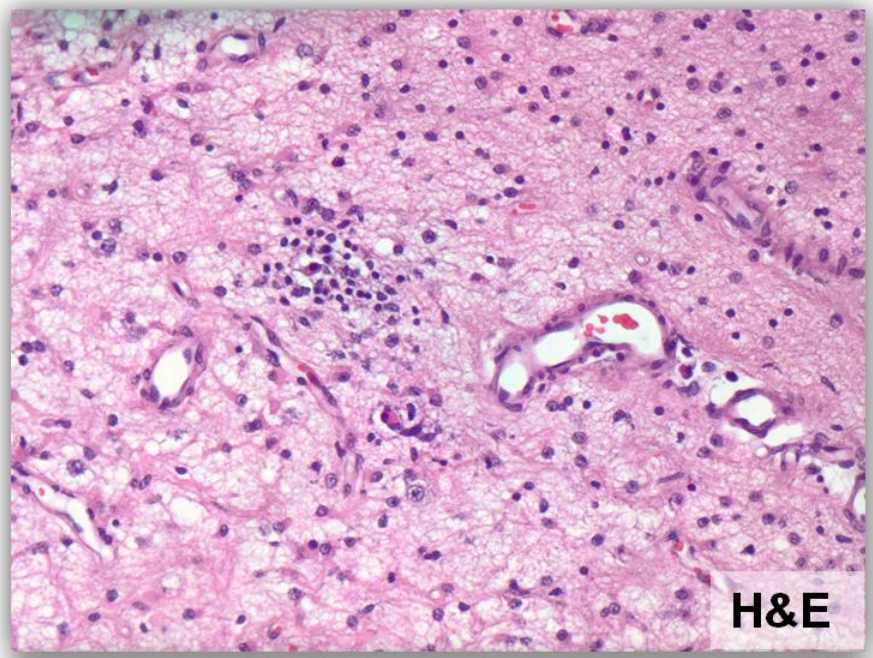




Differential Diagnosis?

Differential Diagnosis

- Cortical laminar necrosis
- Congenital malformation
- Focal cortical dysplasia
- Rasmussen encephalitis
- Viral encephalitis
- Autoimmune encephalitis
- Sturge-Weber syndrome
- Hemimegalencephaly
- Unihemispheric cerebral vasculitis
- Neuroepithelial tumors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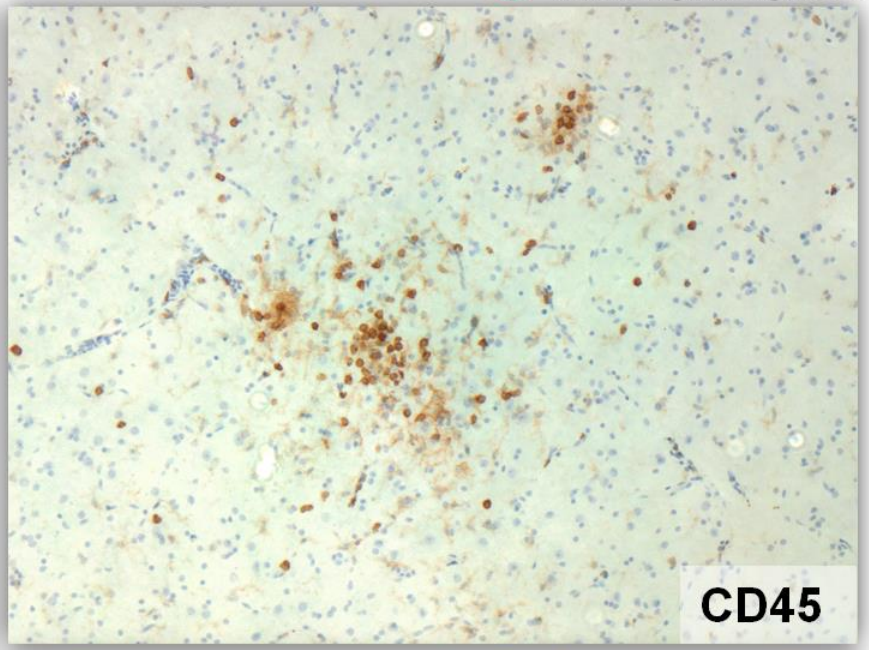
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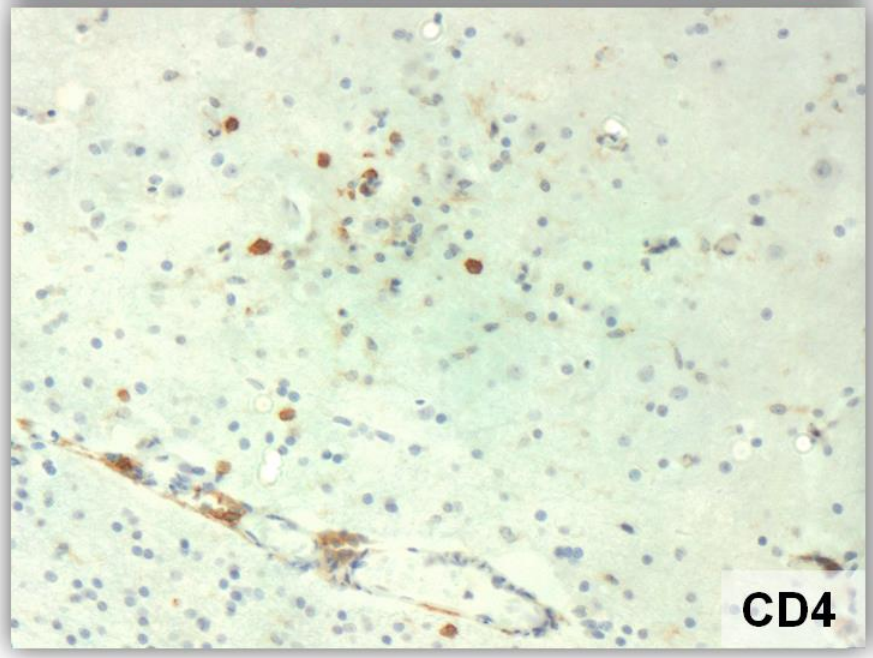
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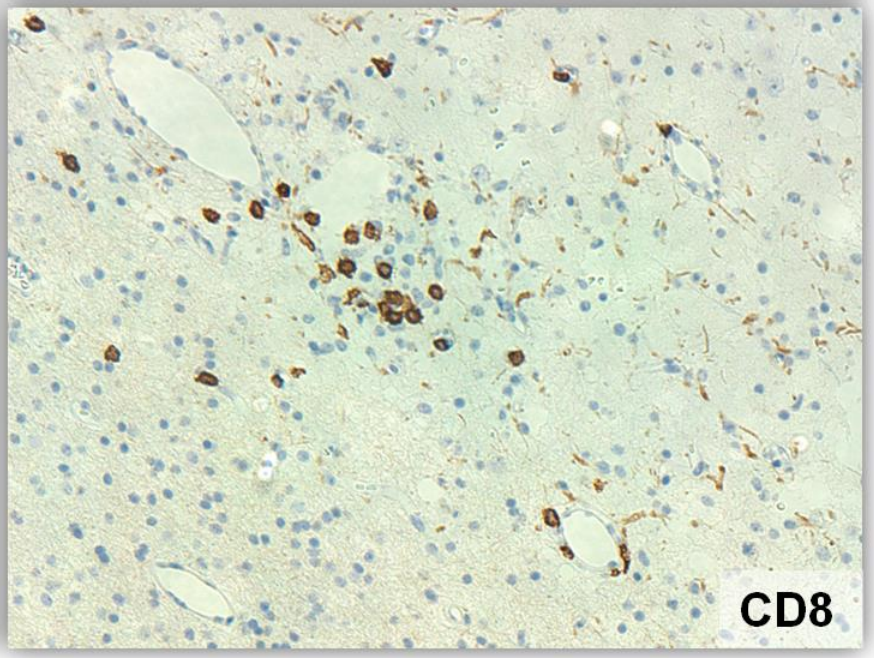
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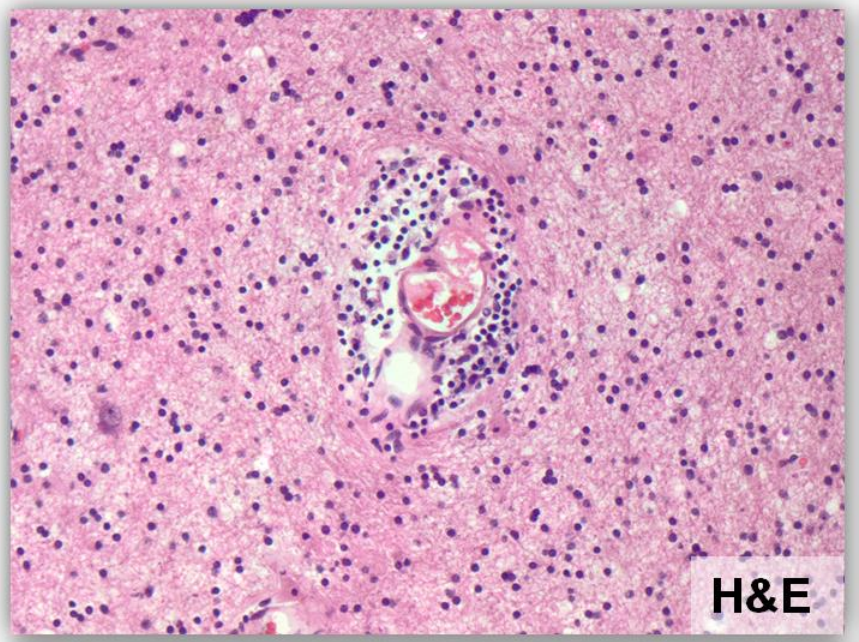
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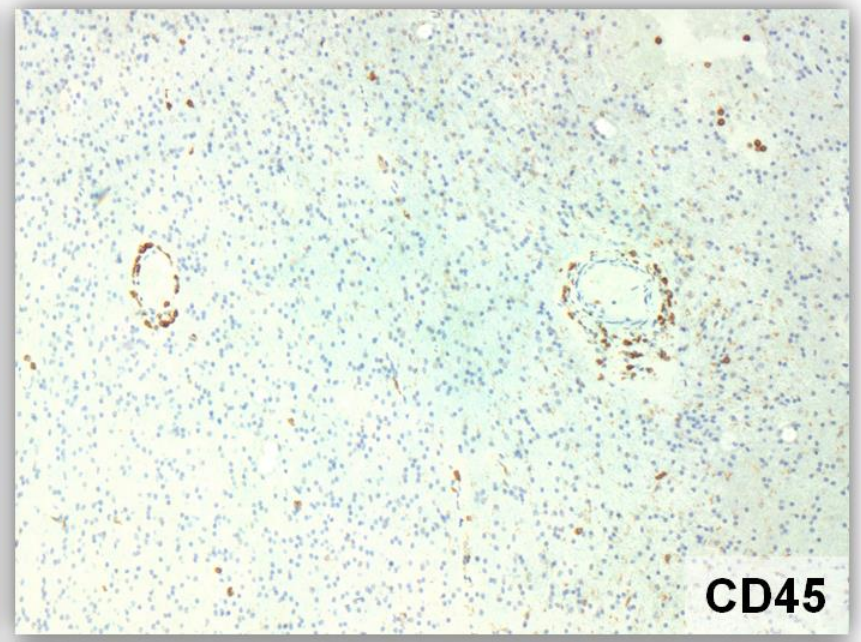
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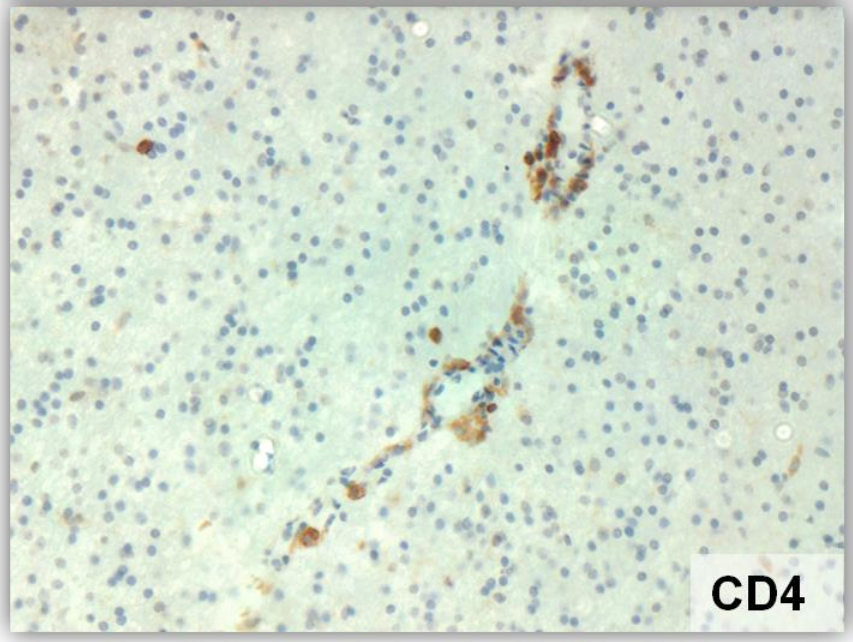
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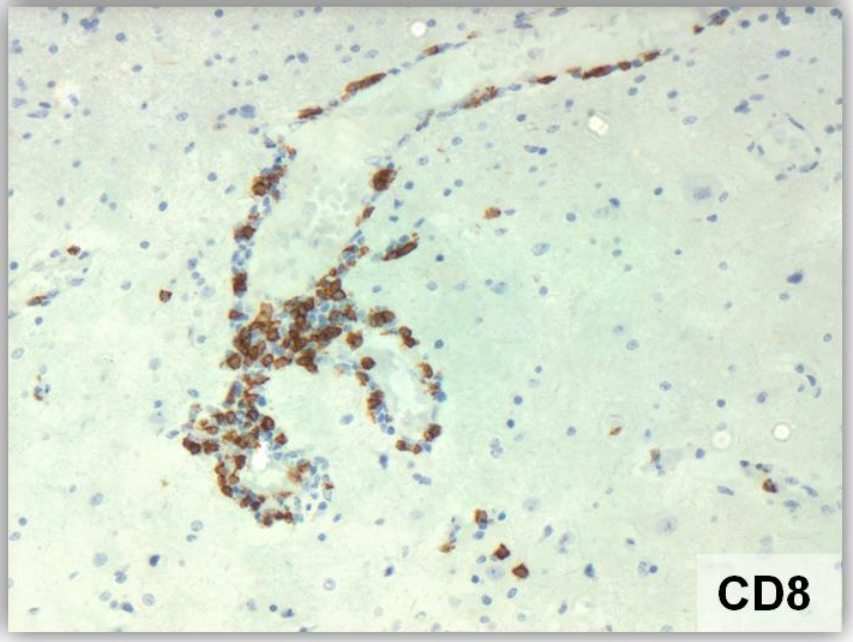
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Neuropathology Diagnosis:

Rasmussen Encephalitis, Stage 4

Rasmussen Encephalitis

- First described by Dr. Rasmussen in 1958
- Age of first seizure as initial presentation: 5-10 years
- Pathogenesis remains unknown:
  - An initial virus infection
  - T-cell response to one or more antigenic epitopes
  - Potential additional contribution by autoantibodies
- Features differentiate from most autoimmune encephalitis:
  - Symptom chronicity
  - Unilateral hemispheric functional and structural involvement
  - Refractoriness to immunotherapy

Rasmussen Encephalitis

- Clinical features:
  - Focal seizures, typically focal (unilateral) motor seizures
  - Epilepsia partialis continua (EPC), 60%
  - Progressive unilateral neurologic deficit (usually hemiparesis)
- Neuroradiologic features:
  - Unihemispheric progressive cerebral atrophy
  - No intracranial calcification or contrast enhancement
- Neuropathologic features:
  - Chronic inflammatory and destructive changes
  - Almost always confined to one cerebral hemisphere
- Treatment :
  - Immunosuppressive therapies: steroids, IVIG
  - Surgery: hemispherectomy, focal corticectomy

Rasmussen Encephalitis

- Neuropathology: four stages in the development of Rasmussen Encephalitis:
  - Stage 1: mainly inflammation, especially presence of perivascular lymphocytes and accumulation of microglial nodules
  - Stage 2: more prominent lymphocytic inflammation, widespread astrogliosis and microgliosis in all cortical layers, patchy neuronal loss
  - Stage 3: severe neocortical degeneration, with a patchy panlaminar pattern and astrocytic gliosis
  - Stage 4: profound cortical atrophy with gliosis and vacuolation of neuropils, and cystic cavitation

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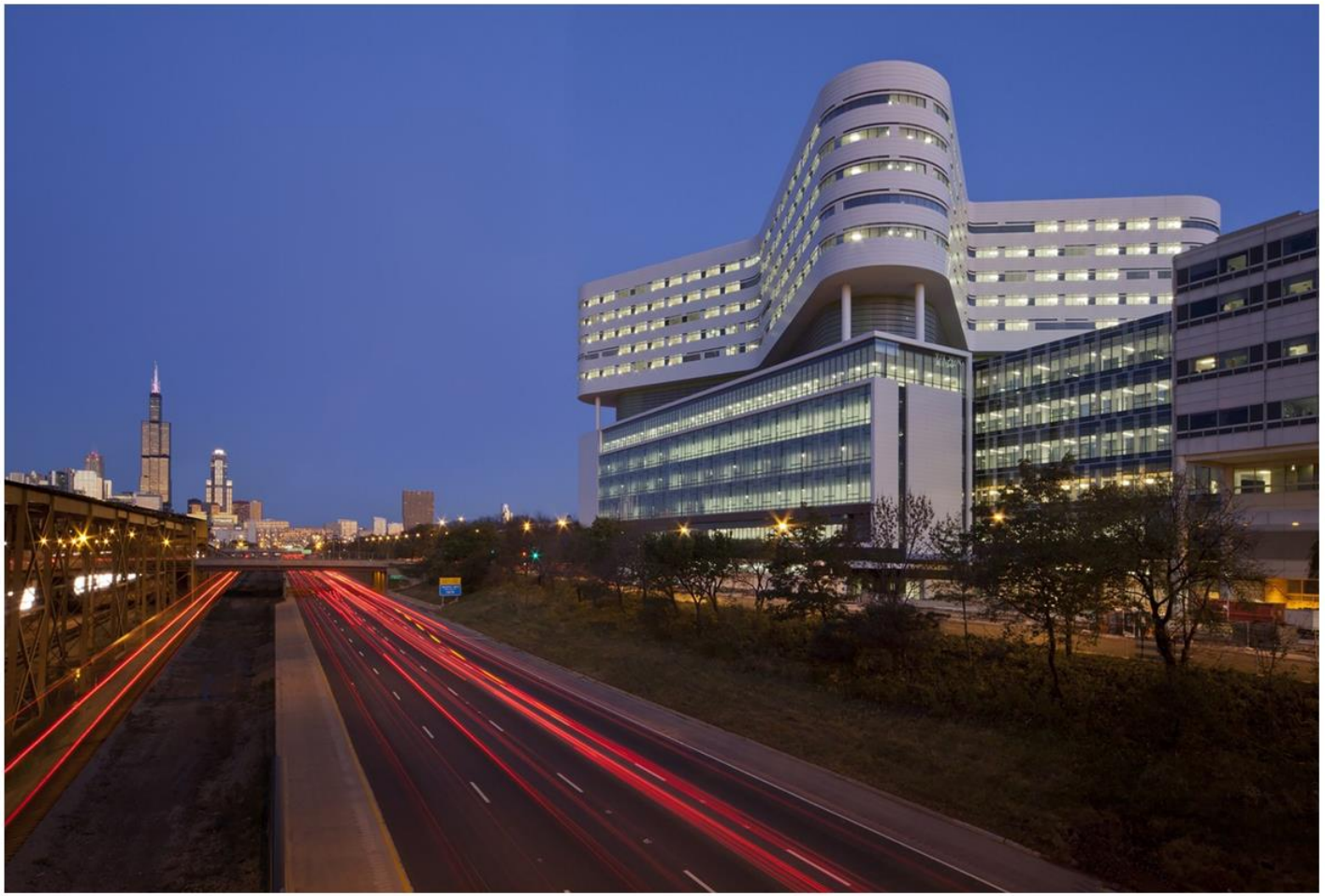

Thank You!