Diagnosis of Primary CNS Vasculitis	CNS Vasculitis (Suspected)	Other Diagnosis

Clinical Presentation:
Newly acquired neurological deficits (focal and/or diffuse) and/or psychiatric symptoms

Evaluation-First Step:
Clinical Assessment:
- General assessment for underlying disease/illness;
- Detailed neurological tests; psychiatric/neurocognitive assessment (when applicable)

Blood Tests:
- Blood inflammatory markers (E.g. ESR, CRP, CBC)
- Auto-antibody testing (E.g. antinuclear antibody, antineutrophil cytoplasmic antibody)

Diagnostic Imaging:
- Cerebral MRI (with gadolinium enhancement, T2, and FLAIR);
- MRA with post-gadolinium vessel-wall imaging and/or other cerebral vessel imaging (E.g. CTA)

CSF analysis:
- Opening pressure; CSF cell count; Infectious work-up (bacterial, fungal, and viral cultures and serology); oligoclonal banding

Evaluation-Second Step: CSF analysis:
- Neuronal antibodies (E.g.; anti-NMDAR, anti-AMPA, anti-AQP4, anti-LG1)

Blood Test:
- Anti-NMDAR, anti-AMPA, anti-AQP4, anti-LGI1
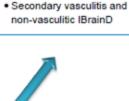
Consider other diagnostic Imaging:
- Conventional angiography

Evaluation-Third Step:
Brain biopsy
Lesional or non-lesional; non-dominant frontoparietal lobe

- Non-inflammatory vasculopathies (E.g, atherosclerosis)
- Non-vascular inflammatory brain diseases (E.g. ADEM, MS)
- Mimic of vasculitis (E.g. RCVS) or IBrainD
- Secondary vasculitis and non-vasculitic IBrainD