

Primary Central Nervous System Vasculitis

Navigating Through Diagnostic Fog

Pr Hubert de Boysson
Department of Internal Medicine
CHU CAEN
France



Why talking about CNS vasculitis ?

Diagnosis is often suspected

Diagnosis is rarely established

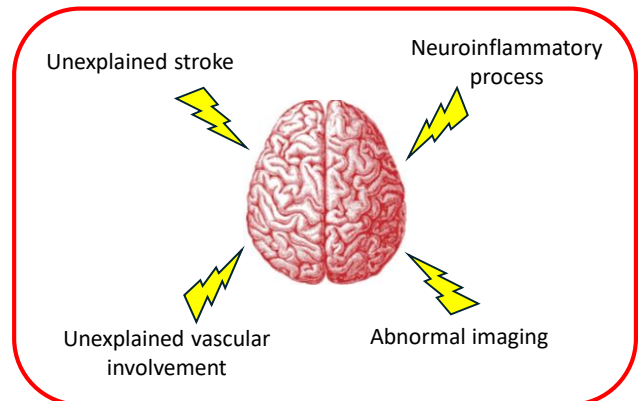
Incidence: 2 cases per million inhabitants¹

Diagnosis is sometimes wrong

Many mimics

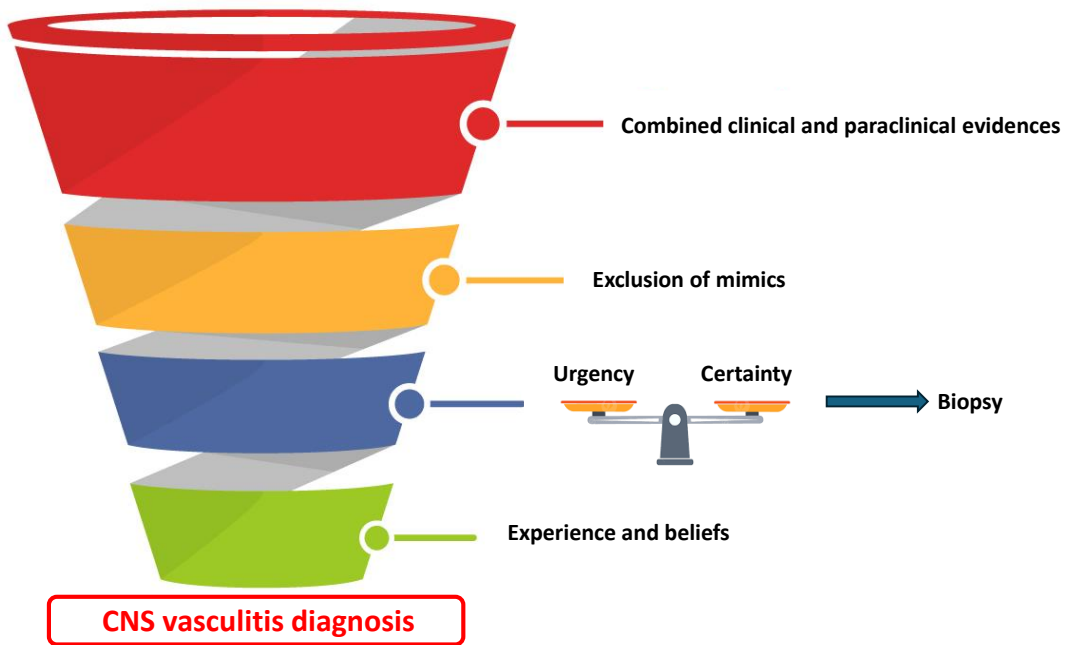
No standardization of the diagnostic approach

Diagnosis criteria not updated²

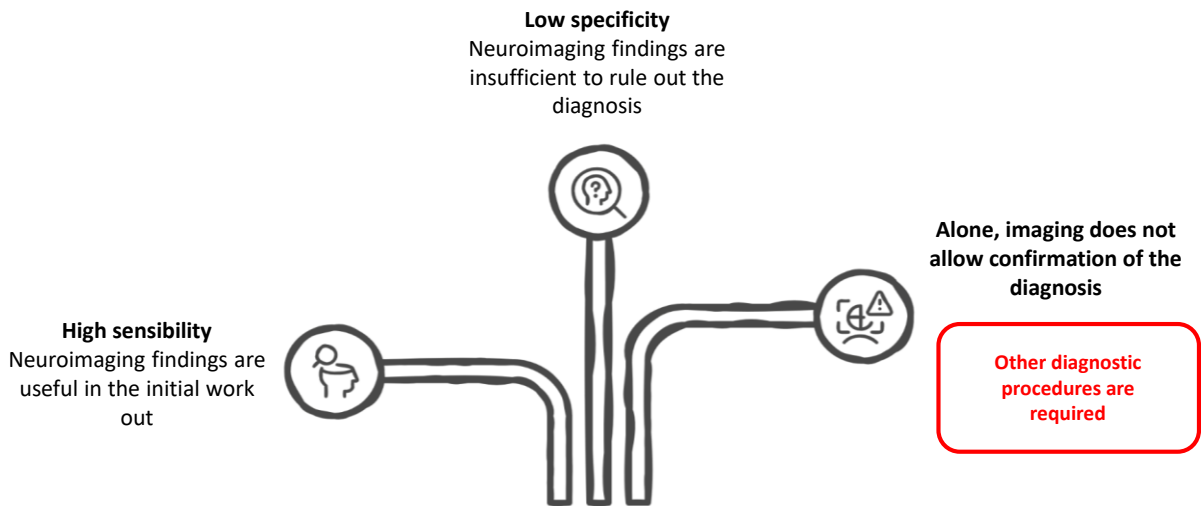


¹Salvarani C et al, Ann Neurol, 2007

²Calabrese & Malek, Medicine, 1988



Neuroimaging findings



Lumbar puncture

Abnormal in 50 to 85% of cases, whatever the CNSV cause



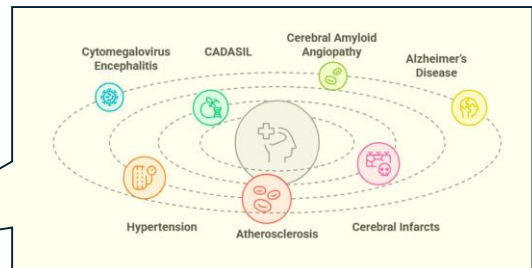
- Between 10 and 400 leukocytes
- Proteinorachy between 0,5 and 2 g/l
- Oligo-clonal bands are possible
- HSV and VZV PCR (+/- infectious explorations)
- Histology, flow cytometry, clonality research

BE CAREFUL

- Mild meningeal reaction post-stroke (5-10 cells, prot: 0.5-0.7 g/l)
- If Leuko >400/mm³ -> look for lymphoma or auto-immune encephalitis

Birnbaum J et al, Arch Neurol, 2009
Hajj-Ali RA, Autoimmun Rev, 2013

- 4% of complications
- Sensibility: 60 to 75%
- Alternative diagnoses



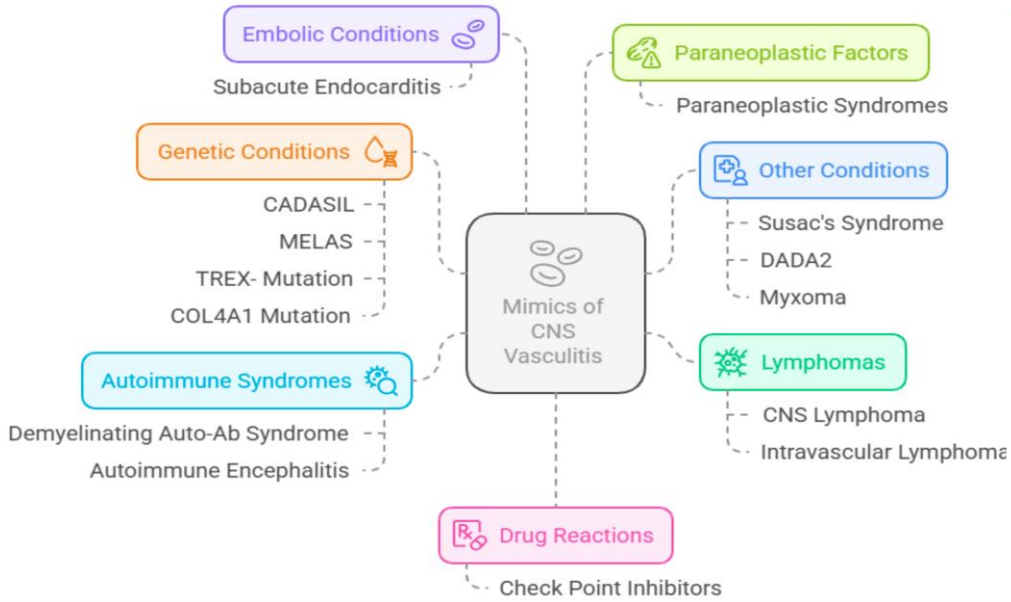
- Cure the wrong disease
- Treatment-related adverse events

Cerebro-meningeal
biopsy

Immunosuppressants

Torres J et al, Stroke, 2016
Bai HX et al, Neurosurgery, 2015
Hajj Ali R, ACR 2025

Primary CNS vasculitis mimics

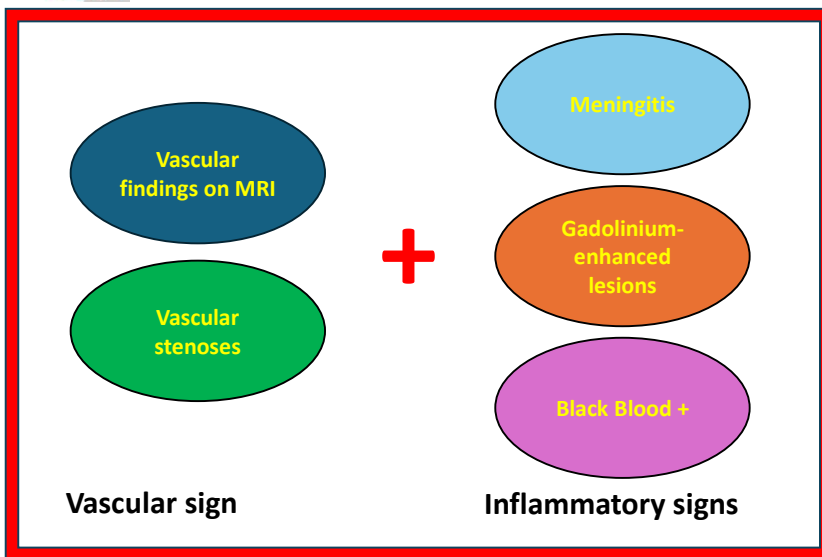


Hajj Ali R, ACR 2025

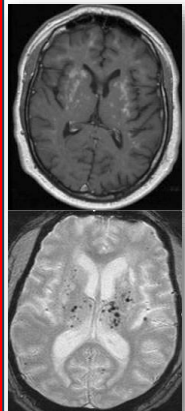
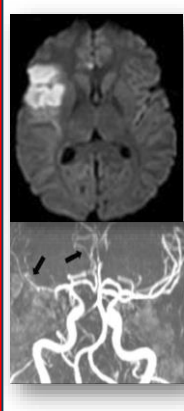


In absence of a positive biopsy

YOU CAN RETAIN THE DIAGNOSIS OF CNS VASCULITIS IF



Clinical and radiological patterns

	<h3>SMALL VESSELS</h3> <ul style="list-style-type: none"> • Encephalopathy • Seizure • Gadolinium-enhanced lesions 		<h3>MEDIUM/LARGE VESSELS</h3> <ul style="list-style-type: none"> • Focal déficits • Cerebral infarctions • Vascular stenoses
---	---	---	---

Therapeutic approach

- **Glucocorticoids (GC) always**
- **GC withdrawal between M12 and M18**
- **Immunosuppressant in all patients**
- **Aspirin according to CV risk factors**



Induction treatment

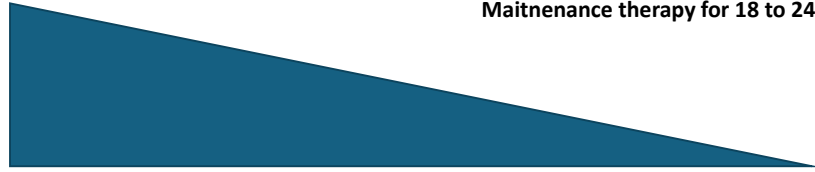
Cyclophosphamide 0,6 g/m² D1, D15 et D29,
then 0,7 g/m² D50, D71 et D92 (x 6)



Maintenance therapy

- Azathioprine 2 mg/kg/j (Max 200 mg/day)
- Methotrexate 0,3 mg/kg/week SC (Max 30 mg/week)
+ folic acid
- Mycophenolate mofetil (2-3 grams/day)
- Rituximab (500 mg or 1000 mg every 6 months IV)

Maintenance therapy for 18 to 24 months



Glucocorticoids for 12-18 months



Rituximab 1000 mg D1 and D15

Associated treatments

- Calcium and vitamin D
- Bisphosphonates
- Vaccines (flu, pneumococcal, COVID, zona)

Take Home Messages

- Multidisciplinary approach (neurologist, internist, neuradiologist, neurosurgeon)
- Identify the two radio-clinical phenotypes
- Discuss a biopsy if « small-vessel » pattern
- In absence of a biopsy: vascular criteria + inflammatory criteria
- Treatment = glucocorticoids + immunosuppressant