Central nervous system vasculopathy refers to any process that affects central nervous system vessels. It includes either inflammation of the vessel wall (vasculitis) or other non-inflammatory etiologies. In the first group, primary central nervous system vasculitides are an uncommon and laborious diagnosis that must be remembered. Secondary causes of vasculitis include systemic vasculitis with central nervous system impairment, infectious causes and others. Non-inflammatory vasculopathies include a wide range of diseases where the reversible cerebral vasoconstriction syndrome should be emphasized due to its distinct presentation, treatment and clinical course.

The challenge remains on precociously differentiating these entities to start the appropriate treatment earlier and thus impact prognosis. Some main differential diagnoses will be discussed in detail. A possible diagnostic algorithm will be presented.