Among all forms of vasculitis, perhaps that affecting the CNS (CNS-V) poses the greatest challenge in diagnosis given the lack of any non-invasive high sensitivity test, the complex and extensive list of mimicking conditions, its inaccessibility for biopsy and its genuine rarity. The diagnosis still requires the presence of an unexplained neurologic sign or symptom(s) following an exhaustive evaluation, evidence of vascular involvement (by direct or indirect angiography) or biopsy and most importantly the meticulous exclusion of all those conditions capable of producing mimicking clinical, radiographic or histologic findings that would confound accurate diagnosis. Despite these challenges CNS-V has been increasingly reported due to a combination of increased diagnostic awareness and advances in diagnostics. Two of these major advances will be discussed including the use of direct vascular wall imaging to differentiate vascular inflammation from spasm or atherosclerosis and the use of next generations sequencing to identify infectious etiologies and obviate the use of biopsy. Clinical examples will be presented.

REFERENCES: