

Isolated Wegener's Pachymeningitis

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The stereotypical CNS manifestations of the ANCA-related vasculitides (specifically Granulomatosis with polyangiitis (GPA, formerly Wegener's) and Microscopic Polyangiitis) have been well described in the literature. Recently, case reports have described a novel presentation of GPA as an isolated pachymeningitis.

A 65 year old man presents to physicians with subacute progressive onset of new headaches and gait instability. Within a month, the patient had gone from healthy to requiring a walker. He was initially diagnosed with aseptic meningitis and treated unsuccessfully with valacyclovir, and presented with increasing symptoms to UVMC Neurology. MR found diffuse pachymeningeal thickening that did not extend into the spinal cord, with the neurologic exam with findings of a wide based ataxic gait requiring 1 assist, subtle bilateral proximal weakness, and bilateral dysmetria.

A comprehensive laboratory workup found CSF 37 nucleated cells with lymphocytic predominance, positive ANA speckled, pANCA, MPO. MR of the neuroaxis along with a PET scan were unremarkable. A dural biopsy found obliterative vasculitis with associated dural necrosis.

The patient was started on a high dose steroid course, and within 2 days had almost complete resolution of symptoms. Unfortunately, on attempted steroid wean, he reported increasing headaches, ataxic gait with falls, and new diplopia on downward gaze. His steroid course was increased, and ultimately was started on cyclophosphamide with residual ataxic gait.

This case illustrates the newly emerging entity known only as an ANCA-associated pachymeningitis. Currently, only approximately 15-16 confirmed case reports exist, though new evidence from Japan suggests a more common and previously undescribed association between pANCA positivity and complex (limited) CNS presentation. This may represent a new previously undescribed clinical entity, and hold clues to underlying neuroimmunologic and neuropathologic processes.