Vasculitic neuropathy as the sole manifestation of microscopic polyangiitis

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Abstract:

Background: Microscopic polyangiitis (MPA) is an anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis characterized by inflammation of small-sized vessels[1]. Common manifestations include glomerulonephritis, pulmonary capillaritis, arthritis, and neuropathy[2]. Anti-myeloperoxidase antineutrophil cytoplasmic antibodies (MPO-ANCA) are detected in up to 80-90% of patients with active MPA[3]. Peripheral nervous system (PNS) involvement is common with MPA, however, exclusive involvement is extremely rare[4]. We report the unusual case of MPA combined with vasculitic neuropathy solely.

Case report: A 78-year-old woman was admitted for evaluation of subacute worsening mental status and profound weakness. Her medical history included hypertension, diabetes mellitus, and Alzheimner's dementia. A recent endothelial keratoplasty with postoperative right eye ptosis was reported. The patient was alert and attentive, followed one step commands, and had no aphasia with intact comprehension and fluent speech. Cranial nerve examination revealed right eye ptosis. Motor exam revealed bilateral wrist and foot drop. Reflexes were absent throughout. Sensation was intact with no sensory level. Initial labs showed high values of erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), and elevated white blood cell count. Her chest X-ray showed no acute abnormalities. Computed tomography (CT) of the head showed no evidence of acute intracranial hemorrhage, large vessel territory infarct, or mass effect. Magnetic resonance imaging (MRI) of the brain showed mild chronic ischemic and involutional changes. Spinal cord imaging revealed no evidence of cord compression. CSF analysis was unremarkable. EMG revealed a severe length-dependent axonal sensory and motor polyneuropathy. A subsequent left sural nerve biopsy identified chronic active vasculitis involving endoneurial and epineurial blood vessels as well as chronic severe peripheral neuropathy. Additional lab workup revealed the presence of a high-titer MPO-ANCA of 8 AI (*normal values* <1 AI) and a positive ANA >1:160 (-cytoplasmic pattern). Anti-proteinase 3 antineutrophil cytoplasmic antibody (PR3-ANCA) was negative. Urinalysis showed no blood or protein and CT of the chest was unremarkable. Extensive testing revealed no infectious etiology for her illness. The clinical presentation of vasculitic neuropathy, in conjunction with a high-titer MPO-ANCA antibody, established an underlying diagnosis of MPA. Intravenous (IV) methylprednisolone (1 g daily for 5 days), followed by oral tapering with prednisone was initiated. Azathioprine was added to the steroid regimen with the goal to control active vasculitis, prevent further nerve damage and potentially other organ involvement. Aggressive immunosuppressive therapy was deferred in light of poor functional status and chronicity of the lesions.

Conclusion: PNS involvement of MPA that leads to bilateral wrist and foot drop solely is extremely rare. Michael P Collins et al have indicated that corticosteroid monotherapy for at least 6 months is considered the first-line in the management of non-systemic vasculitic neuropathies. Addition of immunosuppressive agents can be considered based on disease severity[5]. This case illustrates the need to include nervous system assessment in the evaluation of MPA patients, and the importance of early diagnosis and prompt institution of glucocorticoid therapy, which can prevent severe organ damage and adverse outcomes.

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