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Microscopic Polyangiitis Presenting With A Pure Sensory Peripheral Symmetric Polyneuropathy

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Background

- Microscopic polyangiitis (MPA) is a systemic small-vessel necrotizing vasculitis with a strong association with anti-myeloperoxidase–anti-neutrophilic cytoplasmic antibodies (MPO-ANCA). MPA has little or no immune deposit and is not associated with granulomatous inflammation.  
- In most cases MPA presents with rapidly progressive necrotizing glomerulonephritis and occasionally with pulmonary hemorrhage. Over 70% of patients have constitutional symptoms at diagnosis and disease manifestations may involve any organ system. MPA may also present without renal or pulmonary involvement.  
- Peripheral neuropathy has frequently been reported as a symptom of ANCA-associated vasculitis and is commonly present at disease onset. It may also be an initial manifestation of the disease state, but there is limited evidence comparing the incidence of neuropathy with the other manifestations of systemic vasculitides. 
- Studies have shown that a majority of those with ANCA-associated vasculitis report peripheral neuropathy at some point in the disease process, but less than 25% report peripheral neuropathy in the early phases of the disease process, but less than 25% report peripheral neuropathy. 
- A recent case report of MPA presents a patient with lower extremity polyneuropathy preceding renal manifestations.  
- Previous literature has demonstrated that a majority of those with ANCA-associated vasculitis report peripheral neuropathy at some point in the disease process. 
- Patients may also be treated with plasma exchange during induction phase. 

Case Presentation

A 58 year old woman without significant past medical history presented with the chief complaints of weakness and loss of balance. She described a weakness and heaviness of her bilateral lower extremities and felt off-balance with difficulty ambulating. She also described a clumsiness and inability to correctly use her left hand. This occurred in the context of a one year history of “pins and needles” sensation in her bilateral feet. 

Physical exam revealed a stocky-glove distribution of decreased pinprick sensation, decreased vibration sense in the bilateral toes, ataxic gait, and a positive Romberg sign. The patient underwent extensive imaging revealing the following results:  
- CT Head: no acute abnormality 
- MRA Head/Neck: 50% stenosis of proximal right ICA, 75% stenosis of left ICA, both felt to be artifactual 
- MRI Brain: scattered foci of non-specific supratentorial white matter signal abnormality, but no acute abnormality 
- Chest XR: interstitial prominence laterally at both lung bases most likely represents chronic interstitial disease 
- 2 DCHO: aneurysmal interstitial septum without evidence of shunt, EF 60% 
- CT Chest: interstitial lung disease, mild mediastinal and hilar adenopathy, mild hombecoming at bilateral bases 
- CT Abd/Pelvis: 6cm cystic right pelvis mass, representing likely ovarian cyst 

Laboratory studies revealed an elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), positive rheumatoid factor (RF), and elevated serum creatinine. Patient was ultimately discharged home with a diagnosis of peripheral neuropathy likely secondary to thiamine deficiency. 

Three months after discharge the patient presented with the acute onset of subternal chest pain exacerbated by inspiration and associated with shortness of breath. 

Initial laboratory examination revealed markedly elevated acute oligoanuric renal failure, proteinuria, anemia, and new diffuse bilateral perihilar infiltrates on chest x-ray and CT. Bronchoscopy revealed acute alveolar hemorrhage. Further laboratory studies demonstrated elevated anti-myeloperoxidase–anti-neutrophilic cytoplasmic antibodies (MPO-ANCA). The patient then underwent renal biopsy which revealed acute and chronic pauci-immune necrotizing, sclerosing, and crescentic glomerulonephritis associated with focal necrotizing vasculitis; typical of changes seen with MPO-ANCA seropositivity, leading to a diagnosis of microscopic polyangiitis. 

The patient was initiated on hemodialysis due to her rapidly progressive glomerulonephritis. She was treated with pulse dose methylprednisolone followed by stress dose prednisone, as well as therapeutic plasma exchange and oral cyclophosphamide. Her maintenance phase included azathioprine and prednisone. 

Discussion

Our case is a rare presentation of microscopic polyangiitis (MPA) with concurrent diffuse alveolar hemorrhage and dialysis dependent rapidly progressive glomerulonephritis in the setting of long-standing bA-NaCl neering-glove distribution pure sensory symmetric peripheral polyneuropathy. 

Previous literature has demonstrated that a majority of those with ANCA-associated vasculitis report peripheral neuropathy at some point in the disease process, but few have had active vasculopathic neuropathy at baseline. 

Patients with vasculitic neuropathy at baseline have a higher median number of organ systems involved when compared with those without neuropathy, and also reported higher Birmingham Vasculitis Activity Scores, which is a clinical checklist of items organized by organ system used to determine disease activity. 

The European Vasculitis Study Group trials showed that a pure sensory neuropathy, which our patient developed early in her disease course, was not reported in any patients with MPA. However, our patient did not undergo electrophysiologic confirmation of her neuropathy. 

Studies demonstrate that non-specific symptoms may be present for months to years prior to diagnosis of MPA. This case demonstrates the difficulty of recognizing MPA when only a single organ system is involved and that ANCA-associated vasculitis should be included on the differential diagnosis of peripheral neuropathy with unclear etiology. It also exemplifies the need for further research on peripheral neuropathy in systemic vasculitides, specifically in MPA where these early presenting signs have previously shown to be less prevalent. 

References:


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