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An Observational Study of Polyneuropathy in a Geriatric Patient Due to ANCA Associated Vasculitis (Microscopic Polyangitis) – A Diagnostic Challenge

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Abstract

ANCA associated vasculitis usually presents with features of renal vasculitis however in this observational study this geriatric patient presented with polyneuropathy without any features of renal involvement, thus making the diagnosis challenging.

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I. Introduction

The anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides target small- to medium-sized vessels in multiple organ systems, with sinonasal, pulmonary, and renal involvement being particularly common[2]. Specific diseases included in this category are granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), eosinophilic granulomatosis with polyangiitis (EGPA; also known as Churg-Strauss syndrome), and renal limited vasculitis.These diseases are categorized together, given their association with antibodies directed against antigens in the cytoplasm of neutrophils—proteinase 3 (PR3) and myeloperoxidase(MPO)—and many overlapping clinical manifestations,diagnostic testing, and treatment strategies.

The wide range of prevalences of neuropathies reported in specific vasculitidesreflects differences in how neuropathies were ascertained (clinically,electrophysiologically, and/or through tissue biopsies) and whethercohorts were composed of unselected patients or highly select patients
studied at tertiary care neuromuscular centers.

II. Materials and Methods

62 year old female non hypertensive and non diabetic, known case of hypothyroidism on tab levothyroxine 100µg, presented with weakness of right upper limb an left lower limb for 4 weeks which was insidious in onset and gradually progressive in nature. It was also associated with tingling and numbness of all four limbs which was also insidious in onset and gradually progressive in nature. There was no history of bowel, bladder involvement and cranial nerve involvement. However she complained of joint pain involving multiple small and large joints which was inflammatory in nature for last 1 year. No history of hematuria, dysuria, rash, alopecia etc.

On examination patient was conscious alert and cooperative, Pulse- 84/ min, BP 110/70 mmHg, Pallor-moderate, cyanosis,odema , jaundice, clubbing- nil.

Neurological examination revealed that there was Lower motor type weakness along with sensory involvement. Other system examination was within normal limits.

Her blood reports showed Hb-7.8 gm %, TC- 8100/cmm, Platelet- 2.5 lac/cmm, N88L9E2M1 , ESR-140 , CRP- 83, LFT, urea and creatinine- WNL. Urine R/E, M/E showed pH 6.0, pus cell- 20, RBC- 20-25 with RBC cast. 24 hr urinary protein was 497mg. NCV of all 4 limbs showed axonal type of sensorymotor polyneuropathy. CT Scan thorax showed diffuse parenchymal lung disease. Kidney biopsy shows mesangioproliferative glomerulonephritis with subacute tubulo-interstitial nephritis. Immunofluroscence study was negative. ANA, ENA profile were negative. P-ANCA was positive with high MPO -208.94 RU/ml (n<20 RU/ml). Rheumatoid factor was 42 IU/ML. AntiCCP was negative.

The patient was diagnosed as a case of sensorymotor polyneuropathy due to associated vasculitis (microscopic polyangitis). Patient was treated with IV cyclophosphamide , azathioprime, low dose steroid. Patient's condition improved.

III. Discussion

Microscopic polyangitis (MPA) is a very rare disease characterized by the lesions of arteriolae, venulae and capillaries, as well as medium arteries mainly of the kidneys and lungs, but also of other systems and organs. The reported incidence of MPA in Europe is from 2.7 per million in Norway to 11.6 per million in Spain. Peak incidence of microscopic polyangiitis is between 65-74 years of life. Small vessels vasculitis connected with ANCA are idiopathic diseases of unknown etiology. Compared the clinical aspects of peripheral neuropathy associated with Wegener's granulomatosis (26 patients), Churg-Strauss syndrome (26 patients) and microscopic polyangiitis (12 patients) in a single center cohort study conducted between 1999 and 2006, Cattaneo et al. were confirmed peripheral neuropathy in 27/64 patients . Neuropathy occured earlier and presented with severe form of mononeuritis multiplex in patients with MPA compared to patients with Wegener's granulomatosis. Electrophysiologic findings and sural nerve biopsy confirmed acute axonal changes in MPA . Zhang et al. were observed nervous system involvement in 36.6 % of MPA patients. Microscopic polyangiitis includes mainly mononeuritis multiplex or distal symmetrical polyneuropathy[3].

IV. Conclusion

Results from clinical and laboratory tests can support an immune ANCA vasculitis, but kidney biopsy and histopathology examination is necessary for early and exact diagnosis of ANCA vasculitis microscopic polyangiitis type with ANCA-associated glomerulonephritis, as well as early starting of therapy, which determined outcome of disease. Polyneuropathy of extremities presented much more before kidney symptomatology could be a first manifestation of an atypical presentation of MPA.

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