Systemic vasculitis and nonsystemic vasculitic neuropathy (NSVN)

Authors: E.P. Verrengia, A. Romorini, P. Mantegazza, M. V. Calloni, P. Perrone

UO NEUROLOGIA -STROKE UNIT OSPEDALE LEGNANO-MAGENTA ASST OVEST MILANESE

Objective: ANCA (antineutrophil cytoplasm,atic antibody) associated vasculitis can include Wegener's granulomatosis, microscopic polyangiitis, Churg-Strauss syndrome and necrotinzing glomerulonephritis. Differential diagnosis includes chronic inflammatory demyelinating polyneuropathy (CIDP), mutifocal motor neuropathies, lower motor neuron disorders. Materials: we report a case with neuropathy as a single manifestation of vasculitis (NSVN) compared to a patient with extra-neurological involvement.

Method: both patients underwent the same examinations: general hematochemistry, autoimmunity, cerebrospinal fluid analysis, EMG (Electromyography), chest-abdomen TC (Computerized Tomography), tissue biopsy.

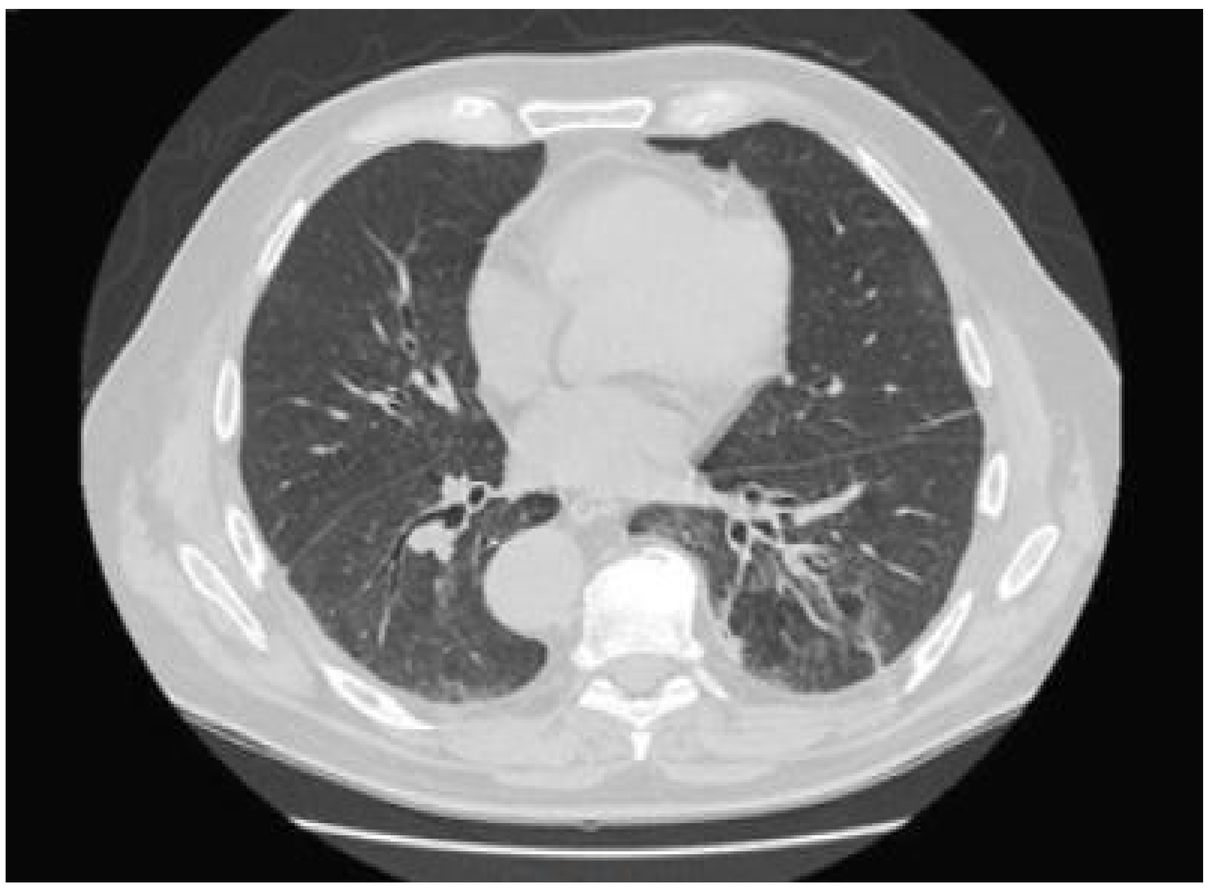

Figure 1. *High Resolution Chest Computed Tomography of 74 yrs old man showing lower lobe lobular bronchiectasis and polished glass areas in subpleural site*

Results: a 79 year-old woman was admitted with painless motor- sensory deficit involving distal lower limbs; examinations revealed elevated p-ANCA antibodies (682 UA/mL; normal value < 10), distal motor sensory axonal neuropathy detected by EMG, inflammatory infiltrate on sural nerve biopsy. A 74 year-old man developed mild motor painless weakness of right hand and left foot; one month later, symptoms progressed in the left hand, haematologic chemistry revealed high p-ANCA (565.4 UA/ml; normal value < 10), kidney dysfunction (creatinine 1.95 mg/dL, normal value < 1.1 mg/dL), renal biopsy showed extracapillary glomerulonephritis, polished glass areas at chest high resolution tomography (figure 1). They were treated in acute phase with IVIg (intravenous immunoglobulin) and high dose of steroid; subsequently they underwent long-term immunosuppressive therapies with azathioprine in the first case and mycophenolate mofetil as man's treatment. Both presented a flue like syndrome prior to the developement of neuropathy.

Discussion: The diagnosis of vasculitic neuropathy should be established by tissue biopsy (nerve or kidney). The immunosuppressive therapy was chosen related to age, potential side effects, presence of systemic manifestations.

Conclusions: Vasculitic neuropathy may appear as a first clinical manifestation of systemic disease, sometimes with long latency between onset of neuropathy and systemic symptoms. In these patients long-term follow-up is necessary to control the history of disease and ensure the best medical management. Neuropathy ANCA-related less commonly represents a NSVN. Microscopic polyangiitis (MPA) and Churg-Strauss syndrome (CSS) share common peripheral nervous system features with an extension of systemic involvement significantly higher in MPA, resulting in poorer survival rates. NSVN does not affect survival but can disturb day-to-day functioning and quality of life. Histopathological evaluation should be considered in all these patients to evaluate the risk to develop systemic manifestations.

Bybliography: Steven A Greenberg. p-ANCA vasculitic neuropathy with 12-year latency between onset of neuropathy and systemic symptoms. BMC Neurol. 2002;2-10.Collins MP. The vasculitic neuropathies: an update. Curr Opin Neurol. 2012 Oct;25(5):573-85. Gorson KC. Therapy for vasculitic neuropathies. Curr Treat Options Neurol. 2006 Mar;8(2):105-17.