ASSESSING COGNITIVE HEALTH OUTCOMES IN ANCA-ASSOCIATED VASCULITIS PATIENTS

Student: **Ruo Yan Chen**
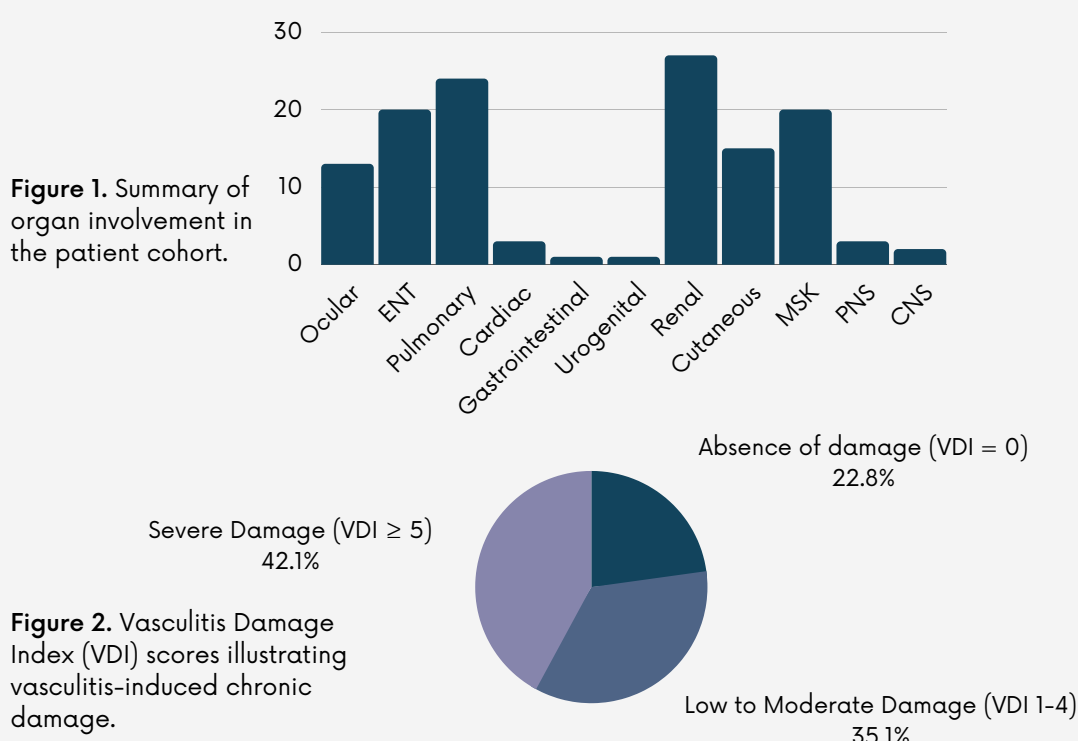Supervisor: **Dr. Lillian Barra**

BACKGROUND

Anti-neutrophilic cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a rare and life-threatening autoimmune disease with severe complications, including damage to various major organs. The disease is caused by small blood vessel inflammation in which white blood cells attack endothelial cells in blood vessel linings, inducing swelling and hemorrhage. In the past, AAV was a fatal disease, but with timely immunosuppressant treatment, the survival of AAV patients has significantly improved. Nevertheless, AAV is still associated with frequent relapses and significant irreversible damage.

Studies looking at the cognitive health outcomes of AAV patients with central nervous system involvement have noted cognitive impairment as a significant contributor to reduced quality of life with unique challenges in care and treatment. It is important to evaluate the cognitive outcomes of AAV patients to aid in early detection at various stages of disease progression. In this output, we summarized the demographics and disease characteristics of AAV patients recruited from renal-rheumatology vasculitis clinics at St. Joseph's Health Care and a glomerulonephritis clinic at London Health Sciences Centre.

DATA & DISCUSSION

We recruited 37 patients (16 male and 21 female) between 18 and 75 years old who meet the American College of Rheumatology classification criteria for AAV. Fourteen patients had the active disease and 23 patients were in remission. The patients had a mean age of 53.07 ± 14.18 at the time of assessment. Disease characteristics (Figure 1) and disease scores (Figure 2) are summarized.

Figure 1. Summary of organ involvement in the patient cohort.

Figure 2. Vasculitis Damage Index (VDI) scores illustrating vasculitis-induced chronic damage.

Absence of damage (VDI = 0) 22.8%

Severe Damage (VDI ≥ 5) 42.1%

Low to Moderate Damage (VDI 1-4) 35.1%

The data collected from this primary output could be combined with global assessments of cognitive function as well as the application of advanced imaging tools to study the relationship between vascular abnormalities, disease activity and cognition. Our findings could prompt the use of non-invasive imaging to identify early signals of AAV complications and prevent irreversible cognitive damage at all stages of disease progression.

REFERENCES

1. Graf J. Central Nervous System Disease in Antineutrophil Cytoplasmic Antibodies-Associated Vasculitis. Rheum Dis Clin North Am. 2017;43:573–578.
2. Mattioli F, Capra R, Rovaris M, Chiari S, Codella M, Miozzo A et al. Frequency and patterns of subclinical cognitive impairment in patients with ANCA-associated small vessel vasculitides. J Neurol Sci. 2002;195:161-6.
3. Novak RF, Christiansen RG, Sorensen ET. The acute vasculitis of Wegener's granulomatosis in renal biopsies. Am J Clin Pathol. 1982;78:367 –371.
4. Salmela A, Ekstrand A, Joutsi-Korhonen L, et al. Activation of endothelium, coagulation and fibrinolysis is enhanced and associates with renal anti-neutrophil cytoplasmic antibody-associated vasculitis. Nephrol Dial Transplant. 2015;30 Suppl 1:i53-9.
5. Pagnoux C, Chironi G, Simon A, Guillevin L. Atherosclerosis in ANCA-associated vasculitides. Ann N Y Acad Sci. 2007;1107:11-2.
6. De Leeuw K, Sanders JS, Stegeman C, et al. Accelerated atherosclerosis in patients with Wegener's granulomatosis. Ann Rheum Dis 2005; 64:753 –9.
7. Masi AT, Hunder GG, Lie JT, et al. The American College of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). Arthritis Rheum. 1990;33:1094-100.
8. JKroenke K, Spitzer RL, Williams JB. The PHQ-9: validity of a brief depression severity measure. Gen Intern Med. 2001;16:606-13.
9. Mukhtyar C, Lee R, Brown D et al. Modification and validation of the Birmingham Vasculitis Activity Score (version 3). Ann Rheum Dis. 2009 Dec;68(12):1827-32.
10. Exley AR, Bacon PA, Luqmani RA, et al. Development and initial validation of the Vasculitis Damage Index for the standardized clinical assessment of damage in the systemic vasculitides. Arthritis Rheum. 1997;40:371-80.