Conclusion: Our small study indicates that clinical and morphological features of renal involvement, as well as renal survival are similar in ANCA-negative and ANCA-positive patients and don't depend on the type of ANCA.

References:

- [1] Berden AE, Ferrario F, Hagen EC, et al. Histopathologic Classification of ANCA-Associated Glomerulonephritis. *J Am Soc Nephrol*. 2010;21(10):1628–1636.
- [2] Brix RB, Noriega M, Tennstedt P, et al. Development and validation of a renal risk score in ANCA-associated glomerulonephritis. *Kidney Int*. 2018;94(6):1177–1188.

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AB0498

CHARACTERISTICS AND OUTCOME OF CORONARY ARTERY LESIONS DUE TO POLYARTERITIS NODOSA: ANALYSIS OF A SINGLE CENTER COHORT IN CHINA

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Background: Polyarteritis nodosa (PAN) is a systemic necrotizing vasculitis that mainly affects medium-sized muscular arteries. The coronary artery could be affected. Some severe cases can lead to spontaneous coronary artery dissection (SCAD) and about 0.02% will die abruptly. Early diagnosis will improve prognosis, but relative studies are all case reports so far.

Objectives: To investigate the clinical characteristics, risk factors and outcome of patients with polyarteritis nodosa (PAN) complicated with coronary artery lesions in China.

Methods: Data of 158 patients with PAN who were admitted to Peking Union Medical College Hospital from September 1986 to September 2019 were retrospective collected. Data were analyzed and compared according to with and without coronary artery lesions due to PAN.

Results: 17 (10.8%) patients with PAN had the coronary artery lesions due to PAN. The age at coronary artery lesion was 36.9±10.3 years. 12 (70.6%) patients were male. There are not statistical differences between two groups in common risk factors of coronary arterial atherosclerosis including smoking, hypertension, diabetes mellitus and hyperlipidemia. Most of them are multi-vessel lesions (8 cases are triple-vessel lesions and 3 cases are bi-vessel lesions). Type of coronary artery affected is shown mainly in stenosis (13 cases). Myocardial infarction are shown in 8 cases (47.1%). Compared to patients without coronary artery lesions, patients with coronary artery lesions had less nervous system involvement (17.6% vs.46.8%) and elevated number of leukocyte (17.6% vs.56%). Besides, patients with coronary artery affected exhibit more cranial and carotid artery involvement (29.4% vs. 5.0%), renal artery involvement (41.2% vs.17.0%), coeliac artery involvement (58.8% vs.27.0%), new onset hypertension (47.1% vs.14.5%), renal infarction (27.3% vs.5.4%), and higher proportion of 2009 Five-factor score (FFS)≥2 (62.5% vs.15.6%). All patients with coronary artery lesions received at least moderate dose of prednisone and CTX except one refused medication. 3 cases underwent interventional therapy. Stent placement was performed on 2 of them, and in-stent stenosis was appeared in a patient one year later. 2 cases died, one for vascular rupture after coronary aneurysms true plus coronary artery bypass grafting, another for myocardial infarction after stopping immunosuppressant therapy himself. Survival analysis showed patients with digital gangrene had poor prognosis though no significant difference (p=0.055).

Conclusion: PAN with coronary artery lesions are not uncommon. These patients exhibit young age, more proportion of multi-vessel of coronary artery involvement, more combined involvements of other organ arteries and more severe disease.

References:

- [1] Munguti CM, Ndunda PM, Muutu TM. Sudden Death From Spontaneous Coronary Artery Dissection Due to Polyarteritis Nodosa. *Cureus*. 2017;9(10), e1737
- [2] Krita nawong C, Kumar A, Johnson KW, et al. Conditions and Factors Associated With Spontaneous Coronary Artery Dissection (From a National Population-Based Cohort Study). *Am J Cardiol*. 2019; 123 (2): 249-253
- [3] Hwang J, Yang JH, Kim DK, Cha HS. Polyarteritis Nodosa Involving Renal and Coronary Arteries. *J Am Coll Cardiol*. 2012;59(7):e13

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AB0499

ABNORMALITY OF PERIPHERAL LYMPHOCYTE SUBSETS IN BECHET'S DISEASE AND EFFECTS OF NEW IMMUNOREGULATORY COMBINATION THERAPIES ON THESE CELLS

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Background: Bechet's disease (BD) is a chronic multisystemic vasculitis. Although its exact etiopathology is unknown, both autoimmune imbalances associated with genetic and abnormal immune response of effector lymphocytes promoted by infectious factors are suggested¹. The increase of effector T cells (Teffs) and the decrease of regulatory T cells (Tregs) are possibly the involving factors in the pathogenesis of BD². Importantly, we have developed new immunoregulatory combination therapies trying to restore the reduction of Tregs in rheumatic patients.

Objectives: To examine abnormal levels of lymphocyte subsets in BD patients at a relatively large-sample size and to investigate whether the immunoregulatory combination therapies have therapeutic efficacy in BD.

Methods: Total 384 BD patients and 206 healthy controls (HCs) were enrolled in this cross-sectional study. Proportions and absolute numbers of peripheral T, B, NK, CD4⁺ T, CD8⁺ T, Th1, Th2, Th17 and Treg subsets were analyzed by flow cytometry (FCM) for all participants. Among these patients, 183 cases of BD patients were treated with immunoregulatory combination drugs (IMiDs) such as low-dose interleukin-2, rapamycin, metformin, retinoic acid and coenzyme Q10. The levels of peripheral lymphocyte subsets were measured before and after the treatment. Compared-T test was used to compare continuous measures and to assess effect of these drugs.

Results: Compared to HCs, the absolute numbers of various Teffs such as T, B, CD4⁺ T, CD8⁺ T, Th1 and Th17 cells were significantly increased in BD group (P<0.01), while the level of Tregs in patients with BD was severely decreased (P<0.05), resulting in increased ratios (imbalance) of Th1/Tregs, Th2/Tregs and Th17/Tregs (P<0.05) (Figure 1). After the IMiDs treatment, the levels of NK, CD4⁺ T, CD8⁺ T, Th1, Th17 cells as well as Tregs were significantly increased (P<0.05). But the increased Tregs was much more dramatical than those of Teffs, resulting in a decrease in ratios of Teffs/Tregs such as Th2/Tregs (P<0.001) (Figure 2).

Conclusion: Impaired balance of pro- and anti-inflammatory immune cells, especially insufficiency of Tregs, might be a cornerstone of the pathogenesis of BD. Immunoregulatory combination therapies could promote the proliferation and functional recovery of Tregs in patients with BD and might help to alleviate disease activity.

References:

- [1] Yazici H, Seyahi E, Hatemi G, et al. Behcet syndrome: a contemporary view. *Nat Rev Rheumatol* 2018;14(2):107-19. doi: 10.1038/nrrheum.2017.208 [published Online First: 2018/01/04]
- [2] Rosenzweig M, Lorenzon R, Cacoub P, et al. Immunological and clinical effects of low-dose interleukin-2 across 11 autoimmune diseases in a single, open clinical trial. *Ann Rheum Dis* 2019;78(2):209-17. doi: 10.1136/annrheumdis-2018-214229 [published Online First: 2018/11/26]

Figure 1: Comparison of lymphocyte subsets in peripheral blood between patients with BD (n=384) and HCs (n=206). All lymphocytes subsets were analyzed by detected by flow cytometry combined with standard absolute counting beads. Data were presented as mean±SD and statistical analysis was determined by two-tailed unpaired t-test. Patients had lower levels of NK, Th2 and Tregs in BD compared with those of HCs. HC: healthy control. * $P < 0.05$, ** $P < 0.01$, *** $P < 0.001$.

Figure 2: Changes of peripheral lymphocyte subsets after immunoregulatory combination treatment (n=183). Data were presented as mean±SD and statistical analysis was determined by two-tailed unpaired t-test. Absolute numbers of NK, CD4+T, CD8+T, Th1, Th2 and Tregs in BD patients were significantly increased after immunoregulatory combination treatment. * $P < 0.05$, ** $P < 0.01$, *** $P < 0.001$.

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AB0500

CLINICAL CHARACTERISTICS AND POTENTIAL BIOMARKERS FOR DISEASE ACTIVITY OF PATIENTS WITH ANCA ASSOCIATED VASCULITIS: A MONOCENTER STUDY IN CHINA

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Background: Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) are a group of multisystem, autoimmune, inflammatory disease characterized by pauci- necrotizing vasculitis affecting small blood vessels. The clinical manifestations of the AAV are diverse and can be confined to one organ, or multiple organs and even life-threatening. However, there has been no specific index for assessing the activity of AAV at diagnosis.

Objectives: The aim of this study was to describe the clinical and serological features of patients with antineutrophil cytoplasmic antibody (ANCA)-associated

vasculitis (AAV) in eastern China using data from a hospital-based study. And looking for indicators that can predict disease activity.

Methods: We retrospectively studied patients with newly diagnosed AAV evaluated from January 1, 2009, to December 31, 2018. In total, 219 patients diagnosed were classified according to the American College of Rheumatology classification criteria and/or revised Chapel Hill 2012 definitions, and their clinical and serological features were evaluated. The association of laboratory data with disease activity was assessed via regression models.

Results: Of 219 incident cases of AAV, 37/219 (16.9%) had granulomatosis with polyangiitis (GPA), 172/219 (78.5%) were microscopic polyangiitis (MPA), and 10/219 (4.6%) had eosinophilic granulomatosis with polyangiitis (EGPA). The mean age at diagnosis of patients with GPA were 51.5 years MPA were 61.7 years, and EGPA were 49.8 years, respectively. Patients with MPA were significantly older than GPA and EGPA at diagnosis ($p < 0.001$). ANCA tested positive in 207 (94.5%) of cases: 167 (80.7%) were MPO-ANCA and 40 (19.3%) were PR3-ANCA. Lung, skin, nervous system symptoms were the most common in EGPA. For GPA, ear-nose-throat (ENT) symptoms and lungs involvement were the most common. Renal and lung involvement occurs most frequently in MPA. In the multivariable logistic regression analysis, higher anti-MPO antibody (149.4 IU/ml), higher hypersensitive c-reactive protein (hs-CRP, 62.5 mg/L), lower hemoglobin (113.5g/L), and higher complement 4 (C4, >0.215 g/L) were proved to be independent risk factors for active disease. Further research showed that C4 had higher sensitivity (70.0%) and specificity (83.4%) than the other three indicators.

Conclusion: MPO-ANCA-positive MPA is the most common form of AAV in Chinese patients. Serum C4 concentrations at diagnosis might be a useful biomarker of disease activity in AAV.

References:

- Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, et al. 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis Rheum* 2013, 65:1-11.
- Choi H, Kim Y, Jung SM, Song JJ, Park Y-B, Lee S-W. Low serum complement 3 level is associated with severe ANCA-associated vasculitis at diagnosis. *Clinical and Experimental Nephrology* 2018, 23:223-230.
- Leavitt RY, Fauci AS, Bloch DA, Michel BA, Hunder GG, Arend WP, et al. The American College of Rheumatology 1990 criteria for the classification of Wegener's granulomatosis. *Arthritis Rheum* 1990, 33:1101-1107.
- Masi AT, Hunder GG, Lie JT, Michel BA, Bloch DA, Arend WP, et al. The American College of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). *Arthritis Rheum* 1990, 33:1094-1100.
- Mukhtyar C, Lee R, Brown D, Carruthers D, Dasgupta B, Dubey S, et al. Modification and validation of the Birmingham Vasculitis Activity Score (version 3). *Ann Rheum Dis* 2009, 68:1827-1832.
- Markiewski MM, Lambris JD. The role of complement in inflammatory diseases from behind the scenes into the spotlight. *Am J Pathol* 2007, 171:715-727.

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