Background: ANCA-associated vasculitis (AAV) is an autoimmune disease that involves abnormal death of neutrophils and leads to necrotic inflammatory reactions in blood vessels, including microscopic polyangitis (MPA), Granulomatous polyangitis (GPA) and Eosinophilic granulomatous polyangitis (EGPA). AAV is mainly involved in small blood vessels, and intermediate arterial lesions can also occur, but the large arteries and their primary branches are rarely involved.

Objective: To summarize the clinical characteristics of aortic involvement in patients with ANCA-associated vasculitis (AAV).

Methods: The clinical manifestations, systemic involvement, laboratory examination, imaging characteristics and treatment of aortic involvement in AAV patients admitted to Peking Union Medical College Hospital from January 2013 to December 2018 were retrospectively analyzed.

Results: Nine patients were enrolled in our study. The ratio of male to female was 2:1 and the median age was 47 years old. Of the 9 patients, 4 were MPA (44%), 4 were GPA (44%) and 1 was EGPA (11%). The aorta is involved in an average of 3 locations per case, mainly in 7 locations: 3 ascending aorta and aortic arch, 4 in the head and arm trunk (including carotid and subclavian artery), 2 in the abdominal aorta, and 1 in the abdominal cavity. There were 2 cases of renal artery, 1 case involving brachial radial artery, 2 cases of iliac artery and lower limb artery, and 1 case involving left main coronary artery, anterior descending branch, circumflex branch, and right coronary artery. Aortic lesions: 3 cases had atherosclerosis, 3 cases had arterial stenosis and / or occlusion, and 1 case had periarteritis. When major arterial involvement was found, the AAV of the patients were mostly active, with an average of 19 points for BVAS vasculitis activity and 1 for FFS score. 6 cases had lung involvement (67%), 6 cases had kidney involvement (67%), 4 cases had ENT involvement (44%), 3 cases had nervous system involvement (33%), and 1 case had gastrointestinal involvement (11%). All patients were treated by steroid and immunosuppressant, while 1 case received the operation of ascending aorta and aortic arch replacement.

Conclusion: Mainly involved in small vessel inflammation, AAV may also have aorta involvement, which was more common in patients who had active disease and need more positive treatment. The affected aorta areas of these patients were mainly ascending aorta, aortic arch, and head and arm trunk, which can be manifested as aneurysms, dissections, and arterial stenosis Periarteritis, etc. If necessary, surgically treatment of the affected aorta could be considered when the situation of AAV was stable enough.

References:

Disclosure of Interests: None declared

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AB0541

IMPROVED SURVIVAL IN PATIENTS WITH GIANT CELL ARTERITIS: A POPULATION-BASED COHORT STUDY

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Background: In previous studies patients with giant cell arteritis (GCA) have had survival rates that are similar to or better than the general population.

Objectives: To investigate survival trends and cause-specific mortality in patients diagnosed with GCA over a 60-year period.

Methods: We assembled a population-based incidence cohort of patients with GCA diagnosed between 1950 and 2009. All patients were included if they met the American College of Rheumatology (ACR) 1990 Criteria for the Classification of GCA. Patients diagnosed between 2000 and 2009 could also be included if they met the following criteria: age greater than or equal to 50 years, elevated inflammatory markers, and radiographic evidence of large-vessel vasculitis attributed to GCA. A non-GCA comparison cohort was assembled from the same underlying population for each patient with GCA. Patients were followed until death, last contact, or December 31st, 2018. Survival trends were analyzed by grouping patients into the following categories according to year of GCA diagnosis: Group A 1950-1979; Group B 1980-1989; Group C 1990-1999; and Group D 2000-2009. Mortality rates were estimated using Kaplan-Meier method and were compared with expected mortality rates for persons of the same age, sex, and calendar year, as estimated by regional population life tables. Cause-specific mortality was obtained from death certificates for patients in both cohorts. The causes were grouped according to ICD-9 chapters and hazard ratios were estimated against the non-GCA comparators.

Results: The study population included 245 incident cases of GCA: 194 (78%) women and 51 (21%) men with mean age (±SD) of 76.2 (±8.3) years and median follow-up of 10.6 years. There was no overall difference in survival between the GCA cohort and the general population. The 2-, 5-, and 10-year survival rates (95% CI) were 89% (86, 93), 76% (70, 81), and 56% (50, 63) respectively with a standardized mortality ratio of 0.99 (0.86, 1.14). The standardized mortality ratios for Groups A, B, C, and D were 0.83 (0.57, 1.17), 0.92 (0.63, 1.3), 1.21 (0.85, 1.69), 0.70 (0.50, 0.96), respectively. The overall all-cause mortality adjusted for age, sex, and calendar year was similar between the GCA patients and their comparators with a hazard ratio of 1.03 (0.84, 1.24). Mortality due to neoplasms was significantly lower in the GCA cohort with a hazard ratio of 0.53 (0.3, 0.92). Other cause-specific mortalities were not significantly different between the groups.

Conclusion: In this population-based cohort of patients with GCA diagnosed over a 60-year period, the survival of patients diagnosed in recent years was significantly better than that of the general population. The explanation for this novel finding is unclear, but likely to be multifactorial. In this study the number of deaths due to neoplasms in the GCA group was significantly lower.

References:

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AB0542

REATIONS TO PNEUMOCOCCAL 13-VALENT VACCINE IN PATIENTS WITH BEHÇET SYNDROME

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Background: The European League Against Rheumatism (EULAR) recommends pneumococcal 13-valent (PCV13) and 23-valent vaccines in patients with rheumatic diseases (1). Adverse reactions to 23-valent pneumococcal vaccine were previously reported in patients with Behçet Syndrome (BS) (2). These were proposed to be associated with the pathergy phenomenon which may be observed in patients with BS.