**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 dilution and/or dissection, 6 cases had arterial lesions also occur, but the large arteries and their primary branches are rarely involved. The objectives are 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 31, 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 the 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 (79%) women and 51 (21%) men with mean age of (±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, 1.35), 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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**Sclerodema, myositis and related syndromes**

**AB0543**

**CLINICAL CHARACTERISTICS OF A GROUP OF CHRONIC GRAFT-VERSUS-HOST DISEASE PATIENTS WITH POSITIVE AUTOIMMUNITY.**

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**Background:** Graft-versus-host disease (GVHD) is a commonly severe multiorgan complication in patients undergoing allogeneic transplantation of hematopoietic progenitors. Its chronic form reflects a complex immune response with different degrees of inflammation, immune dysregulation and fibrosis. In some chronic graft-versus-host disease (cGVHD) patients, positive antibodies have been detected, which represent the presence of immune activity and suggest the possible involvement of B lymphocytes in the disease etiopathogenesis, but their clinical utility is controversial.

**Objectives:** To describe the clinical characteristics of a group of cGVHD patients with positive autoimmunity treated in a multidisciplinary consultation of Rheumatology-Dermatology-Hematology of GVHD.

**Methods:** Observational and retrospective study to describe the clinical characteristics of the patients with positive autoimmunity collected in the database of the multidisciplinary consultation of GVHD. The variables reviewed for this study, in addition to the demographic ones, were type of antibody, disease causing the transplant, presentation, severity and type of involvement. The statistical analysis was done with Epi-info 7.2.2.6.

**Results:** Only 16 (16%) of the 100 patients included in the database had positive autoimmunity. Twelve (75%) tested positive to ANA, although 5 (31.25%) in a lower titer (1/80). The most common immunofluorescence pattern was the nuclear (88.89%), 66.66% nucleolar and 22.22% nucleolar + cytoplasmic. Other antibodies detected were: 6 anti-Ro52, 2 anti-dsDNA, 1 anti-RP155, 1 anti-Fibrillarin, 1 anti-SAE1, 1 p-ANCA and 1 anti-NOR-90. The mean of age was 51.31±14.03 years. As for sex 4 (25%) were female and 12 (75%) were men. The most frequent disease that caused the transplant was acute myeloid leukemia (58.3%). Ten (62.5%) patients presented de novo cGVHD, 1 (6.25%) progressive and 5 (31.25%) quiescent. The time since receiving the transplant until the first visit was 14 to 79 months. Ten (62.5%) patients had nonspecific symptoms (arthralgia and myalgia), 2 (12.5%) edema, 8 (50%) contractures, 5 (31.25%) fascitis and 6 (37.5%) esophagitis. Eight (50%) patients had ocular involvement and 6 (37.5%) of the oral mucosa in the form of dry syndrome (Sjögren-like syndrome). Ten (62.5%) patients had limitation of joint mobility detected by the range of motion scale (ROM), of which 6 were mild and 4 moderate. Only 5 (31.25%) patients had general condition impairment. As for the skin involvement 10 (62.5%) patients had sclerodermiform involvement (8 of them being eosinophilic fasciitis-like), 2 (12.5%) lichenoid, and 3 (18.5%) mixed (sclerodermiform + lichenoid). Only 1 patient didn’t meet diagnostic criteria for GVHD. The sclerodermiform was the most common type of involvement in the positive ANA patients. Regarding the severity according to the of the American National Institute of Health (NIH) classification: 8 (50%) had severe affection, 5 (31.25%) moderate and 2 (12.5%) mild, with 4 (25%) exuits.

**Conclusion:** In our cohort of patients with cGVHD, serum detection of autoantibodies is uncommon, being the ANA with nuclear pattern the most frequent. Although the small sample size does not allow correlations with the clinical variables it’s worth highlighting a greater positivity of autoantibodies in the scleroderma skin forms.

**References:**


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