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Letter

Prevalence of Takayasu Arteritis: A Population-based Study

To the Editor:

Takayasu arteritis (TA) is a rare inflammatory condition of the large blood vessels that affects the aorta and its branches. Young females of Asian descent are typically the most affected by this disease; however, in the United States, most patients with TA are White[1,2,3]. Data regarding the epidemiology of TA is limited, with most studies of the incidence and prevalence of the disease coming from Japan, where TA is more prevalent.

The described incidence rate of TA ranges from 0.3 to 3.3 per million per year and the prevalence ranges from 4.7 to 360 cases per million[4]. Only 2 studies have evaluated the epidemiology of TA in the US; one of them demonstrated an incidence rate of 2.6 cases per million per year in Olmsted County, Minnesota[5], and the other study described a prevalence of 0.9 cases per million in New York, which is the lowest prevalence reported to date[6].

The present study was performed with the objective of estimating the epidemiology of Takayasu arteritis in Olmsted and 26 surrounding counties. The study was conducted using the extended Rochester Epidemiology Project (REP), which was developed in 2010[7,8]. The protocol was approved by the Mayo Clinic and Olmsted Medical Center institutional review boards (IRB #09-004685 and IRB #025-OMC-19, respectively).

Records from patients residing in Olmsted and surrounding 26 counties and with a diagnosis of TA between January 1, 2010, and December 31, 2018, were individually reviewed to confirm the diagnosis of TA. Patients who met the 1990 American College of Rheumatology (ACR) criteria for TA were included. Patients who denied use of their medical records for research were excluded. Point prevalence was calculated on January 1, 2015, using the number of cases as the numerator and population counts from the REP census as the denominator. The adult (age ≥ 18 yrs) population of the 27-county region covered by the extended REP in 2015 was 585,000. Prevalence rates were age- and sex-adjusted to the US White 2010 population. CI were computed by assuming that the number of prevalent cases followed a Poisson distribution.

Between January 1, 2010 and December 31, 2018, forty patients were identified on initial screen by diagnosis code and underwent comprehensive medical record review. Patients with noninflammatory vascular disease and those with vasculitis not meeting criteria for TA were excluded; additionally, only patients who were residents at the time of diagnosis or on January 1, 2015, were included in the study. One incident case and 5 prevalent cases of TA meeting ACR criteria were included in the study.

As there was 1 incident case, the incidence rate could not be determined. With the 5 prevalent cases, a point prevalence was calculated on January 1, 2015, establishing a prevalence of TA of 8.4 per million population (95% CI 1–15.8) in this group. The prevalence was higher in females (13 per million population; 95% CI 0.2–26.2) compared with male patients (3.6 per million population; 95% CI 0–10.7).

The prevalent cases included 4 females and 1 male. Four patients were White and 1 Asian. The mean age at diagnosis was 20.5 years (SD 4.5 yrs). Four patients had a vascular distribution of involvement classified as Hata type V and 1 as Hata type I. As part of their treatment, all the patients received glucocorticoids, 2 methotrexate, 2 infliximab, 2 adalimumab, and 1 azathioprine at any given time. One patient underwent 4 separate vascular interventions, which included right subclavian bypass, bilateral carotid endartectomies, abdominal aortic aneurysm repair, and left iliac stenting. No restenosis or occlusion of the intervened sites was observed at last follow-up. The remaining 4 patients did not require vascular or endovascular procedures.

In the present population-based study conducted in Midwestern USA, the estimated prevalence of TA was found to be 8.4 per million population. This is similar to the findings of other epidemiology studies outside of Asia that report a prevalence in the range of 4.7–25.2 per million population[4]. Additionally, we found a higher prevalence in female patients, consistent with the reported female predominance of this disease. The majority of patients in our study were White, reflecting the demographic distribution of residents of Olmsted and surrounding counties.

The strengths of our study include the extended REP, which is a unique record linkage system in Southern Minnesota and Western Wisconsin, allowing full access to the medical records of the studied patients. The limitation of our study, inherent to a retrospective study design, is that the information is entirely dependent on what is reported in the medical records.

Understanding the epidemiology of TA is essential for the advancement of research in this challenging disease.

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The authors declare no conflicts of interest.

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