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Chapter

Epidemiological Aspects of Giant Cell Arteritis

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Abstract

Giant cell arteritis (GCA) is a systemic vasculitis that affects medium-to-large-sized arteries, in which the inflammatory reaction destroys the artery wall with the fragmentation of the elastic lamina. Such phenomena can result in vision loss if not treated promptly. Other nonocular symptoms noted include GCA, headache, tenderness in the temporal area of the scalp, myalgias and arthralgias, fever, weight loss, and jaw claudication. Clinical suspicion is an essential pathway to the diagnosis of this disease. Thus, immediate Westergren sedimentation rate and C-reactive protein should be obtained. A temporal artery biopsy, however, remains the most definitive diagnostic tool. The incidence of GCA remarkably increases with each decade of age among those aged 50 years or over. Additionally, there have been notable differences among patients of different ethnicities. The epidemiological characteristics of GCA have been primarily researched in populations from the United States as well as several European countries with emphasis on the Caucasian population. In more recent years, a handful of studies have emerged from non-European countries regarding the epidemiology of GCA. The results of these findings are in parallel with previous observations, which presumed GCA to be more common in European and North American populations.

Keywords: giant cell arteritis, epidemiology, visual loss, temporal artery biopsy, incidence rate

1. Introduction

The first account of giant cell arteritis (GCA) can be traced back to tenth-century Baghdad by Arab ophthalmologist of medieval Islam Ali ibn Isa al-Kahhal. It was then more precisely described by Sir Jonathan Hutchinson in 1890, who noted the peculiar thrombotic appearance of the temporal artery (TA), a defining feature of the course of GCA. With the progression of the discovery of this disease and various case studies exploring its nature, ophthalmologists have additionally attempted to view how GCA could affect certain populations. During the second half of the twentieth century and the course of the twenty-first century, facilities across continents have published their findings on the tendencies of GCA to affect certain individuals more than others. In this chapter, we describe the epidemiology of GCA across continents and countries from individual reports and studies presenting the incidence rate of this vasculitis in their respective locations or populations.
2. Europe

Europe remains the continent with the most abundant publications pertaining to the epidemiology of GCA. One of the longest studies on the epidemiology of GCA was conducted in Western Norway, a retrospective study encompassing cases from 1972 to 2012 [1]. This study was one among many that noted how changing the criteria for the identification of GCA could greatly alter its incidence, especially due to the rarity of this vasculitis. For instance, the incidence rate of patients potentially affected by GCA satisfying the ACR 1990 criteria was 16.7 per 100,000 persons over 50 years of age. The incidence of patients clinically diagnosed as having GCA was 18.4 per 100,000 persons aged 50 years or more. Meanwhile, the incidence of biopsy-proven GCA was 11.2 per 100,000 persons over 50 years of age. The extended period of this study additionally contributed to the lowering of the mean annual incidence. When solely evaluating cases during a certain 5-year period (from 1992 to 1996), the incidence rate was found to be 26.7 per 100,000 persons over 50 years of age. Regardless of this, the prevalence remained among the highest recorded globally. This study also describes the increasing GCA incidence with age, where an older individual has a greater susceptibility to acquiring this vasculitis, as well as a greater ratio of women having this disease. Similarly, in the southern region of Norway, comparisons with past records from the same set of population were provided [2]. In a study spanning a period of 14 years (2000–2013), the Hospital of Southern Norway presented a GCA occurrence rate of 16.8 per 100,000 persons over the age of 50 years, one of the highest recorded globally and in line with other epidemiological findings from the Scandinavian region. As previously noted, one must consider that a small study sample and a short study period both have the potential of overestimating the incidence of a disease, as demonstrated by this study. In the same vicinity, the rate of giant cell arteritis in Western Nyland, Finland was examined [3]. From 1984 and 1988, 54 patients were diagnosed with GCA, among which 16 patients had a positive biopsy. The retrospective annual incidence of GCA was 69.8 for every 100,000 individuals older than 50 years. From 1984 to 1990, 133 patients in Iceland fulfilling the ACR criteria for GCA were identified, rendering an incidence rate of 27 in 100,000 people older than 50 years [4]. This study also suggests that the clinicians’ greater tendency to suspect GCA and perform TA biopsies (TABs) may have contributed to a higher statistical incidence. The results of these studies and their reported high mean annual incidence rates go on to highlight the possibility of a greater susceptibility to GCA among Scandinavian population. These are among the highest globally, supporting the claim that the Scandinavian population is most considerably afflicted by this inflammatory disease.

When gauging the incidence of GCA cases in other parts of Europe, we witness a lowering in the number of cases. For instance, in Italy, 285 cases of biopsy-proven GCA were observed in the Reggio Emilia area from 1986 to 2012 [5]. The adjusted incidence rate was 5.8 per 100,000 people older than 50 years of age and was significantly greater in women. In Lugo, Spain, a retrospective study was conducted from 1986 to 1995 to identify the occurrence rate of biopsy-proven GCA [6]. The mean annual incidence was computed for each 5-year period, rendering a rate of 8.26 and 10.49 per 100,000 people older than 50 years, respectively. Nearby countries and regions in the southern part of Europe presented similar incidence rates, demonstrating the moderate tendency of individuals from this area of the continent of being acquiring GCA. Namely, the incidence rate in France was concluded to be between 7 and 10 individuals out of 100,000 older than 50 years [7]. Likewise, in Slovenia, the estimated annual incidence rates of GCA were overall 8.7 per 100,000 aged greater than 50 years. This lowered rate suggests a different ethnical make
up in the region that is perhaps less susceptible to acquiring this vasculitis, suggesting a genetic factor, while the geographical location in a lower latitude than the Scandinavian region may imply an environmental etiology. The exact etiology remains unknown.

Epidemiological studies from 2002 to 2008 in Southern Europe and Northwestern Turkey aimed to assess the epidemiology of GCA by following patients at Trakya University Medical Faculty [8]. During this period, the incidence of GCA was found to be 1.13 patients per 100,000 persons 50 years of age or older. The incidence of GCA for females was slightly greater than that for males. The fact that this study relied on a single center presents the possibility of missing individuals who sought care in a different location or simply neglected their condition. Regardless of this, the contribution of this report is crucial due to the paucity of epidemiological outlook on GCA in this space.

3. Asia

A retrospective study of patients with giant cell arteritis in China was performed from August 1992 to May 2014 at the Peking Union Medical College Hospital [9]. A total of 70 patients were diagnosed with GCA. The demographic data of these patients differed from that in the previously discussed epidemiological studies in Europe. First, the average age of Chinese GCA patients was 65.2 years. This age at onset is lower than the mean reported age in other populations, which hovered between 70 and 80 years. In addition, male patients with GCA predominated the study, which differed from most reports globally. Chinese male may be more susceptible to GCA than female or they may present greater health-seeking behavior. It is important to note that patients in this study were identified from a single healthcare center, which may substantially underestimate the occurrence of this vasculitis despite its current rare occurrence. On a similar note, statistical records, pathology records, and case records from university hospitals were gathered to estimate an annual incidence of one out of 100,000 people aged older than 50 years in Hong Kong [10]. These findings suggest the particularly lower frequency of GCA among the Chinese population. In 1998, a nationwide survey was performed in Japan, revealing 690 patients treated for GCA in the previous year [11]. An incidence rate of the population was calculated to be 1.47 per 100,000 people older than 50 years of age. In conclusion, the epidemiological reports of GCA from East Asian countries reveal extremely low prevalence of GCA among this population.

From 2008 to 2014, a total of 17 patients fulfilling the classification criteria for GCA in India were identified [12]. Comparably to a previously discussed study in China, the mean age of GCA patients in the Indian population was 67 years, lower than the mean age from European reports. In addition, individuals with GCA in India were predominately male. The reasoning behind a lower mean age and a male predominant patient status is unknown and was hypothesized to be due to the greater likelihood of individuals with these characteristics to seek healthcare.

The rarity of GCA among the Indian population was demonstrated at Moorfields Eye Hospital, a center in London, UK [13]. From 2006 to 2014, patients of Indian descent were significantly less likely to have a biopsy-positive GCA. Perhaps, some ethnicities are less likely to present a positive result to the TA biopsy or clinicians may simply be more likely to diagnose these individuals with GCA. A study of this nature, in which ethnicities are compared in a population, could provide important findings on the vulnerability of certain individuals to present with this vasculitis.
4. Middle east

The true incidence of GCA in the Arab population is difficult to assess due to the absence of a more nationwide perspective as well as a lack of population-based study in Arab countries. In a 22-year study, the epidemiology of GCA in Saudi Arabia was investigated [14]. From 1983 to 2004, 102 patients at King Khaled Eye Specialist Hospital underwent TAB, as seen in Figure 1, and seven patients were identified with biopsy-proven GCA. They noted that the incidence of GCA increases with age. Regardless of this, many aspects of the healthcare system in Saudi Arabia closely resemble that of the United States, with a similar life expectancy and a ratio of ophthalmologists relative to the size of the population.

In 1980, the incidence of giant cell arteritis in Jerusalem over a 25-year period was evaluated in a study involving four general hospitals in Jerusalem [15]. Among them, 170 patients with GCA had a positive TA biopsy. Furthermore, 36 biopsy-negative cases were also considered as they fulfilled the 1990 ACR criteria for GCA classification and responded adequately to steroid therapy. The age-adjusted incidence rate was computed to be 11.3 per 100,000 people ≥50 years of age for all incorporated GCA cases, but lower at 9.5 for the biopsy-proven cases. Moreover, this study observed seasonal patterns with a statistically insignificant rise in GCA diagnosis during the summer. The incidence rate of GCA in this study is comparable with those in other Mediterranean countries, with a less prominent frequency of female patients.

The results of a cohort of 114 patients who met the 2016 rACR criteria for the diagnosis of GCA and underwent TAB over a 10-year period in a tertiary center, Rassoul Akram Hospital, in Tehran, Iran were described [16]. This finding reflects the increase in GCA incidence with age. Although this study did not sufficiently provided a macroscopic account of the incidence and manifestation of GCA in a population, it was the first study performed in Iran assessing the intricacies of characterizing GCA and the discrepancies that may arise as a result of heterogeneous studies, especially due to the absence of definite criteria for the diagnosis of GCA.

Figure 1.
A temporal artery biopsy involves acquiring a small section of the artery, which can potentially appear thrombotic. The length of the segment can vary across studies and may influence the results of the biopsy [14].
5. Africa

Africa stands out for its scarcity of information on GCA and its epidemiology. Perhaps, this could be due to an underdeveloped healthcare system, which hinders the proper equipment and tools for an adequate diagnosis, which could ultimately serve as data to be studied on a larger scale. It may also be that the African population has a lower susceptibility to GCA. In addition, the life expectancy in Africa is lower, which could influence statistics related to a disease with an increased likelihood of manifesting at a later stage in life. Therefore, it can be hypothesized that this region presents with lower rates of this vasculitis. The two studies discussing the epidemiology of GCA in the African population both pertain to French-colonized islands. From 1991 to 2016, data from two pathology units in Martinique, West Indies were reviewed to discuss the features of cases of biopsy-proven giant cell arteritis [17]. The findings fortified the assumption that GCA is less prevalent in an African descent population. Nevertheless, the retrospective nature of the study and the exclusion of a biopsy-negative GCA may have led to an underestimation of cases of GCA.

In a retrospective study from La Reunion near the Southwest region of the Indian Ocean from 2005 to 2017, an incidence rate roughly 4–12 times lower than in most European countries was calculated [18]. An exact count was difficult to provide due to the presence of a diverse group of ethnicities in La Reunion, especially from regions of the world with a lower prevalence of GCA. A shorter life expectancy may contribute to a lower frequency of cases observed as GCA increases with age. Other characteristics were found to be analogous observations made in previous epidemiological studies.

6. North America

Studies conducted in the United States have the potential of presenting important findings due to the possibility of comparing and contrasting features of a disease between ethnicities. A retrospective study spanning 11 years was conducted in the Texas Gulf Coast. Twenty-seven out of 101,239 patients aged 40 years or older had GCA. Intriguingly, 13 of these patients were black females, rendering it a noteworthy aspect of this study in which a significantly greater proportion of patients with GCA were black individuals [19].

A report from a study spanning from 1971 to 1980 in Shelby County, Tennessee identified 26 cases of GCA [20]. The average annual incidence was computed to be 1.38 per 100,000 individuals older than 50 years of age. The predominant patient from this study was white and female. This study presents one of the lowest frequencies of GCA cases across the globe. This could partially be due to the racial makeup of this population, which has a high percentage of black residents. African descent population is assumed to present a lower incidence rate of GCA. Among other contributions to a low incidence rate such as a retrospective design as well as inconsistencies in the diagnosis criteria, this study urged the need to consider environmental factors as potential causes for the onset of the vasculitis, such as the climate, exposure to the sun, frequency of rainfall, elevation, etc.

In another region of the United States, Olmsted County, Minnesota holds a population with northern European ancestry, which appears to be the group of people most severely afflicted by GCA. Therefore, the observation of a greater incidence rate may indicate a genetic factor in the onset of GCA. Between 1950 and 1991, 125 Olmsted County inhabitants were diagnosed with giant cell arteritis. The incidence per 100,000 persons 50 years of age or older was 17.8, which was significantly higher in women than in men. The incidence of GCA had increased to 19.8 from 2000 to 2009. The annual
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incidence rates substantially increased over the study period and with congregated cases of GCA, suggesting a regular cyclic pattern over time, which suggested the possibility of an infectious root for giant cell arteritis.

Previous studies suggested a low incidence of GCA in black patients, although conclusions were drawn from relatively small sample sizes. Nevertheless, the impression that GCA rarely impacts black individuals is generally assumed. Some reports have sought to compare GCA more directly between two races. A multicenter study involving 10 healthcare institutions was conducted to evaluate the presentation of GCA in African Americans [21]. An African American group of patients was compared with a cohort of Caucasian patients with a positive biopsy for ophthalmic GCA. Both the groups appeared to have a similar sex distribution, as around 70% of patients in both the cohorts were females. At Johns Hopkins Wilmer Eye Institute, findings notably challenging the commonly held belief that GCA is uncommon in African Americans were presented [22]. However, annual rates may not be directly calculated due to racial distributions in patients not reflecting that of the census population of the city of Baltimore, a detail that needs to be approached diligently prior to establishing conclusions. Furthermore, the screening and diagnosis process may differ among races and ethnicities due to physicians and clinicians holding preconceived perception of a greater prevalence in certain populations.

When comparing the rate of GCA between Caucasians and Asians, a significant lower occurrence rate of GCA in Asians was identified, which they computed to be 0.26–3.8 per 100,000 individuals older than 50 years of age, in parallel with studies from Asia [23]. The data for this study were collected from the University of California San Francisco computer database for patients from July 1989 to July 2006.

Similarly, giant cell arteritis has been reported to be very rare in Hispanics. From 1996 to 2002, patients with GCA at the Bascom Palmer Eye Institute were assessed [24]. Rates of a positive temporal artery biopsy were similar among Hispanic and non-Hispanic patients. Thirty-two patients with biopsy-proven GCA revealed similar mean age, symptoms, and final visual acuity between Hispanic and non-Hispanic cohorts. Hispanic and non-Hispanic cases are similarly impacted by the onset of giant cell arteritis.

A retrospective review was performed of all the biopsy-positive cases of giant cell arteritis presenting to a neuro-ophthalmology practice in Saskatoon, Saskatchewan [2]. Records of 141 consecutive patients who underwent temporal artery biopsy at the Saskatoon Eye Centre from July 1998 to June 2003 were reviewed. The average age of the biopsy-positive patients was 76.5 years, and the patients were 2.4 times more likely to be women. A total of 35 patients had a European ancestry, while two patients were of Aboriginal descent. The estimated incidence of GCA for Saskatoon was 9.4 per 100,000 for people over the age of 50 years. This study reveals the prospect of GCA to affect the people of Aboriginal descent despite a probable low incidence rate.

7. South America

Very few studies pertaining to the epidemiology of GCA have come from South American countries. One that most closely attempted to depict the status of GCA nationwide collected findings from three university hospitals in Brazil for patients with GCA between 2009 and 2010 [25]. This was, in fact, the first study addressing the features of GCA in Brazilian patients having the disease. Most GCA patients were Caucasians, while a few were of a combined European and Indigenous lineage. The Caucasian cohort was mostly of Portuguese, Italian, or Spanish ancestry. These suggested the possibility of asymptomatic manifestations, which may skew the epidemiological perspective of this disease.
8. Oceania

The last geographic region to be discussed is Oceania, which can be hypothesized to most closely resemble findings from Europe. From 1992 to July 2011, 314 cases of biopsy-proven GCA in South Australia were studied, in which the incidence for people over the age of 50 was 3.2 per 100,000 individuals [26]. Most characteristics of the disease were in line with observations described in studies from Europe, including a similar mean age and female predominance. Seasonal variations were additionally perceived, with a greater amount of diagnosis occurring during the summer season.

Cyclical variations were similarly noted in a study conducted in Otago, New Zealand. Records of 363 consecutive patients who underwent temporal artery biopsy at Dunedin Hospital between 1996 and 2005 were reviewed, with biopsy-proven GCA diagnosed in 70 patients. The mean annual incidence of GCA in Otago for people older than 50 years was 12.73 per 100,000 persons ≥50 years of age [26].

9. Conclusion

Nordic countries present the highest annual incidence rates of GCA. This vasculitis moderately affects southern European countries (Italy, Spain, France, etc.). The lowest incidence rates have been reported in East Asia. The diverse ethnical populations in countries such as United States lead to variations across regions, such as a higher incidence rate in the Northern states due to Scandinavian ancestry. Different ethnicities may present varying susceptibility because clinicians may exhibit different degree of suspicion with certain races, leading to influence on the number of biopsies performed and diagnosis made. In some regions, race and ethnicity is self-identified, which may reveal limited information on genetic background. Figure 2 reveals the varied incidence rate observed in different populations across the globe.

The incidence rate increases substantially with age and a greater ratio of patients are women in most regions, except for Asian countries. Whether female susceptibility is genuinely lower in that region or whether this discrepancy is due to different health-seeking behavior is unknown. Although seasonal and cyclic patterns were observed in a few studied and environmental factors were suggested, such influence remains inconclusive.

Figure 2. Graphical representation of incidence rates of GCA among some of the populations described in the literature. The highest incidence rates appear to be among the Scandinavian countries, regardless of the criteria utilized to diagnose the incidence of GCA.
The definition of giant cell arteritis is inconsistent across literature, resulting in the inclusion of heterogeneous data during extensive review. Hence, there may be an over- or underestimation of statistical values. The criteria for the diagnosis of this disease substantially varied, with incidence rates presented based on biopsy-proven cases, ACR-criteria-fulfilling cases, or unspecified clinical diagnosis. Therefore, data may vary depending on which inclusion criteria were used.

Moreover, the technicality for biopsy-proven cases (length of the segment or threshold for diagnosis) may also alter the rate of incidence. In many reviews, the length of the arterial specimen remains unmentioned.

In 2016, an alteration to the list of criteria for a more comprehensive diagnosis of GCA was submitted. Furthermore, additional diagnostic tools have recently emerged, including the color Doppler ultrasound (CDUS), despite requiring extensive experience for utilization and a proper diagnosis. Other high-resolution magnetic resonance imaging technologies include magnetic resonance angiography (MRA), positron emission tomography (PET), computed tomography (CT), CT with angiography, and conventional MRA, which alternatively permit the visualization of the temporal artery. Although most reports attempted to thoroughly describe the equipment and tools for diagnosis, the heterogeneous approach across studies hinders appropriate comparisons, which may limit a precise epidemiological outlook of the disease in question.

Although this study repeatedly describes the rarity of GCA, it remains the most common vasculitis with severe consequences if remained untreated, ultimately resulting in permanent visual loss. Therefore, clinicians should remain diligent when coming across individuals presenting symptoms of the disease because an immediate course of action may greatly influence a person’s course of life and impact their well-being physiologically and psychologically.
References


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