Epidemiology of Takayasu's arteritis in Turkey

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ABSTRACT

Objective. *To estimate the prevalence and incidence of Takayasu arteritis (TA) among the residents of the city of Izmir, the third largest metropolis in Turkey.*

Methods. *Five tertiary care teaching hospitals, which were the only ones that provided rheumatology specialty care during the study period in the city of Izmir from 2006 through 2010, were invited to take part in the present study. A case search was performed electronically in the information systems of these hospitals using The International Classification of Diseases Tenth Revision (ICD-10) code for Takayasu arteritis (M31.4). The diagnosis was confirmed through chart review by a rheumatologist according to the 1990 American College of Rheumatology (ACR) criteria. Annual prevalence was calculated based on the number of patients that were alive at the end of 2010. Age- and sex-adjusted prevalence rates were standardised according to the 2010 Turkish population, based on 2010 Turkish Census.*

Results. *A total of 41 patients were confirmed to have TA and also to live within the targeted area. The annual prevalence was estimated as 12.8 (95% CI 12.0–13.6) per million; 23.5/million (95% CI 21.9–25.0) in females and 1.9/million (95% CI 1.5–2.4) in males. The prevalence was higher 8.8/million (95% CI 7.7–10.0) in the population >40 years of age. During the study period, the mean annual incidence of TA was estimated as 1.11/million (95% CI 0.54–1.67).*

Conclusion. *The first epidemiologic study of TA in a Turkish population suggests that TA is a relatively common vasculitis in Turkey.*

Introduction

Takayasu's arteritis (TA), which is also known as "pulseless disease," "aortic arch syndrome" or "occlusive thromboarthropathy" is a rare, systemic, chronic, granulomatous large-vessel vasculitis of unknown aetiology, affecting large elastic arteries, especially the aorta and its main branches (1). Several sets of classification criteria have been proposed over the years (2-5); however, the American College of Rheumatology (ACR) 1990 criteria for the classification of TA are most widely used (2). TA is generally thought to affect young people less than forty years of age; however, newly diagnosed patients aged from their 20s to mid-60s have been reported. The clinical picture varies based on the artery involved and on the severity of inflammation. TA tends to be more insidious at onset or even completely asymptomatic; however, presentation with atypical and/or catastrophic disease may also be seen. Since there are no specific diagnostic blood or urine laboratory tests or autoantibody positivity, many patients experience considerable delay in their diagnosis (1).

The relative rarity of TA is a barrier to collecting large numbers of patients and to performing detailed studies. Clinical and epidemiological features of TA show variations among different regions, such as Asia (6-8), Europe (9-11), and North America (12-15). More patients with TA have been reported from Asian countries, especially from Japan (16-19), compared to Europe (10, 20-24) or the United States of America (25). Therefore, the exact incidence and prevalence of TA are unknown in Europe and North America. However, it is believed that TA may be more frequent than that was previously thought and it may be seen in all ethnic groups. Moreover, female-to-male ratio in TA shows geographic differences; female predominance is observed in Japan, Korea, and China, but not in India. Onset and progression of the disease also show regional differences. The disease primarily involves the aortic arch and extends through the abdominal aorta in

Japanese patients, whereas the abdominal aorta is primarily involved in Indian patients (9-11).

Clinical observations have suggested that TA, which is more prevalent in Japan, India, and other Eastern Asian countries, is also a relatively prevalent vasculitis in Turkey. Although patient characteristics of TA have been previously reported in a number of studies in Turkish population (26-28), there are no data regarding the prevalence or incidence of the disease in our country. The present study, which is the first epidemiological TA study in Turkey, aimed to determine the prevalence and incidence rates of TA and to evaluate patient characteristics using medical records of university hospitals and training and research hospitals in central Izmir, the third largest city in Turkey. Izmir province is located on the west coast of Turkey and is the gateway to the Aegean Region.

Materials and methods

Study population

Izmir is the third largest city in Turkey; it is divided into 30 districts. Eleven of them (Cigli, Karsıyaka, Bayraklı, Bornova, Buca, Gaziemir, Karabaglar, Konak, Balcova, Narlidere, and Guzelbahce) are central districts, while the others are satellite districts and classified as suburban or rural communities of Izmir. The census data of central Izmir, including the distribution of population by age groups and their gender ratios, were obtained from Izmir Regional Office of the Turkish Statistical Institute. In 2010, the whole population of Izmir was 3,965,232. Nearly 70% of this population (2,786,863) resided in central Izmir, of whom 62% (1,729,149) were below 40 years of age.

Only university or state hospitals having rheumatology in-patient and/or out-patient departments with available rheumatologists were targeted for inclusion in this study and all of them agreed to participate. Under these specifications, the Rheumatology Departments of Dokuz Eylül University Medical Faculty, Ege University Medical Faculty, Izmir Atatürk Training and Research Hospital, Bozyaka Training and Research Hospital, and Tepecik Training and Re-

search Hospital were included. Pediatric departments of these hospitals were not included in our case finding strategy. According to address records at the time of diagnosis, patients who were not residents of central Izmir were not included in the analysis.

Since follow-up and treatment of patients with TA are complicated, these patients are generally referred to rheumatologists at tertiary health care centres. In central Izmir, currently, there is no other tertiary referral centre for patients with TA aside from those five centres included in the present study. We did not include the population living in the satellite districts of Izmir. This was mostly due to the high possibility of referral of patients from the satellite districts to neighbouring city hospitals. In other words, patients with TA living in the satellite districts of Izmir are more likely to seek medical care in hospitals of neighboring cities, which are closer to their places of residence, instead of traveling longer distances to obtain medical care in central Izmir.

Subject evaluation

As there is no national registry for TA in Turkey, principally clinical coding systems for electronic medical records (EMR) of referral hospitals were used for calculating incidence and prevalence rates. With this regard, two independent strategies were applied to identify TA patients in those centres. Initially, we reviewed all of the hospital contact records between January 1, 2006 and December 31, 2010. The EMR systems were integrated widely and have been effectively used in Turkish hospitals. The disease codes used in the EMR of the hospitals were consistent with the 10th Revision of the International Statistical Classification of Diseases coding system (ICD-10), and the code of M31.4 was used for searching candidate patients with TA. Additionally, the patients were also detected based on the information obtained from the direct medical records of rheumatologists in each hospital where the authors were employed.

At the second stage, all of the patients coded as M31.4 were re-evaluated to

confirm the given diagnosis. Only those patients who met the ACR criteria for TA (2) were included in the study. Demographic and clinical information of the patients including date of onset of symptoms, date of diagnosis, presenting symptoms, clinical findings, and results of angiography, were recorded to verify the diagnosis of TA. Angiographic classification was performed as defined in 1994 at the International Conference on TA (29).

At the re-evaluation period, among patients who were recorded as the ICD-10 code of M31.4, an accurate diagnosis of TA was confirmed in 88 patients using objective clinical data and the ACR criteria. Three patients presenting with symptoms suggestive of TA were excluded from the study. One of these patients was a 40-year-old female patient who had vision loss, claudication in the right upper extremity, blood pressure difference between two arms, and a pulseless upper extremity. Based on occlusion in the left common carotid artery, internal carotid artery and right subclavian artery in a magnetic resonance angiography, she was considered to have TA; however, she refused to participate in the study and to undergo classical angiography. Another 53-year-old female with a claudication of bilateral lower extremities had classic angiographic findings in the left vertebral, right internal iliac and right superficial femoral arteries, as well as stenosis in the bilateral subclavian arteries, left deep femoral artery, and left superficial femoral artery. However, since she did not meet the ACR 1990 criteria for the classification of TA because of her age of disease onset, she was excluded from further analysis. Finally, a 90-year-old female with a diagnosis of TA was lost to follow-up for three years and we were unable to contact her.

After excluding patients with TA who were living outside Izmir, 41 patients with TA were included in the final analysis. All the identified patients survived throughout the study period. Patients who had resided in central Izmir at the time of diagnosis, but who later moved from Izmir were excluded from prevalence calculations from that point forward.

The study protocol was evaluated and approved by the Ethics Committee of Dokuz Eylül University Faculty of Medicine.

Statistical analysis

All statistical analyses were performed using the Statistical Package for the Social Sciences (SPSS, Inc., Chicago, IL, USA) program version 15.0. Data were expressed as mean, standard deviation, and percentage (%) unless stated otherwise. Ninety-five percent confidence intervals (95% CIs) for prevalence and incidence were computed under the assumption of Poisson distribution. Incidence rates for age groups and gender were calculated according to the annual number of newly diagnosed patients. Annual prevalence rates in 2006-2010 were estimated by dividing the number of diagnosed TA cases in each or any previous year by the total population of central Izmir in the same year. Annual incidence rates were also calculated for the same time period by dividing the number of newly diagnosed TA patients in that year divided by the total population at risk in the same year. The mean annual incidence of TA was calculated as the observed number of TA cases during the study period divided by the cumulated population in central Izmir during 2006–2010. Prevalence and incidence rates were calculated overall, by sex and by age (for age groups below 40 and 40 years and older).

Results

Of the 41 patients with TA, 50% (n=21), 41% (n=17), 5% (n=2), and 2% (n=1) received follow-up care at Ege University Hospital, Dokuz Eylül University Hospital, Atatürk Training and Research Hospital, and Tepecik Training and Research Hospital, respectively.

The mean age of the 41 TA patients was 45.3±10.5 years (range, 24–63 years); of these patients, 38 (92.7%) were female and 3 (7.3%) were male (female-to-male ratio was approximately 13:1). The mean age at the time of diagnosis was 37.2±10.7 years (range, 20–56 years), the mean age at the onset of symptoms was 33.6±10.8 years (range, 13–55 years), the mean latency period for diagnosis was 42±60 months (range,

Table I. Annual incidence and prevalence rates of Takayasu's arteritis by gender in central Izmir.

Year		Total cases (n)	New cases (n)	Population of central Izmir	Incidence	Crude point prevalence	(≤40 years of age) (n) Total cases / new cases
2006		30	1	2,605,327	0.38 (0-1.1)	11.5 (7.4-15.6)	7/1
	Female	28	1	1,307,385	0.76 (0-2.27)	21.4 (13.4-29.4)	
	Male	2	0	1,297,942	0 (0)	1.5 (0-3.7)	
2007		34	4	2,649,582	1.5 (0-3.0)	12.8 (8.5-17.1)	9/2
	Female	32	4	1,329,598	3.0 (1.1-8.0)	24.1 (15.7-32.4)	
	Male	2	0	1,319,984	0 (0)	1.5 (0-3.6)	
2008		36	3	2,683,842	1.1 (0-2.4)	13.4 (9.0-17.8)	11/3
	Female	33	2	1,348,592	1.5 (0-3.5)	24.5 (16.1-32.8)	
	Male	3	1	1,335,250	0.7 (0-2.2)	2.2 (0-4.8)	
2009		40	4	2,740,306	1.5 (0-2.9)	14.6 (10.0-19.1)	12/1
	Female	37	4	1,376,001	2.9 (0-5.7)	26.9 (18.2-35.6)	
	Male	3	0	1,364,305	0 (0)	2.2 (0,3-4.6)	
2010		41	3	2,786,863	1.1 (0-2.3)	14.7 (10.2-19.2)	12/0
	Female	38	3	1,395,827	2.1 (0-4.6)	27.2 (18.6-35.9)	
	Male	3	0	1,391,036	0 (0)	2.2 (0-4.6)	

Values are expressed as per million population per year (95% confidence intervals).

1–660 months), and the mean follow-up period was 104±91 months (range, 2–360 months). It was determined that a diagnosis of TA was established after the age of 40 and older in 29 (71%) patients.

Between 2006 and 2010, the mean annual incidence of TA for the general population was estimated as 1.11 per million (95% CI: 0.54–1.67) in Izmir. The mean annual incidence among females and males was 2.06 per million (95% CI: 0.88–3.23) and 0.15 per million (95% CI: 0–0.56), respectively. This showed that the mean annual incidence of TA among females was 7.3 times that of males. During the same years, the mean annual incidence among the population less than 40 years of age was estimated as 0.83 per million (95% CI: 0–1.67). There was a gradual increase in the prevalence of TA during the course of this study; from 11.52 per million in 2006 to 14.71 per million in 2010. In 2010, the crude annual prevalence was estimated to be 6.9 per million (95% CI: 3.9–12.2) among the population equal to or less than 40 years of age. For the overall population, the minimum and maximum incidence rates were found to be 0.38 per million and 1.50 per million, respectively, between 2006 and 2010. When only the population of those less than 40 years

of age was considered, the mean annual incidence rate was found to be 0.83 per million (95% CI: 0–1.67). On December 31, 2010, the age and gender-adjusted prevalence rates of TA in central Izmir was 12.8 per million (95% CI: 12.0–13.6) for overall population. The incidence and prevalence rates according to year and gender are presented in Table I. A summary of age and gender-adjusted prevalence of TA based on the 2010 Turkish population census are shown in Table II.

Discussion

Much of the literature related to TA is originated from Asian countries, where the disease was once thought to be restricted to these regions. On the other hand, the majority of studies on TA concentrate on clinical presentations and characteristics of the disease, pathogenesis, and diagnostic imaging, as well as assessment of disease activity and management. Being a relatively rare disease, there is limited number of epidemiological studies concerning TA. The present study is the first epidemiological TA study performed in Turkey. We used the medical recording systems of the five available referral hospitals located in central Izmir, which is Turkey's third largest city. Izmir is located on the western coast of Turkey

Table II. Age and gender-adjusted prevalence of Takayasu's arteritis based on the 2010 Turkish population census.

	Prevalence for all ages	Prevalence for ≤40 years of age	Prevalence for >40 years of age
Total	12.8 (12.0-13.6)	4.0 (3.5-4.6)	8.8 (7.7-10.0)
Female	23.5 (21.9-25.0)	7.2 (6.2-8.3)	16.2 (14.1-18.6)
Male	1.9 (1.5-2.4)	0.7 (0-1.1)	1.2 (0-2.0)

Data are presented as per million (95% confidence interval) by direct standardisation method.

and serves as the gateway to the Aegean Region. The present study demonstrated that the prevalence rate of TA in Izmir gradually increased from 11.52 per million in 2006 to 14.7 per million in 2010. For the overall population, the minimum and maximum incidence rates were found to be 0.38 per million and 1.50 per million, respectively, between 2006 and 2010 (Table I). The incidence rates remained roughly stable for TA; the mean incidence rate was 1.1 per million (95% CI: 0.54–1.67). When only the population of those less than 40 years of age was considered, the mean annual incidence rate was found to be 0.8 per million (95% CI: 0–1.67). On December 31, 2010, the age- and gender-adjusted prevalence rates of TA in Izmir was 12.8 per million (95% CI: 12.0–13.6) for the overall population (Table II). The incidence and prevalence rates derived from this study, together with the available global epidemiological data from the literature, are summarised in Table III. Our data are consistent with the previous studies, confirming the rarity of TA.

Table III. Summary of epidemiologic data reported incidence and prevalence rates of Takayasu's arteritis in various countries.

Period	Study location	Data source	Total number of cases	Incidence (per million per year)	Prevalence (per million)	Reference
2006-2010 (5 years)	Turkey (Central Izmir)	Hospital-based and individual records of rheumatologists	41	0.38-1.51 (overall) 0.83 (<40 years)	14.7 (overall) 6.9 (<40 years)	Current study
1973-1975 (3 years) 1982-1984 (3 years)	Japan (nationwide)	Hospital-based (A combined results of two different studies)	Study-I: 2148 Study-II: 2606	≅1-2*	NR	Koide (17)
1994	Japan (nationwide)	Hospital-based	4500	NR	40.0*	Toshihiko (18), Numano and Kobayashi (30)
1958-1973 (16 years)	Japan (nationwide)	Autopsy-based	76	NR	360	Nasu (31)
1975-1984 (10 years)	Japan (nationwide)	Autopsy-based	115	NR	330	Nagata (32) Hotchi (16, 33)
1989-1994 (6 years)	Kuwait (nationwide)	Hospital-based	13	2.2 (overall) 3.3 (<40 years)	7.8 (overall) 9.5 (<40 years)	el-Reshaid et al. (34)
1971-1983 (13 years)	USA (Olmsted County, MN)	Hospital-based	32	2.6***	NR	Hall et al. (35)
1986-1990 (5 years)	USA (New-York State, NY)	Hospital-based	154	NR	0.9	Cotch et al. (12)
1969-1976 (8 years)	Sweden (Uppsala County)	Hospital-based	15	NR	6.4	Waern et al. (20)
1998-2002 (5 years)	Germany (Federal state Schleswig-Holstein, north Germany)	Regional registry (mainly from hospitals)	7	0.4-1.0	NR	Reinhold-Keller et al. (10)
1990-1999 (10 years)	Lithuania (Vilnius City)	Mainly hospital-based	6	1.3	NR	Dadoniene et al. (21)
2000-2005 (6 years)	United Kingdom (Norfolk region, East England)	Regional registry	14	0.8 (overall) 0.3 (<40 years)	4.7 (overall)	Watts et al. (22)
1990-2009 (20 years)	Denmark (Eastern part)	Central registry (1990-2006) and hospital-based (2007-2009)	19	0.4 (overall) 0.6 (<40 years)	8.0 (overall) 12.0 (<40 years)	Dreyer et al. (23)
1994 and 2010 (17 years)	Spain (Southern part)	Hospital-based	5	1.1	10.5	Romero-Gómez et al.(36)

NR: not reported. *Estimated from the given data in the literature (17, 18), ** Incidence estimate based on population over 16 years old, *** Incidence estimate based on population under 50 years old.

While evaluating these results, it should be kept in mind that our data reflected the minimum incidence and prevalence rates of TA in Izmir. There are other important considerations for our study. Firstly, the present study included only adult patients who were at least 18 years of age at the time of diagnosis of TA. Secondly, we did not include local clinics, district hospitals, specialised secondary care hospitals, and hospitals operated by private enterprise. Although private health insurance is well developed in Turkey, we did not collect any data for the patients attending private hospitals. Additionally, TA is not covered by a screening program in Turkey and some clinicians do not have enough experience in diagnosing TA. Therefore, some cases may have been misclassified or not recognised during the study period. Based on those factors, we admit that the actual incidence and prevalence rates of TA might be higher than that reported herein. However, many hospitals and health centres generally tend to refer TA cases to tertiary health care centres, once a presumptive or definitive diagnosis is established. Furthermore, since the majority of the population is covered by social security and national public health insurance, which is funded by the government, vulnerable patients with chronic diseases like TA generally do not prefer to go to private hospitals. Therefore, the inclusion of only five referral hospitals located in central Izmir presumably would not have significantly influenced our results. Likewise, since it is quite rare to encounter TA cases diagnosed before the age of 18 years, we assume that our screening strategy which did not cover pediatric patients would have only a negligible effect on our estimates.

The retrospective design of our study is another limitation. However, due to the lack of prospective data sources on TA, it is difficult to obtain more accurate data in Turkey. On the other hand, our study is not a nationwide epidemiologic survey. These results might not be representative for the whole country; however, it might reflect an informative disease occurrence rate for the population of Turkey. To date, nationwide epidemiologic surveys of TA related to frequency patterns have only been conducted in Japan.

The combined use of EMR from five large tertiary hospitals and medical file reviews conducted by individual rheumatologists is one of the strengths of the present study. Combining information from independent sources has the potential to provide a positive impact on the quality of patient data collected. Moreover, the present study reviewed the medical records covering a 5-year time period, which was long enough to reach an epidemiological analysis. This is a strength of this study since analysis of shorter time periods for a rare disease may not be enough to identify reliable data for epidemiologic investigations. Because the observation period in most epidemiologic studies comprise of more than 5 years, our study recruited considerably more patients within 5 years. Another strength of the present study was evaluation by a single clinician (Y.K.) who provided a comprehensive standardised approach to the collection of historic data for all patients.

The incidence and prevalence of TA have been reported to be higher in Southeastern and Eastern Asian countries than in other countries in the world. In line with those reports, the prevalence of TA reported in our study was significantly lower than that reported in Japan (18), where the estimated prevalence is 40 per million. However, our prevalence was significantly higher than those reported in the United Kingdom (22), Denmark (23), Sweden (20) and southern Spain (36). Our incidence rates were close to the rates reported from Japan (Table III). In agreement with clinical observations of many Turkish rheumatologists, the results of this study also confirm that TA is a relatively prevalent systemic vasculitis in Turkey.

In the literature, autopsy studies, performed mostly in Japan, document higher prevalence of TA; some have found prevalence to be as high as 1 per 3,000 (16, 31-33). However, similar postmortem studies have not been performed elsewhere to provide comparative data.

Turkey is a transcontinental country often described as a bridge between Europe and Asia. Comparing our results with the findings from other European studies demonstrated that the incidence rates for TA were similar to the incidence rates determined in Lithuania (21) and southern Spain (36) roughly comparable with Germany (10), but remarkably higher than the incidence rates in Denmark (23) and the United Kingdom (22). Such variations in the incidence could be due to true inter-population differences, but might also be due to differences in rates of reporting of a rare disease due to low awareness about disease occurrence. Insidious onset of disease, difficulty in diagnosing TA, and occasional misdiagnosis also cause considerable variations (37, 38). Other possible causes of variation between these studies may be imprecise estimates due to the fact that most incidence rates are based on a small number of cases.

As awareness of this disease increases among physicians, the incidence is expected to increase. During the past few decades, patients with TA have been increasingly recognised in Africa (39-41), Europe, and North America. This supports the belief that the true frequency of TA was probably underestimated previously in the West. Available data needs to be re-examined to be certain about possible ethnic skewing.

Disease onset for TA is reported mostly between the ages of 20 and 30 years. In previous Turkish studies, the mean age at onset was 34 years (26) and the median age at onset was 30.2±14 years (28). In this study, the mean age at onset of symptoms was 33.6±10.8 years. Of note, 29% (12 out of 41) of the patients had disease onset after 40 years of age and 71% (29 out of 41) of them were older than 40 years of age at the time of diagnosis. In summary, our results showed that the prevalence of TA among those aged ≤40 years was significantly lower than the prevalence among those older than 40 years of age. In other studies, the proportions of TA patients with disease onset after 40 years of age were 9% (42), 13% (25), 15% (43), 17.5% (24), and 23.1% (44). Moreover, in a French study (45), 32% of patients had disease onset after 40

years of age, 18.3% after 50 years, and 4.9% after 60 years, suggesting that late onset is not a rare feature of TA. These findings emphasise that those over 40 years of age should be considered for a diagnosis of TA and included in these kinds of studies. However, one female patient (53 years old) was excluded from this study as she did not meet the age requirement of the ACR 1990 criteria. We believe that revised classification criteria for TA are required to determine whether or not the giant cell arteritis can be distinguished from TA, regardless of age at onset.

In conclusion, according to the results of the first epidemiological TA study in Turkey, which was performed using the medical recording systems of five tertiary referral hospitals in central Izmir, TA is a relatively prevalent vasculitis. Although this study was not a nationwide epidemiologic survey and the results might not be representative for the whole country, the figures could provide an informative estimation of the frequency patterns of TA in Turkey. Hopefully, this study will provide a valuable addition to the current information on frequency rates of TA in South-East Europe.

The gradual annual increase of the prevalence rate of TA observed in the present study might be due to an increasing awareness of this disease. In brief, although lower than those reported in Japan, the reported frequency rates in Turkey were higher than in those for many European countries. As discussed above, it is highly likely that the true frequency of TA in Turkey is somewhat higher than assessed in this current investigation. Therefore, we agree with many other Turkish rheumatologists who believe that TA is the second most frequent systemic vasculitis seen in Turkish adults, after Behcet's disease.

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