EDITORIAL

New data on sarcoidosis in Poland

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Sarcoidosis is a multiorgan inflammatory disease, characterized by granuloma formation in the affected organs.[1] The disease most commonly involves the respiratory system, although various other sites are often affected.[2] Sarcoidosis usually remits spontaneously; however, in some cases, it leads to significant pulmonary fibrosis and impairment of lung function, or it can be life-threatening due to central nervous system or cardiac involvement.[3] Despite extensive studies, the etiology of sarcoidosis remains unknown. The genetic and environmental traits have been reported, and the most common hypothesis suggests that an environmental trigger (possibly mycobacteria, propionibacteria, or other organic and inorganic substances) provokes a specific response in susceptible individuals.[2] However, there is no firm evidence to support this concept.

There are considerable differences in reported data on the prevalence of sarcoidosis, with the rates ranging from less than 4/100 000 to more than 140/100 000.[4] Moreover, the prevalence varies across countries and races. In general, papers from the United States and Scandinavian countries report higher prevalence and suggest that sarcoidosis is much more common among individuals of African-American origin. Also publications from Europe differ in reported prevalence, with estimations ranging from 5.9/100 000 in Greece to more than 150/100 000 in Sweden.[5,6] There is a need for actual data because the prevalence of sarcoidosis has been shown to be rising.[7] Most reviews report female predominance, while recent epidemiological data have suggested the opposite.

In healthcare settings where most patients with suspected sarcoidosis are hospitalized for diagnosis, hospital-based registry data can be used to study the epidemiology of sarcoidosis. This also holds true for the Polish healthcare system, and this methodology can provide valid estimations on disease incidence and prevalence. Indeed, some recent studies on the epidemiology of sarcoidosis in Poland have been performed using this method. A study conducted in the Silesian Voivodeship estimated the incidence of sarcoidosis to range from 3.8 to 4.5/100 000 and reported local differences in incidence rates.[8] A large single-center study from the National Tuberculosis and Lung Diseases Research Institute, published in 2018, aimed to characterize 1810 patients hospitalized from 2010 till 2013.[9]

In this issue of the *Polish Archives of Internal Medicine* (*Pol Arch Intern Med*), Bogdan et al[10] reports on sarcoidosis epidemiology in Poland, based on data obtained using similar methods. Their study provides important information because they assessed nationwide data based on 23 097 patients hospitalized with the diagnosis of sarcoidosis from 2008 to 2015. The authors estimated the annual incidence rate of sarcoidosis to be 7.5/100000. The 95% confidence interval was narrow (7.1–7.9), reflecting the study design. The prevalence for the end of the studied period of time (2015) was estimated at 60/100 000. It should be noted that this estimation concerns the whole country population, so the prevalence rates in the most commonly affected age groups could be about twice as high. Estimations based on hospital discharge records are probably slightly underrated, because some patients with sarcoidosis might not have been hospitalized for some reasons (eg, lack of consent to a hospitalization or an invasive diagnostic procedure). However, knowing the Polish healthcare settings, the number of such patients is most probably small.

The incidence rate reported by Bogdan et al[10] is higher than that reported by previous local studies.[8,11] However, this reflects regional differences rather than the increasing disease incidence, because Bogdan et al[10] did not observe a significant increase in the number of hospitalizations due to sarcoidosis throughout the study period (2008–2015). A higher proportion of men (54.7%) is consistent with some recent findings from Poland and other countries,[9,12] which is in contrast to a widespread opinion that sarcoidosis is more common among women.[2] The mean age of hospitalized patients was about 45 years; however, this measure is not informative because of

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significant sex differences. The peak incidence was observed among individuals aged 30 to 39 years for men and 50 to 59 years for women, a pattern that has been reported in some, but not all, countries.[2] Similar data on sex-age distribution were delivered by a recent multicenter European study, which included also sites from Poland.[13]

Data on the affected systems or organs reported by Bogdan et al[10] should be interpreted with consideration of the limitations inherent to the methods used. There are a couple of caveats related to the use of *International Classification of Diseases, Tenth Revision* (*ICD-10*) codes for sarcoidosis. Firstly, the classification provides only one common code for sarcoidosis with lymph node involvement (D86.1). It is safe to assume that most patients coded as D86.1 were those with mediastinal lymph node involvement; however, this approach excludes cases of sarcoidosis with extrapulmonary lymph node involvement. Secondly, there is only one common code for extrapulmonary sarcoidosis (D86.8), and this rules out a detailed analysis of extrapulmonary sarcoidosis (except for the possibility of using an additional code for eye involvement [H22.1], not reported by Bogdan et al).[10] Overall, the proportions of respiratory system involvement are within the expected range and are similar to those reported by another large single-center study from Poland.[9]

In the study by Bogdan et al,[10] the proportion of sarcoidosis with skin involvement was only 1.4%, which is much lower than the rates reported in other studies.[2,13] This may suggest that some forms of extrapulmonary sarcoidosis might be underdiagnosed in the Polish population. Indeed, we rarely see patients with skin or ocular involvement in our everyday practice, despite the fact that skin and eye involvement is reported in about 15% and 10% to 30% of patients, respectively.[2] This is also reflected in some other previous Polish reports.[9] Apart from underdiagnosis, this discrepancy might have been caused by regional differences in sarcoidosis presentation or bias related to the use of *ICD-10* codes (patients with lung and extrapulmonary site involvement should be theoretically assigned 2 codes, which is most probably rarely done).

The paper by Bogdan et al[10] provides an important insight into the current epidemiology of sarcoidosis in Poland. Furthermore, the database used by the authors offers some possibilities for further analyses, for example, of geographical differences within Poland. Geographical variations within countries have been reported in Europe,[14,15] and such an analysis could combine the available data on residence, environmental factors, and clinical presentation, possibly leading to some interesting observations.

ARTICLE INFORMATION

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REFERENCES

1 Hunninghake GW, Costabel U, Ando M, et al. ATS/ERS/WASOG statement on sarcoidosis. American Thoracic Society/European Respiratory Society/World Association of Sarcoidosis and other Granulomatous Disorders. Sarcoidosis Vasc Diffuse Lung Dis. 1999; 16: 149-173.

2 Valeyre D, Prasse A, Nunes H, et al. Sarcoidosis. Lancet. 2014; 383: 1155-1167.

3 Iannuzzi MC, Fontana JR. Sarcoidosis: clinical presentation, immunopathogenesis, and therapeutics. JAMA. 2011; 305: 391-399.

4 Gerke AK, Judson MA, Cozier YC, et al. Disease burden and variability in sarcoidosis. Ann Am Thorac Soc. 2017; 14: S421-S428.

5 Karakatsani A, Papakosta D, Rapti A, et al. Hellenic Interstitial Lung Diseases Group. Epidemiology of interstitial lung diseases in Greece. Respir Med. 2009; 103: 1122-1129.

6 Arkema EV, Grunewald J, Kullberg S, et al. Sarcoidosis incidence and prevalence: a nationwide register-based assessment in Sweden. Eur Respir J. 2016; 48: 1690-1699.

7 Gerke AK, Yang M, Tang F, et al. Increased hospitalizations among sarcoidosis patients from 1998 to 2008: a population-based cohort study. BMC Pulm Med. 2012; 12: 19.

8 Kowalska M, Niewiadomska E, Zejda JE. Epidemiology of sarcoidosis recorded in 2006-2010 in the Silesian voivodeship on the basis of routine medical reporting. Ann Agric Environ Med. 2014; 21: 55-58.

9 Martusewicz-Boros MM, Boros PW, Wiatr E, et al. Systemic treatment for sarcoidosis was needed for 16% of 1810 Caucasian patients. Clin Respir J. 2018; 12: 1367-1371.

10 Bogdan M, Nitsch-Osuch A, Kanecki K, et al. Sarcoidosis among hospitalized patients in Poland: a national hospital register-based study. Pol Arch Intern Med. 2019; 129: 580-585.

11 Szafrański W. Interstitial lung diseases among patients hospitalized in the Department of Respiratory Medicine in Radom District Hospital during the years 2000–2009. Pneumonol Alergol Pol. 2012; 80: 523-532.

12 Fidler LM, Balter M, Fisher JH, et al. Epidemiology and health outcomes of sarcoidosis in a universal health care population: a cohort study. Eur Respir J. 2019. [Epub ahead of print].

13 Schupp JC, Freitag-Wolf S, Bargagli E, et al. Phenotypes of organ involvement in sarcoidosis. Eur Respir J. 2018; 51: pii: 1700991.

14 Arkema EV, Grunewald J, Kullberg S, et al. Sarcoidosis incidence and prevalence: a nationwide register-based assessment in Sweden. Eur Respir J. 2016; 48: 1690-1699.

15 Jamilloux Y, Maucort-Boulch D, Kerever S, et al. Sarcoidosis-related mortality in France: a multiple-cause-of-death analysis. Eur Respir J. 2016; 48: 1700-1709.