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EDITORIAL

Measuring sarcoidosis around the world: Using the same ruler



Sarcoidosis is a worldwide disease, although the incidence and severity between areas of the world varies considerably.^{1,2} This variability of presentation and outcome of the disease has been evident for years.³ These differences are in part due to differences not just in genetic background and environment, but also use of different criteria for diagnosis and rigor of evaluation of pulmonary and extra-pulmonary disease. In this issue of the Journal, Cardoso and colleagues report on the presentation and outcome of sarcoidosis in the Oporto region of Portugal.⁴ Using standardized criteria, the investigators have provided information so one can compare and contrast sarcoidosis in their clinic versus other parts of the world. The authors directly compared their results to other reports from Europe, Asia, and the United States. While their findings were similar to most European reports, there are some interesting differences. For example, respiratory symptoms were much more common in Oporto than Lisbon. The variation between sites could be due to how patients were identified for study. For example, recruiting patients from a specialized pulmonary center may identify different patients from those encountered in a rheumatology clinic.

Another major source of variation between reports is the measuring stick used to define organ involvement. Over the years, several groups have tried to minimize the variability of descriptions of sarcoidosis patients by applying standard evaluations to all patients studied. In one study, the same team of investigators evaluated all patients in two different countries (Finland and Japan).⁵ The differences in the two clinic populations were mostly based on genetic differences. However, those investigators noted that there was a difference in the frequency of screening chest X-ray (every year in Japan and every three years in Finland).

In the United States, a case controlled epidemiologic study of sarcoidosis (ACCESS) involved ten centers of newly diagnosed sarcoidosis. An instrument was developed to

standardize organ involvement.⁶ That instrument allowed one to demonstrate difference in organ involvement based on race, age, and gender.⁷ While this was a useful instrument, over the years it became clear that not all forms of organ involvement were detected. Also, new techniques especially PET and MRI scanning were not included in the original instrument.⁸ An updated instrument was reported in 2014⁹ included these newer diagnostic modalities. In that report, the classifications “highly probable”, “at least probable”, and “possible” were used to indicate the level of certainty of organ involvement in a patient with known sarcoidosis. For example, bilateral hilar adenopathy in a patient with known sarcoidosis is very likely to indicate pulmonary involvement. However, not all patients with bilateral hilar adenopathy have sarcoidosis.

While the WASOG instrument is a useful ruler to measure different organ involvement, it still has its limitations. For example, there was no comment on Lofgren’s syndrome. The presence of bilateral hilar adenopathy in a patient with erythema nodosum or periarticular swelling is almost always associated with sarcoidosis.¹⁰ This manifestation of sarcoidosis is often associated with a good clinical outcome. The good outcome has been associated with certain genetic markers.¹¹

The application of the new WASOG instrument may increase the detection of specific organ involvement. For example, the criteria for cardiac involvement has widened between the two instruments. The current guidelines lists nine criteria for at least probable cardiac involvement⁹ as opposed to the five criteria for the earlier instrument.⁶ In a recent report of 73 patients with cardiac sarcoidosis, all but one of these criteria identified cardiac involvement in one or more sarcoidosis patient.¹² For cardiac sarcoidosis, other have demonstrated the harder one looks, the more likely one is to find cardiac sarcoidosis. At the Mount Sinai New York sarcoidosis clinic, an early report found that only eight percent of their sarcoidosis patients had evidence of cardiac sarcoidosis.¹³ However, a report from the same clinic twenty years later identified cardiac involvement in over forty percent of patients.¹⁴ While this second report

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may have been of a select group of patient, this dramatic increase in the rate of cardiac involvement emphasizes the need for standard criteria.

The use of a standardized method of reporting organ involvement is an important step in better understanding sarcoidosis. Use of such a tool allows the researcher to identify potential differences based on genetic background and environmental exposures.¹⁵ Such studies have been useful in understanding the importance of HLA markers in Lofgren's syndrome.¹¹ HLA markers are most predictive in Sweden and have been generally helpful in other parts of Europe. However, they have not been as useful in African American sarcoidosis patients. Interaction between environmental exposures and other HLA markers have been identified with various manifestations of pulmonary and extra pulmonary disease.¹⁶

An accurate picture of what sarcoidosis looks like is also helpful for the clinician. For example, Lofgren's syndrome was noted in over fifteen percent of patients from both Oporto and Lisbon. This is twice as frequent as seen in Germany and United States. In Portugal, the clinician should be sensitive to the fact that the presence of ankle swelling and pain as well as erythema nodosum are key features of Lofgren's syndrome and will often lead to a diagnosis of sarcoidosis. Since these features are often self limited, one should ask specifically about these problems.

One of the interesting aspects of sarcoidosis is its multiple ways of presentation. However, patterns of organ involvement are well recognized. In the report by Cardoso et al., a table compares the different manifestations of sarcoidosis for various parts of the world. As we become more accurate in defining organ involvement, we will be able to determine which of these differences are due to the ruler and which are due to the disease.

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