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The ability to predict the clinical course of pulmonary sarcoidosis from data that is right in front of us

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The natural course of sarcoidosis is highly variable. It is a disease that may never lead to symptoms or functional impairment. However, sarcoidosis may cause distressing symptoms that greatly impair quality of life. Sarcoidosis may also lead to significant organ dysfunction that may incapacitate patients and lead to death. The majority of poor outcomes from sarcoidosis relate to the development of pulmonary fibrosis[1] and specific organ involvement (heart and central nervous system).[2] In addition, even chronic, well-controlled disease is often debilitating because of the need for chronic corticosteroid therapy and its resultant complications.[3]

This wide variation in sarcoidosis outcomes creates problems for the caregiver treating patients newly diagnosed with the disease. The clinician wants to avoid overaggressive treatment of inconsequential cases as well as adequately treat cases destined for a poor outcome. Clearly, sarcoidosis patient care could be improved by the construction of reliable prognostic tools. However, at the present time, such tools are lacking including reliable screening algorithms for cardiac sarcoidosis[4] and reliable biomarkers for the development of pulmonary fibrosis.[5] Proposed prognostic biomarkers for sarcoidosis include imaging studies and technical genetic, proteomic, or immunologic tests, as well as gene transcription tests, that are all unproven and unavailable in most parts of the world.[5,6]

In this issue of the Brazilian Journal of Pulmonology, Castro et al.[7] performed a longitudinal retrospective analysis of the clinical presentation and the results of routine medical tests in 200 pulmonary sarcoidosis patients treated at three Brazilian medical centers. These authors recorded patient demographics, symptom onset, date of diagnosis, routine chest imaging results, and sarcoidosis organ involvement. All these clinical data are standard, and a clinician should obtain them in the routine management of a sarcoidosis patient. These authors[7] then analyzed the association of these clinical data with the sarcoidosis outcome of self-limited disease versus persistent disease at two years. The authors performed this analysis on the 160 non-fibrotic patients, as those with fibrotic disease would be expected to have chronic pulmonary symptoms and frequently require treatment, although this is not always the case. In a multivariate analysis, the following clinical features were statistically associated with persistent disease in non-fibrotic pulmonary sarcoidosis patients: reduced FVC, the presence of dyspnea, parenchymal lung involvement on radiographic imaging, involvement of ≥ 2 non-thoracic organs with sarcoidosis, and a delay in the diagnosis of sarcoidosis of > 12 months after symptom onset. The authors went on to construct a scoring system based on weighing these factors that was able to discriminate the likelihood of persistent disease at two years after diagnosis between 13% (low score) and 82% (high score).

This analysis has several problems and limitations. First, many of the factors that were found to be associated with poor sarcoidosis outcomes were not novel and had been identified in previous studies. These include symptoms versus no symptoms,[8] parenchymal disease versus no parenchymal disease,[9] extrapulmonary disease,[9] and reduced FVC.[10] Second, the results may not be generalizable to other populations living where there is a low prevalence of tuberculosis. Tuberculosis is not only a very common granulomatous lung disease in Brazil, but a misdiagnosis of tuberculosis as sarcoidosis may result in inappropriate corticosteroid treatment with disastrous consequences. Therefore, despite the association of a delayed diagnosis of sarcoidosis and persistent disease at two years, it may be prudent to delay the diagnosis and treatment of sarcoidosis in areas of high tuberculosis prevalence to ensure that the latter disease is clearly excluded. In other words, a rapid diagnosis of sarcoidosis may improve sarcoidosis outcomes, but it is unclear what consequences may result from the misdiagnosis of tuberculosis as sarcoidosis, especially in areas with a high frequency of tuberculosis. Finally, a rapid diagnosis of sarcoidosis may have been associated with a good prognosis simply because physicians who are knowledgeable in the management of sarcoidosis may establish the diagnosis more rapidly.

Despite these issues, these researchers[7] should be commended for performing this analysis. Even though many of the factors that these authors identified as associated with the prognosis of pulmonary sarcoidosis have been known, they are often not appreciated by the clinician. In addition, these prognostic factors involve routine clinical tests that should be available. We await accurate and more reliable biomarkers to clearly distinguish the clinical course of pulmonary sarcoidosis. However, in the meantime, the clinician should use all the tools that are currently available. The prognostic utility of this scoring system is accurate enough to give the pulmonary sarcoidosis patient a reasonable estimate of a long-term outcome and give the physician a framework of how aggressively to monitor and treat these patients.

CONFLICT OF INTEREST

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