# Sarcoidosis – time for a clinical refresher!

The diagnosis and management of sarcoidosis continue to be a challenge, as Morar and Feldman[1] remind us in this issue of *AJTCCM*. As noted by them, and in the general South African specialist experience, sarcoidosis is regularly misdiagnosed as tuberculosis (TB). A little extra effort in radiology training and expertise is required, but tends to be a deficiency in undergraduate training throughout the country. Being an orphan disease, sarcoidosis is also rarely a topic of CME activities among medical officers and general practitioners, limiting their expertise. And when it is misdiagnosed, it is difficult to understand why treatment is often continued for prolonged periods – often 9 months or longer – before the diagnosis is reconsidered. One resulting complication is that patients are unnecessarily exposed to the side-effects of anti-tuberculosis therapy. Another is that complications of sarcoidosis may continue unabated: pulmonary fibrosis, and ocular and metabolic complications such as hypercalcaemia and its sequelae. One can understand that the interstitial abnormalities may be confused with TB, but we should remember that mediastinal and paratracheal adenopathy are generally not features of TB in the immunocompetent, and should trigger a reconsideration of the diagnosis.

The traditional radiographic staging of sarcoidosis has been questioned for its accuracy and poor interobserver concordance.[2] It is probably anachronistic and ought to be relegated to history, as it is incorrect and has minimal clinical significance for the following reasons: (*i*) patients do not progress through the stages; (*ii*) computed tomography scanning is superior and is likely to demonstrate lymphadenopathy and parenchymal changes that cannot be appreciated on chest radiographs;[2] (*iii*) a chest radiograph with normal lung fields does not mean that there is no parenchymal involvement – non-caseating granulomas have been found in up to 100% of such patients undergoing lung biopsy;[3] and (*iv*) one cannot be certain about the so-called end-stage fibrosis, because there may still be active granulomas that are difficult to discern amid the fibrosis.[4]

Rather, one should merely note the radiological abnormalities and take them into consideration in decision-making without assigning a specific stage.

As regards lung function testing, 21% of patients were noted to have an obstructive defect. With sarcoidosis being an interstitial lung disease, one would have expected all to have a restrictive defect. However, the characteristic peribronchial distribution of granulomas results in an obstructive defect as the commonest abnormality, and should also not result in a misdiagnosis of asthma.[5] The fact that more patients had restrictive defects is a reflection of the larger proportion having interstitial changes, and has bearing on the following observation in the study.

A high proportion of Morar and Feldman's patients needed corticosteroids and relapsed on cessation of therapy. This finding probably reflects the spectrum of sarcoidosis seen at referral centres. Patients with acute and self-limiting forms of the disease are, not unexpectedly, seldom referred to specialist clinics. Those with recurrent or persistent symptoms and activity limitation are more likely to be referred to specialists and require long-term immunosuppression or corticosteroid-sparing agents. In a total population of sarcoidosis patients, ~10 - 30% will develop progressive pulmonary disease.[6]

Chronic pulmonary sarcoidosis can be a therapeutic challenge. A prompt and accurate diagnosis has important clinical implications, as has delineating the extent of involvement and surveillance for new organ involvement and the pulmonary course. These have an influence on morbidity and mortality and ensuring optimal patient outcomes.

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