Re-visiting association between systemic sclerosis and sarcoidosis: prevalence and clinical features.

Ana Lages1,2, Huw Beynon2, Christopher P. Denton2, Voon Ong2

1Internal Medicine Department, Hospital de Braga, Braga, PORTUGAL
2Centre for Rheumatology and Connective Tissue Diseases, University College London, London, UNITED KINGDOM

Background
Systemic sclerosis (SSc) and sarcoidosis are both uncommon connective tissue diseases (CTD) with reported prevalence of about 3.08 in 100,000 (1) and 20 in 100,000 (2) respectively. Case reports and a recent review suggest an association between the two diseases as well as the possibility that the CTD may trigger granulomatous inflammation (3). We evaluated the prevalence and clinical features of patients with both these diseases.

Methods
We retrospectively examined the clinical database of all SSc patients (n=2500) in our centre over the last 12 years.

Results
We identified a sub-cohort of 827 patients. From these, 11 patients (1.33%) were found to have both SSc and sarcoidosis. The median age was 63.0±11.2 (years±SD). The majority of patients (63.7%) were Caucasians. The diagnosis of sarcoidosis preceded SSc in 7 patients and was contemporaneous in one patient. The median interval for diagnosis between the two diseases was 14.0±11.4 years. All patients, but one, were anti-nuclear antibodies (ANA) positive with homogenous pattern identified in 5 patients (45.5%) and centromere pattern in 4 (36.4%). Extra-nuclear antibodies (ENA) were positive in 5 patients, mainly anti-Scl70 (36.4%). Sarcoidosis was biopsy proven in 6 patients, one had a positive Kveim test and 4 were considered to have sarcoidosis based on clinical features and supportive investigations. Five patients had single-organ involvement related to sarcoidosis (lung, lacrimal gland, lymph nodes). Interstitial lung disease (ILD) was present in 5 patients (45.5%), a majority of these harboured anti-Scl70 antibodies. 40% of these had radiological changes attributable to both diseases and the remaining 3 patients had changes related to SSc or sarcoidosis alone. Three of the patients with ILD required immunosuppression. Apart from two patients who developed pulmonary arterial hypertension and one with suspected cardiac sarcoidosis, no other major organ involvement was documented.

Conclusions
The data suggests that the observed prevalence for coexisting diseases is higher than expected. Interestingly, the gender preponderance is more similar to that observed in SSc than sarcoidosis (4,5). In contrast to recent review, a majority in this cohort had sarcoidosis prior to SSc suggesting that it is less likely that SSc may be associated with granulomatous formation (3). The pulmonary involvement shared between the two diseases is consistent with the concept of similar aetiopathogenesis for SSc and sarcoidosis.