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

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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.

Case	Gender	Age	Race	SSc subset	Sarcoidosis involvement	Year of diagnosis		Major organ involvement	Lung disease	ANA	ENA	Immunossupression
						SSc	Sarcoidosis					
1	F	65	Ca	lcSSc	Lung + LN	1988	1970	-	-	H	-	-
2	F	64	Ca	dcSSc	Lung	1984	1995	ILD	NSIP	H	Scl70	Previous AZA
3	F	49	Ca	lcSSc	Lung + LN	1982	2008	ILD	Sarcoid nodules	-	-	-
4	F	66	W/B	lcSSc	Lung + Skin	2000	1994	ILD	NSIP	H	Scl70	MMF + Steroids
5	M	36	W/A	lcSSc	Lung + LN	2007	2006	Suspected cardiac sarcoidosis	-	FS	PM/Scl	MMF + Steroids
6	F	66	Ca	lcSSc	Lacrimal glands	2008	1994	-	-	C	-	-
7	M	73	Ca	lcSSc	Lung + LN	2009	1978	-	-	C	-	-
8	F	45	Ca	lcSSc	Lung	2002	2002	ILD	NSIP + Sarcoid nodules	H	Scl70	MMF + Steroids
9	M	63	Ca	lcSSc	LN + Eye	2006	1975	PAH	-	C	-	-
10	F	50	B	dcSSc	Lung	2011	2008	ILD	NSIP + Sarcoid nodules	H	Scl70	Steroids
11	F	60	I	lcSSc	LN	1999	2013	PAH	-	C	-	-

Table 1: Table 1. Patient's clinical and analytical features..

F – female; M – male; Ca – caucasian; W/B – mixed caucasian and african-american; W/A – mixed caucasian and asian; B – african-american; I – indian; lcSSc – limited cutaneous systemic sclerosis; dcSSc – diffuse cutaneous systemic sclerosis; LN – lymph nodes; ILD – interstitial lung disease; PAH – pulmonary arterial hypertension; NSIP – non-specific interstitial pneumonia; H – homogenous; FS – fine speckled; C – centromere; AZA – azathioprine; MMF – mycophenolate mofetil

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.

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