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RC039 - Clinical usefulness of ordering tests of anticentromere antibodies in patients with known or suspected scleroderma in routine care

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Resumen

Introduction: Anticentromere antibodies (ACA), antinuclear antibodies that target the centromere, have been classically associated with systemic sclerosis (SSc) and rarely found in patients with other connective tissue diseases (CTDs) and healthy controls. ACA are useful in distinguishing CREST patients from other patients with SSc or primary Raynaud's phenomenon, with a sensitivity of 33% and a specificity of 99,9%.

Objectives: To assess the value of ACA determination in a tertiary academic centre and to explore the referral pattern to the autoimmunity laboratory for testing among medical specialists.

Methods: All requests and samples received in the autoimmunity laboratory from 2008 to 2014 for ACA determination by immunofluorescence were included for analysis. The test was confirmed by immunoblot. The following data were collected retrospectively after reviewing the electronic clinical charts: confirmed or suspected diagnosis at baseline, demographics (age and gender), clinical manifestations and the medical specialist requesting the ACA test. We determine sensitivity, specificity and predictive values in our sample and present a description of the patients and referral patters for these patients.

Results: A total of 1,756 samples were included, only 141 (8%) were ACA positive. We found in our sample a sensitivity of 64.4%, specificity of 96%, positive predictive value (PPV) 53.9%, negative predictive value (NPV) of 97.3% (Table 1). A total of 118 patients with SSc were identified in the sample, 76 (64%) ACA positive and 42 (36%) ACA negative. A total of 65 patients were identified with positive ACAs but no SSc, most of them without diagnosis for any autoimmune disease and less frequently with autoimmune hepatitis, lupus, Sjögren´s syndrome, or rheumatoid arthritis (Table 2). Rheumatologists were the most frequent referral specialist, followed by internists. Around 70% of patients with ACA+ have neither diagnosis of SSc nor other autoimmune disease at the time of determination. However, Raynaud&#39s phenomenon was present in 37% of patients in this group.

Table 1. Anticentromere antibodies (ACA) in diagnosis of systemic sclerosis (SSc)

	Systemic Sclerosis Diagnosis		Total
ACA	Yes	No	

Positive, n (%)	76 (4.4%)	65 (3.7%)	141 (8%)
Negative, n (%)	42 (2.4%)	1,560 (89.5%)	1,602 (92%)
Total	118 (6.8%)	1,625 (93.2%)	1,743 (100%)

Table 2. Demographic and clinical characteristics of the patients and referral pattern

	ACA+		ACA-
	SSc (N: 76)	No SSc (N: 65)	SSc (N: 42)
Age Median	64,6	56,8	57,9
Female (%)	72 (95%)	56 (86%)	37 (88%)
	SSc limited: 70 (91%)	Non-autoimmune disease: 46 (70%)	SSc limited: 26 (61%)
	SSc diffuse: 6 (8%)	Autoimmune Hepatitis: 6 (9%)	SSc Diffuse: 16 (37%)
Diagnostic (%)	Lupus: 1 (1%)	Lupus: 5 (8%)	Lupus: 1 (2%)
		Sjögren´s syndrome: 5 (8%)	
		Rheumatoid Arthritis: 3 (5%)	
Clinical manifestations, n (%)			
Raynaud’s phenomenon	70 (92%)	24 (37%)	34 (81%)
Digitals ulcers	13 (17%)	3 (5%)	11 (26%)
Dysphagia	11 (14%)	5 (8%)	13 (31%)
Referral pattern, n (%)			
Rheumatology	46 (60%)	54 (82%)	39 (93%)
Internal Medicine	16 (21%)	9 (14%)	2 (5%)
Clinical Immunology	2 (3%)	1 (2%)	0 (0%)
Family Medicine	6 (8%)	1 (2%)	0 (0%)
Dermatology	3 (4%)	0 (0%)	1 (2%)
Gastroenterology	3 (4%)	0 (0%)	0 (0%)

Conclusions: Because of its high specificity and negative predictive value ACA testing is very useful in SSc diagnosis and needs to be performed in patients with high suspicion of SSc or other systemic autoimmune disease. Long-term studies are necessary to know the predictive value of ACA to develop SSc in patients with Raynaud´s phenomenon without a clear diagnosis.

References

1. Reveille JD, Solomon DH. Arthritis Rheum. 2003;49(3):399-412.