SAT0361 CHEST COMPUTED TOMOGRAPHY AS AN ALTERNATIVE TO HIGH RESOLUTION MANOMETRY FOR THE DIAGNOSIS OF SCLERODERMA ESOPHAGUS

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Background: Severe esophageal disease in patients with systemic sclerosis (SSc), referred as scleroderma esophagus, is characterized by ineffective or absent peristalsis along with hypotensive esophagogastric junction (hEGJ). The associations between scleroderma esophagus and different clinical and laboratory manifestations of SSc are still controversial.
Objectives: To assess associations between scleroderma esophagus, diagnosed by high resolution manometry (HRM), and other manifestations of disease.
Methods: Fifty-four consecutive SSc patients (49 women, mean age 50.6±11.6 years) with esophageal symptoms underwent clinical interview, medical records review and HRM. HRMs were analyzed according to the Chicago Classification in order to provide esophageal motility diagnosis; EGJ <9 mmHg was considered hypotensive. Demographic characteristics, patient-reported symptoms, SSc subtypes, pulmonary fibrosis, cutaneous ulcers, and anti-Scl-70 positivity were compared between SSc patients with or without scleroderma esophagus. Comparison was also performed in chest computed tomography (CT) findings of esophageal lumen in 26 patients with available data. Esophageal dilatation was deemed present when the diameter was >9 mm.
Results: Absent contractility was present in 37 (68.5%) patients; among these patients hEGJP was found in 32, thus 32/54 (59.2%) patients had classic scleroderma esophagus. There were no associations with gender, age, esophageal symptoms, skin involvement extent, anti-Scl-70, pulmonary fibrosis and cutaneous ulcers. Notably, esophageal dilation on chest CT was more frequent in patients with scleroderma esophagus compared to those without (77% *vs.* 8%, p=0.04, respectively).

Table 1. Associations between scleroderma esophagus and various SSc manifestations

	Scleroderma esophagus (n=32)	No scleroderma esophagus (n=22)	p
Age (yrs)	52. 5±11.9	52. 5±11.9	
Sex (F, %)	28 (87.5)	21 (98.4)	0.362
Diffuse SSc (n, %)	12 (55.5)	14 (43.8)	0.646
Pulmonary fibrosis (n,%)	25 (78.1)	15 (68.1)	0.750
Cutaneous ulcers (n, %)	19 (59.4)	12 (54.5)	0.854
Anti-Scl-70 (n, %)	23 (71.9)	12 (54.5)	0.540
CT coronal diameters >9 cm (n, %)*	10 (76.9)	1 (7.7%)	0.045
Presence of symptoms (%)			
– dysphagia	18 (56.3)	13 (39.1)	0.914
– heartburn	25 (78.1)	21 (95.4)	0.620
– regurgitation	28 (87.5)	19 (86.4)	0.126
– chest pain	13 (56.3)	7 (31.8)	0.653

*Data available in 26 SSc patients (13 with and 13 without scleroderma esophagus).

Conclusions: Scleroderma esophagus diagnosed by HRM was present in less than 2/3 of symptomatic patients with SSc and associated only with esophageal dilation (>9mm) in CT. Although further studies are needed, esophageal dilation on chest CT may be a non-invasive alternative for evaluation of SSc patients with esophageal symptoms.
Disclosure of Interest: None declared
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SAT0362 RISK FACTORS AND OUTCOME OF THAI PATIENTS WITH SCLERODERMA RENAL CRISIS (SRC): A DISEASE DURATION-MATCHED CASE CONTROL STUDY

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Background: Data regarding the prevalence, risk factors and outcome of scleroderma renal crisis (SRC) in Asian patients with systemic sclerosis (SSc) are limited.
Objectives: To determine the prevalence, risk factors and outcome of SRC in Thai SSc patients.
Methods: Medical records of all SSc patients, fulfilling the 1980 American College of Rheumatology classification criteria for SSc, seen at the Division of Rheumatology, Maharaj Nakorn Chiang Mai Hospital, Thailand, from January 1990 and December 2015 were retrospectively reviewed. Patients younger than 18 years old or overlapping with rheumatoid arthritis, systemic lupus erythematosus or mixed connective tissue disease were excluded. Cases of SRC were identified based on the international Scleroderma Renal Crisis Study criteria[1]. Controls were selected from consecutive SSc patients without SRC from our database that was ranked closest to the cases based on hospital number, and matched for disease duration from the first non-Raynaud's phenomenon, by±1 year. The ratio for SRC patients to control patients was 1:4.
Results: Of 608 SSc patients seen during the study period, 19 SRC were identified resulting of the SRC prevalence of 3.13% and there were 76 matched controls. Of the 19 cases, mean±SD age and median (IQR1, 3) disease duration was 56.2±13.8 years and 9 (5.0, 16.0) months, respectively. Seventeen patients (89.5%) had dcSSc. There were 12 (63.2%) patients with hypertensive renal crisis (HRC) and 7 (16.8%) patients with normotensive renal crisis (NRC). Univariate conditional logistic regression analyses showed that current myositis (Odd ratio [OR]21.75, 95% CI 2.65– 178.71,p=0.004), cardiac involvement (OR5.68, 95% CI2.03 – 15.84,p=0.001), age >60 years (OR2.82,95% CI 1.01 – 7.94,p=0.049), WBC>10,000 cell/mm^3 (OR2.69,95% CI 0.99 – 7.26,p=0.050), serum albumin<3.0 mg/dl (OR11.08,95% CI 3.02 – 40.66,p<0.001), and current prednisolone used≥15 mg/day (OR19.36,95% CI 2.29 – 163.66,p=0.007) were associated with SRC. Digital gangrene tended to show an association with SRC in the univariate analysis (OR8.00,95% CI0.72 – 88.22,p=0.090). Variables with p<0.15 in univariate conditional logistic regression analysis were included in the multivariate conditional logistic regression analysis. When multivariate conditional logistic regression analysis was performed, digital gangrene (adjusted odd ratio [AOR]31.41,95% CI 1.16 – 852.23,p=0.041), current prednisolone use≥15 mg/day (AOR31.22, 95% CI 1.59 – 613.85,p=0.024), serum albumin <3 mg/dl (AOR7.97, 95% CI 1.49 – 42.56,p=0.015) and cardiac involvement (AOR 6.62,95% CI1.08 – 40.63,p=0.041) were confirmed to be independent risk factors for SRC. During a median (IQR 1, 3) follow-up of 1 (0, 2) months, 15 (78.9%) patients required hemodialysis, including 9 of 12 (75.0%) patients with HRC and 6 of 7 (85.7%) with NRC. Twelve (63.2%) patients received ACEI, including 9 of 12 (75.0%) patients with HRC and 3 of 7 (42.9%) patients with NRC. Ten (52.6%) patients died, including 5 of 12 patients (41.7%) with HRC and 5 of 7 (71.4%) patients with NRC.
Conclusions: SRC was an uncommon complication in Thai patients with SSc, but associated with high mortality. Digital gangrene, current prednisolone used≥15 mg/day, serum albumin<3 mg/dl and cardiac involvement were independent risk factors of SRC.
References:
[1] Hudson M, et al.Semin Arthritis Rheum 2014.
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SAT0363 INTERSTITIAL LUNG DISEASE IN IDIOPATHIC INFLAMMATORY MYOPATHIES: A REPORT FROM THE REMICAM REGISTRY

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Background: Interstitial lung disease (ILD) in idiopathic inflammatory myopathies (IIM) may appear at any time in evolution and is associated with a worse prognosis.
Objectives: To investigate sociodemographics data and clinical characteristics of patients with IIM and ILD from the REMICAM registry.
Methods: A multicenter retrospective study (1980–2014) was performed. Patients were classified as polymiosytis (PM), dermatomyositis (DM), IIM with anti-synthetase antibodies (AB) and overlap syndrome. In addition, ILD was classified according to HRCT pattern: usual interstitial pneumonia (IP), non specific IP, organizing pneumonia, and acute IP. We compared sociodemographic data, clinical characteristics, AB and treatments of patients with and without ILD, patients with ILD according to subgroup of myositis or ILD.
Results: 478 patients were included, of whom 129 (27%) had ILD. Patients with ILD had a higher age at diagnosis, ESR and CRP and worse initial respiratory function. They were characterized by increased frequency of arthritis, systemic and cardiac manifestations, Raynaud, cardiovascular disease, pulmonary hypertension, ischemic ulcers, sclerodactyly and anti-synthetase AB (p<0.001). The differences among patients with ILD were: less arthritis in PM, greater frequency of Raynaud and sclerodactyly in overlap and antisynthetase syndrome, more anti-RNP AB in the overlap syndrome, more anti-synthetase and anti-Ro AB in the antisynthetase syndrome, and a higher prevalence of cutaneous signs and mechanical hands in DM (p<0.001). There were no differences according to the type of ILD or treatments.
Conclusions: We found some clinical manifestations and AB that may help for detection of patients with IIM associated to ILD, as well as clinical manifestations to better differentiate myositis subtypes in patients with ILD.
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SAT0364 HISTORY OF DIGITAL ULCERS IN PATIENTS WITH SYSTEMIC SCLEROSIS IS ASSOCIATED WITH PERIPHERAL VASCULOPATHY AS ASSESSED BY VIDEOCAPILLAROSCOPY AND 22-MHZ POWER DOPPLER ULTRASONOGRAPHY

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Background: Vasculopathy is considered a primary pathogenic event in systemic