DO PATIENTS WITH SYSTEMIC SCLEROSIS HAVE ULTRASONOGRAPHIC MODIFICATIONS OF SALIVARY GLANDS SUGGESTIVE OF SJOGREN SYNDROME?

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Background: Modifications in ultrasonographic aspect of major salivary glands have been reported in patients with primary Sjogren Syndrome (pSS) with good diagnostic accuracy. Sicca symptoms are frequently observed in Systemic sclerosis (SSc).

Objectives: To assess the ultrasonographic echostructure of major salivary glands in patients with SSc and compare the modifications with those of patients with pSS or controls with sicca symptoms.

Methods: We performed a monocentre case-control study between 2014 and 2017 in the university hospital of Clermont-Ferrand (France). Patients with SSc and pSS were fulfilling the American College of Rheumatology/European League against Rheumatism (ACR/EULAR) 2013 and the ACR 2012 classification criteria respectively. Controls patients were complaining of sicca symptoms but did not meet the ACR 2012 criteria. Bilateral parotid and submandibular glands ultrasound (US) were performed in all patients by the same operator blinded to the diagnosis. Inhomogeneity of each of the 4 major salivary glands in B-Mode was graded using the Jousse-Joulin scoring system (scale 0 to 4) as previously described.[1] The highest grade among the 4 glands was retained as suggestive of Sjogren Syndrome if ≥ 2.

Results: A total of 108 patients were included: SSc (n=25), pSS (n=48) and controls (n=35). Among the 48 patients with pSS, 12 were receiving hydroxychloroquine, 4 an immunosuppressor, 77% had antinuclear antibodies at a significant level (≥1/640), 26 (54%) anti-SSA antibodies, 14 (29%) anti-SSB antibodies, 40/45 (89%) had a labial salivary biopsy suggestive of pSS (Chisholm and Mason score ≥3). Comparing the pSS and the control groups, performance of a US echostructure grade ≥ 2 for the diagnosis of pSS was good: Se=75%, Sp=91.4%, positive predictive value= 92.3%, negative predictive value= 72.7%.

Among the 25 SSc patients, 7 had immunosuppressor therapy, 8 had a localized SSc. Shimr's test ≤ 5 mm in 5 minutes was present in 9/18 (53%), unstimulated salivary flow ≤ 0.1 ml/minute in 8/14 (57.1%). In the SSc group, 12 patients had an US echostructure grade 0, 6 grade 1, and 7 patients (28%) had an US echostructure grade ≥ 2: US score=3 (n=5), US score=4 (n=2). Anti-SSA antibodies were found in 1/7 patients with an US echostructure grade ≥ 2 and 2/18 patients with US echostructure grade 0 or 1.

Conclusion: Nearly one third of patients with SSc have US echostructure changes suggestive of Sjogren Syndrome regardless of the presence of anti-SSA antibodies.

REFERENCES:

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THU0329

RED FLAG SIGNS OF SYSTEMIC SCLEROSIS ARE PREVALENT IN SUBJECTS WITH RAYNAUD’S PHENOMENON IN THE GENERAL POPULATION AND MAY BE A PROXY FOR LUNG INVOLVEMENT

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Background: Pulmonary involvement in systemic sclerosis (SSc) is very difficult to treat when diagnosed too late. Therefore, in order to optimally use “the window of opportunity” more attention should be given to the early identification of SSc. To the best of our knowledge, no studies exist that have structurally assessed the epidemiology of red flag signs and potential signs of pulmonary involvement in patients with Raynaud’s phenomenon (RP).

Objectives: To assess the prevalence of red flag signs in participants with RP. Moreover, we aim to investigate the occurrence of pulmonary signs and symptoms in participants with red flag signs.

Methods: We retrospectively analyzed data from the LifeLines Cohort Study, which is a large population-based cohort study in the Northern parts of the Netherlands. A total of 74011 participants completed the connective tissue disease questionnaire. The presence of RP and red flag signs for SSc (i.e., puffy fingers, skin thickening distal, skin thickening proximal, and pitting scars) were obtained. Patients were classified as having red flag signs by the presence of at least one red flag sign in addition to RP. In addition, patient characteristics, self-reported pulmonary complaints, spirometry (screening for interstitial lung disease (ILD)), and uric acid (global screening for pulmonary arterial hypertension (PAH)) were also obtained. Three groups of participants were formed, namely: participants with RP and red flag signs (n=981), participants with RP without red flag signs (n=2946), and participants without RP and without red flag signs (n=70037).

Results: The prevalence of red flag signs was 5 fold higher in participants with RP, as compared to non-RP groups. RP 25% [23.7-26.4], non-RP 5% [4.9-5.2], p<0.001. A total of 413 (42.1%) of the participants with RP and red flag signs reported dyspnea, which was 1.5 fold higher than in those with RP but without red flag signs, and two-fold higher compared to participants without RP and without red flag signs (p<0.001). Moreover, dyspnea in rest and after exertion was more prevalent in participants with RP and red flag signs (p<0.001). In addition, participants with RP and red flag signs more frequently reported to suffer from pulmonary fibrosis (p<0.001, table 1), and had the lowest forced vital capacity, as compared to the other groups (p<0.001). Conversely, uric acid was not found to be elevated in participants with RP and red flag signs.

Conclusion: This unselected cohort study from the general population demonstrates that the prevalence of red flag signs in subjects with RP may be as high as 25%. Potential signs and symptoms of pulmonary complaints are more prevalent in participants with RP who also reported red flag signs. This could indicate an increased risk of pulmonary involvement (i.e., ILD and PAH) in RP patients with red flag signs, although additional specific tests are mandatory to substantiate definite disease.

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Abstract THU0329 – Figure 1. Prevalence of red flag signs in participants with and without Raynaud’s phenomenon.

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