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Pulmonary arterial hypertension and interstitial lung fibrosis in systemic sclerosis: One-stop shop assessment with cardiac and chest ultrasound

F. Frassi[1], M. Doveri[2], E. Vesprini[2], L. Gargani[1], MC. Scali[3], ML. Tafuri[1], ML. Bazzichi[2], S. Bombardieri[2], E. Picano[1]

[1]Cnr, Institute Of Clinical Physiology, Pisa, Italy; [2]Rheumatology Clinic, Pisa, Italy; [3]Siena University, Siena, Italy

Background: Interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH) are common complications of systemic sclerosis (SSc). Echocardiography evaluates PAH, and chest sonography detects even mild ILC as ultrasound lung comets (ULC), i.e. multiple comet-tails fanning out from the lung surface and originating from subpleural interlobular septa thickened by fibrosis.

Aim: to assess ILD and PAH by integrated cardiac and chest ultrasound in SSc.

Materials and methods: We enrolled 30 consecutive SSc patients (age=54±13 years, 23 females) in the Rheumatology Clinic of Pisa University. In all, we assessed Systolic Pulmonary Arterial Pressure (SPAP), from maximal velocity of tricuspid regurgitation flow, and ULC score with chest sonography (summing the number of ULC from each scanning space of anterior and posterior right and left chest, from second to fifth intercostal space). All patients underwent plasma assay for anti-topoisomerase antibodies (anti-Scl70), associated with development of pulmonary fibrosis. Twenty-eight patients also underwent High Resolution Computed Tomography, HRCT (from 0=no fibrosis to 3=honey combing).

Results: ULC number - but not SPAP - was correlated to HRCT fibrosis and presence SSc-70 antibodies (see figure). ULC number was similar in localized or diffuse forms (16±20 *vs.* 21±19, p=ns) and was unrelated to SPAP (r=0.216, p=ns).

Conclusions: Cardiac and chest sonography assessment of SPAP and ULC allow a complete, simple, radiation-free characterization of vascular and interstitial lung involvement in SSc - all in one setting and with the same instrument, same transducer and the same sonographer. In particular, ULC number, but not SPAP, is associated with HRCT evidence of lung fibrosis and presence of Scl-70 antibodies.