(modified Rondan total skin thickness score [mRSS], and percent predicted forced vital capacity [%FVC]), safety profiles, and outcomes after TCZ introduction regardless of TCZ continuation were extracted from the database.

Results: Of 404 patients enrolled in the database, 13 dcSSc patients were eligible for this study. Baseline characteristics included a mean age of 51 ± 9 years, 85% female, disease duration of 27 ± 24 months, and mRSS of 19.5 ± 10.6. Seven patients (54%) had HRCT-confirmed ILD at baseline, and 9 (69%) were positive for anti-topoisomerase I antibody. Two (14%) and 11 (85%) were on mycophenolate mofetil and low-dose prednisolone (72 ± 6.0 mg/day), respectively. Seven patients (54%) each had active skin disease and elevated inflammatory markers defined in the phase III clinical trial [2], while only 4 (31%) fulfilled the inclusion criteria. TCZ was initially administered intravenously (8 mg/kg every 4 weeks) in 8 patients and subcutaneously in 5 (162 mg every 2 weeks and every week in one). At one year, mRSS was improved from 20.9 ± 11.4 to 10.7 ± 8.9 in 11 patients (p < 0.007), and 0%FVC was stable in 7 patients with ILD (75.8 ± 15.0 to 78.6 ± 16.1). During the observation period of the first 26 ± 28 months, 4 patients were treated with a stable dose of TCZ, while TCZ dose was reduced and/or discontinued in 9. Four of them discontinued TCZ due to adverse events (n = 2), acute lung injury and pleghmon) or prominent improvement of skin thickening (n = 2). Of 9 patients with dose reduction/discontinuation of TCZ, 4 patients who discontinued TCZ (n = 3) or received dose reduction of TCZ (n = 1) experienced a recurrence of progressive skin thickening together with inflammatory complications, including edematous induration of the skin, progression of ILD, polyarthralgia, and/or pericarditis with increased inflammatory markers. The interval between dose-reduction/discontinuation of TCZ and clinical worsening ranged from 2 to 11 months. These manifestations were promptly improved by dose-escalation or resumption of TCZ in all patients except one who experienced progressive ILD and died of respiratory failure 27 months later.

Conclusion: In dcSSc patients who experienced improvement of skin thickness during treatment with TCZ, dose-reduction or discontinuation of TCZ may result in a recurrence of the disease. Randomized controlled studies are necessary to examine optimal timing for dose-reduction or discontinuation of TCZ in dcSSc patients after improvement of skin thickness.

REFERENCES:


AB0437
OESOPHAGEAL DILATATION ON HIGH-RESOLUTION CT CHEST IN SYSTEMIC SCLEROSIS: SIGNIFICANT INDEX?
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Background: Systemic sclerosis (SSc) is chronic, autoimmune multisystem disorder presented by thickening and fibrosis of the skin and internal organs. Esophageal involvement is one of the most common manifestation. Esophageal enlargement of HRCT on chest is a common finding in scleroderma patients and may also be associated with other sclerosis-related clinical findings [1].

Objectives: The aim of this study to evaluate the association between esophageal dilatation on chest HRCT at diagnosis with the other SSc features.

Methods: The study was planned for SSc patients registered between October 2007 and September 2020 in Dokuz Eylul University Rheumatology Department database. Demographics, clinical features and medical history were recorded. The presence of HRCT on chest was compared with the presence of esophageal dilatation. Then, the initial HRCT images were assessed for esophageal dilatation by an experienced chest radiologist according to recommendation of Pirz et al.

Results: In our study, there were 233 SSc patients (f:206 M:27, mean age 59.9±12.7 years) 71 (31.4%) of them diagnosed with diffuse disease. Median follow-up of study was 73 (1-272) months. Esophageal dilatation on HRCT was detected in 60 (26%) of the 233 SSc patients (43.2%). Had proof of esophageal involvement in esophageal transit scintigraphy. There is no statistical correlation was found between esophageal dilatation on HRCT with gender, smoking, arthrosis, pulmonary hypertension and autoantibody subtypes.

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AB0438
SERUM HOMOCYSTEINE AND BONE STATUS IN SYSTEMIC SCLEROSIS PATIENTS
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Background: Systemic sclerosis (SSc) is a complex autoimmune connective tissue disease characterized by self-amplifying microvascular damage sustained by autoimmune response and progressive skin and visceral fibrosis. Besides, SSc patients show higher incidence of bone micro/macrophageal changes and bone fractures. Emerging data demonstrate that high serum levels of homocysteine (Hcy) could modulate osteoclastogenesis and are recognized as risk factors for osteoporosis (2). Furthermore, serum levels of Hcy were found to be higher in SSc patients than in healthy controls (3).

Objectives: to evaluate the bone status according to Hcy serum levels in a cohort of SSc patients.

Methods: 20 female patients fulfilling ACR 2013 criteria for SSc underwent a dual-energy X-ray absorptiometry scan (DXA) (Lunar Prodigy) to evaluate bone status. We analysed bone quantity and quality respectively by bone mineral density (BMD) and trabecular bone score (TBS). According to the WHO criteria, osteoporosis was defined as a bone density of 2.5 standard deviations below that of a young adult (T-Score). Fasting blood samples were obtained from all patients in order to test serum Hcy level and bone turnover markers after obtaining the informed consent. All subjects underwent morphometric spine X-Ray to evaluate vertebral fractures. Statistical analysis was performed using non-parametric tests.

Results: The mean age of patients was 64.15 ± 10.8 years with a mean disease duration of 9.1 ± 2.3 years. The mean modified Rodnan Skin Score (mRSS) was 10.7 ± 8.5. All patients showed a "scleroderma pattern" at nailfold videocapillaroscopy (NVC); in particular, 7 patients showed the "Late" pattern, 9 patients the "Active" pattern and 4 patients the "Early" NVC pattern. Hyperomocisteinemia (HHCy) was found in 25% of patients. Interestingly, SSc patients with the "Late" NVC pattern showed a significantly higher serum level of Hcy compared to the "Early/Active" group (11.5 ± 4.4 vs 17.1 ± 6.4, p=0.03). No significant differences were observed in relation to the autoantibody profiles. Of note, 60% of patients with HHcy were found osteoporotic and 40% had bone fractures.