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Scleroderma: Clinical and immunological profile

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❑Introduction:

➢Systemic scleroderma (Scl) is a rare disease of unknown etiology.
➢It belongs to the connectivitis group.
➢It is characterized by involvement of arterioles and micro-vessels, and connective tissue.
➢The disease causes life-threatening cutaneous and/or visceral sclerosis lesions.
➢Biological markers of autoimmunity are also present.
➢Various immune system abnormalities are illustrated by the presence of highly specific antibodies.
➢Some antibodies are specific to certain associated manifestations.

❑Aim:

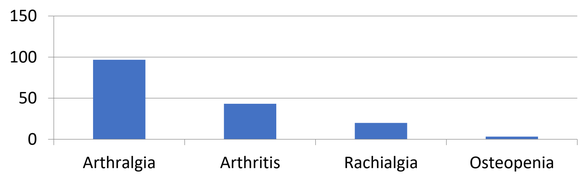
➢The objective is to determine the clinical, para-clinical, immunological, evolutionary, and therapeutic characteristics of scleroderma.
➢This is done through a retrospective study.

❑Materials and Methods:

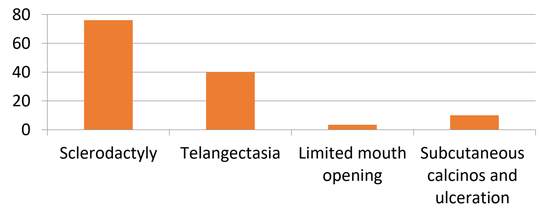
➢This is a retrospective study including 30 patients followed in a rheumatology department.
➢They presented with a clinico-biological or histological picture suggestive of scleroderma.
➢The study spans a 23-year period from 2000 to 2023.

❑Results:

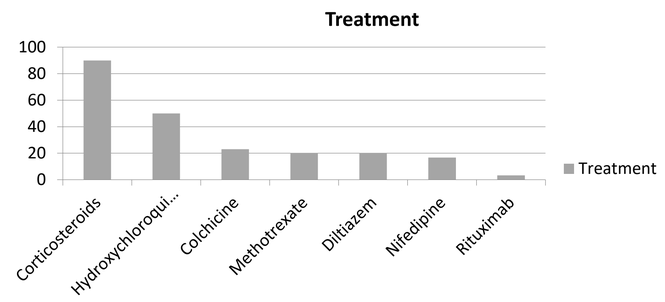
➢Thirty patients were included: 6 men (20%) and 24 women (80%).
➢The mean age was 44 years, ranging from 10 to 81 years.



***Osteoarticular manifestations:**
***Raynaud's phenomenon:** Observed in 76.6% of patients.
***Respiratory symptoms:**
-43% had NYHA stage II–III exertional dyspnea.
-Chest X-rays showed reticulo-micronodular opacities in 38%, suggestive of interstitial syndrome.
-Chest CT confirmed diffuse interstitial pneumopathy (DIP) in 46.7% of patients.
-Restrictive EFR syndrome was found in 20% of cases.
***Muscular involvement:**
-Found in 26.7% of patients.
-Symptoms included diffuse myalgia and increased muscle enzymes.
***Digestive involvement:**
-36.7% had dysphagia.
-6.7% had eso-gastric involvement (congestive and erosive gastropathy, esophagitis).
-Esophageal manometry showed hypotonia of the lower esophageal sphincter and reduced peristalsis in 13.3% of patients.
***Neurological involvement:** Found in 10% of cases.
***Pulmonary arterial hypertension (PAH):** Found in 16% of patients.
***Cardiac involvement:**
-Pericardial involvement in 6.7%.
-Myocardial involvement in 3.3%.
***Skin manifestations:**



***Capillaroscopy findings:** Disorganization and rarefaction of the periungual capillary bed in 43% of patients.
***Biological findings:**
-Inflammatory syndrome in 49% of cases.
-Normocytic normochromic anemia in 33%.
-Renal function was satisfactory in 96% of patients.
-One patient had proteinuria elevated to 0.5 g/l.
***Immunological findings:**
-35% of patients had positive anti-topoisomerase antibodies.
-These were significantly associated with Raynaud's phenomenon (p=0.037), digestive disorders (p=0.028), and DIP (p=0.05).
-No correlation was found between anti-Scl70 and neurological disorders (p=0.16), muscular disorders (p=0.4), PAH (p=0.6), or arthritis (p=0.06).
***Treatment:**
***Evolution:**



Partial improvement was seen in Raynaud's phenomenon, arthralgia, and general symptoms.
83% showed improvement of the biological inflammatory syndrome.
Dyspnea worsened in 3.3% of cases.
One patient died.

❑ Conclusion:

➢These results highlight the complexity and variability of clinical, biological, and immunological manifestations in systemic scleroderma.
➢Anti-topoisomerase I antibodies were found in 35% of patients.
➢Their investigation is useful not only for diagnosis but also to predict associated phenotypes.
➢These antibodies help identify patients at risk of severe forms of Scl.
➢This supports better follow-up and prevention of complications.