AUTOIMMUNE CONNECTIVE TISSUE DISEASES

CREST SYNDROME

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Background: CREST syndrome is a limited subtype of systemic sclerosis variant consisting of calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, teleangiectasis. Therefore, because the diagnosis of this syndrome is difficult to distinguish from diffuse subtype of SS, requiring complete history taking, physical and support examination.

Case: A 41 years old female with black and white spots are hardened skin on face, chest, stomach, back, hands and feet, red spots on nose, purplish's fingertips and stiffness, difficulty swallowing and indigestion. Physical examination shows Raynaud's phenomenon, esophageal dysmotility, sclerodactyly and teleangiectasis. Supporting examinations wih histopathological shows dense of fibrocollagen in dermis and hyalinization of subcutis, X-ray on superior extremities with calcification of blood vessels, X- ray on thorax with pulmonary fibrosis, anti Scl-70 was negative and endoscopy with esophagitis This patient has given therapy of methotrexate 7,5 mg/week injection, lansoprazole 30 mg/d, sucralfate 100 ml thrice a day, prednisone 20 mg/d and nifedipine 30 mg/d, soft u derm cream apply twice daily.

Discussion: Systemic scleroderma is a multisystem disease with microvascular abnormalities, autoimmune disorders, excessive production of collagen and fibrosis in the skin with visceral organ involvement. Scleroderma systemic has 2 forms, there are diffuse and limited. Syndrome of CREST is subtype of limited systemic scleroderma. Establishing of CREST syndrome in this patient findings calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly and teleangiectasis and the patient has improvement after given therapy in 2 months.

Keywords: Crest syndrome, systemic sclerosis