International Journal of Research and Review
Vol. 9; Issue: 7; July 2022
Website: www.ijrrjournal.com
Case Report *E-ISSN: 2349-9788; P-ISSN: 2454-2237*

The Challenges of Diagnosis and Management of Geriatrics with Systemic Sclerosis: A Case Report

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DOI: https://doi.org/10.52403/ijrr.20220701

ABSTRACT

Scleroderma (systemic sclerosis) is a rheumatic autoimmune disease characterized by skin fibrosis, internal organ damage, and systemic vasculopathy. Scleroderma in elderly patients presents with non-specific symptoms that overlap with another medical condition.

A woman, 63 years old, came with a chief complaint of thickening and darkening of the skin since ten years ago. The patient also complained of shortness of breath and decreased urine in 3 weeks. On physical examination, the patient was having ascites. Multiple diffuse hyperpigmented in the thoracal, abdominal, and extremities areas. Based on the 2013 ACR/EULAR (American College of Rheumatology; European League Against Rheumatism) classification criteria, the patient was diagnosed with scleroderma with systemic sclerosis, overlapping with other medical conditions such as diabetes mellitus, chronic kidney disease stage V, urinary tract infection, and hypoalbuminemia.

Most of the systemic symptoms in systemic sclerosis patients were not specific. The progressive cardiopulmonary disease is critical in systemic sclerosis, especially in geriatrics. Interstitial lung disease can progress to pulmonary hypertension as a late and terminal complication. It is recommended for routine screening for interstitial lung disease.

Keywords: Scleroderma, systemic sclerosis, geriatric.

INTRODUCTION

Scleroderma (systemic sclerosis) is a rheumatic autoimmune disease characterized by skin fibrosis, internal organ damage, and systemic vasculopathy. Scleroderma can occur at any age. The incidence of scleroderma has a peak onset of 50 to 60 years.[1] Scleroderma in elderly patients presents a complex systemic disease challenge that overlaps with other chronic medical conditions whose incidence also increases with age. The dangers of polypharmacy, frailty, and nutrition are all of the great importance when dealing with a potentially damaging disease as an organ system.[2]

Here we report the case of an elderly with scleroderma. This case can increase understanding of risk factors, proper diagnosis, and management to reduce morbidity and mortality.

CASE ILLUSTRATION

A woman, 63 years old, came with the chief complaint of darkening of the skin on her hands, feet, head, and body since ± ten years ago. Complaints begin with itching all over the skin; then, the skin feels thickened, stiff, and darkened. In the morning or when the weather is cold, the patient's fingers and toes often turn pale or blue until the knuckles on their feet fall off on their own. The patient also complains of pain in the entire low back that does not radiate and improves with changes in position and when lying down. The patient also complains of shortness of breath that comes and goes and worsens with activity and the supine position. Complaints of shortness of breath

improved with oxygen and sitting position. The color of the bowel is said to be black; the urine is decreased in 3 weeks. The patient has had a history of type 2 diabetes mellitus.

On physical examination, the patient was conscious; vital signs were within normal limits. The abdominal were ascites. Multiple diffuse hyperpigmented macules were demarcated, geographically shaped, and covered by thin scales and xerotic scars in the thoracal, abdominal, and extremities area (Figure 1.).

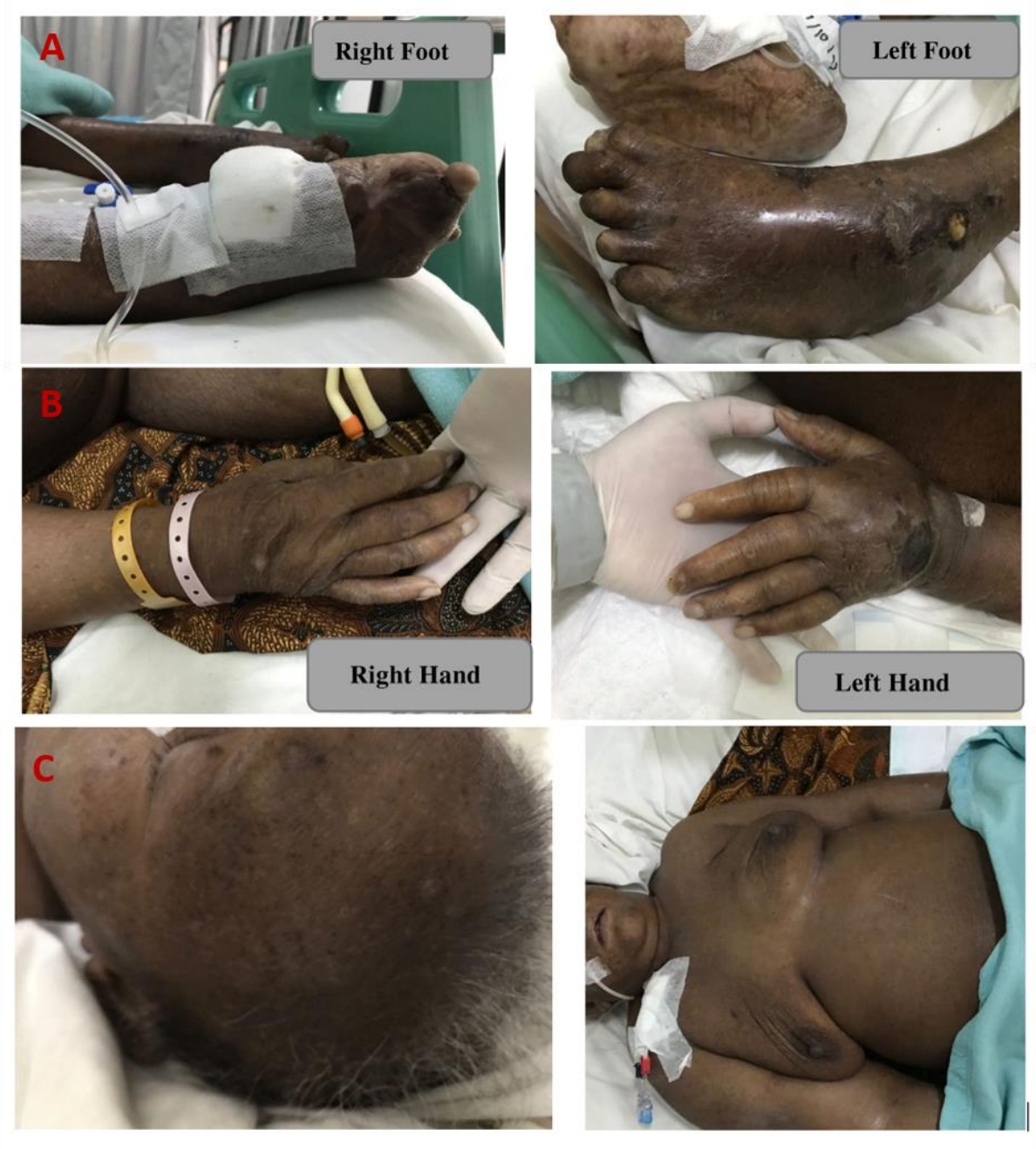

Figure 1. Multiple diffuse hyperpigmented macules were demarcated, geographically shaped, and covered by thin scales and xerotic scars in the head, thoracal, abdominal, and extremities area.

Complete blood count showed leukocytosis with a predominance of neutrophils with mild normochromic normocytic anaemia. Hypoalbuminemia (1.7), with elevated renal function (BUN 32.3 mg/dL and Creatinine 4.8 mg/Dl). Complete urine examination showed urinary tract infection (leukocytes ++, protein +++, ketones ++, erythrocytes

+++, sedimentary leukocytes 8-12). ANA-IF test of patient 1:100. Roentgen of the thorax revealed cardiomegaly with congestive lung (Figure 2.). Ultrasound of the abdomen showed a bilateral chronic renal disease. The tissue biopsy results from the skin provide a morphological picture of scleroderma.

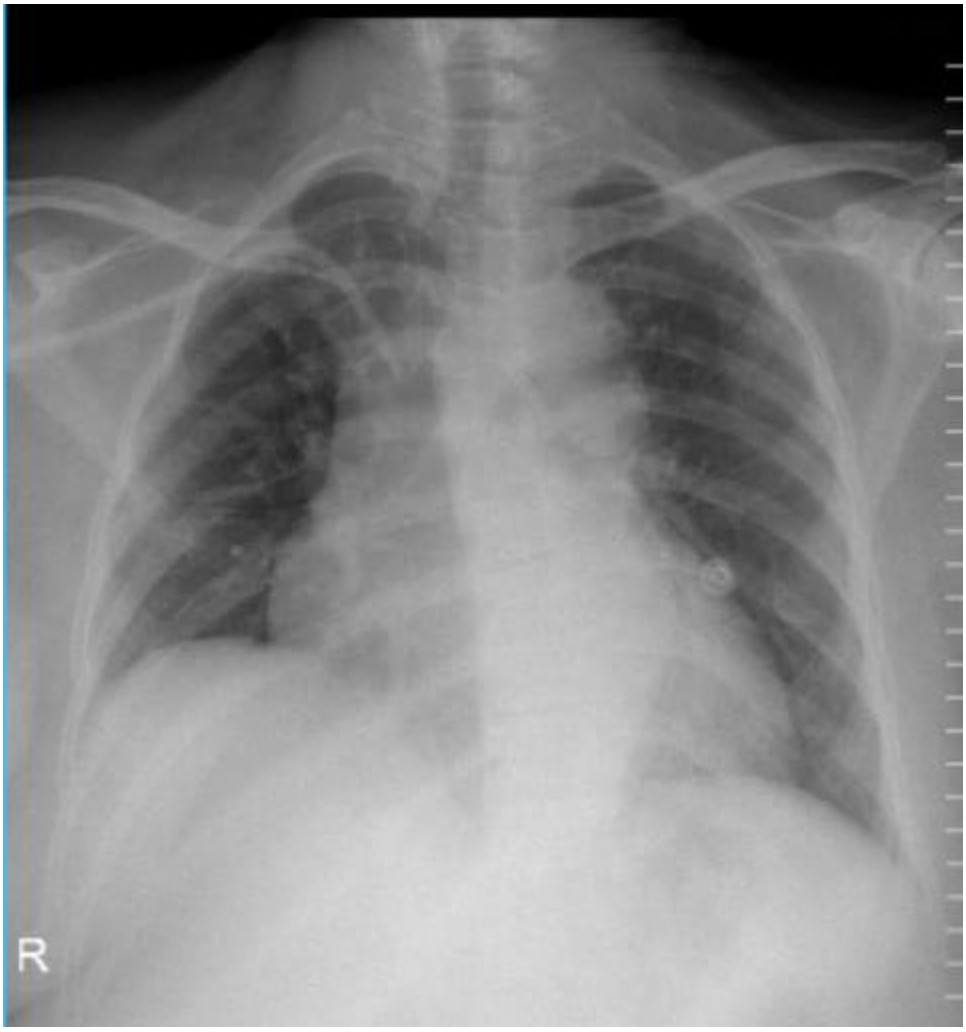

Figure 2. Roentgen of thorax showed cardiomegaly with congestive lung.

Based on the 2013 ACR/EULAR classification criteria, the patient was diagnosed with scleroderma with systemic sclerosis. The patient was also diagnosed with type 2 diabetes mellitus, chronic kidney disease (CKD) stage V, urinary tract infection, and hypoalbuminemia due to chronic inflammation. During treatment, the patient was treated with albumin transfusion with target serum albumin 2.5g/dL and insulin. Systemic antibiotic, Cefoperazone 1 gram every 12 hours IV, Captopril 25 mg every 12 hours orally, CaCo3 500 mg/8 hours orally, and planned to undergo elective hemodialysis 2x/week. From the dermatologist, patients were given urea 10% cream every 12 hours on dry skin lesions. From rheumatology, give therapy methylprednisolone 3x4mg orally. The patient was hospitalized for ten days and later died from complications of sepsis.

DISCUSSION

Systemic sclerosis with the subtype of diffuse cutaneous scleroderma (dcSSc) has various degrees of cutaneous sclerosis, such as thickening of the skin on the fingers and the proximal areas of the elbows and knees, trunk, and face - patients with new-onset dcSSc usually present with Raynaud's phenomenon and swelling and pain in the extremities.[3] In addition, it can also be

accompanied by weight loss, decreased appetite, and increased inflammatory markers. Most of the time, patients with early symptoms of dcSSc are diagnosed with rheumatoid arthritis, peripheral oedema, heart failure, and carpal tunnel syndrome.[4] The skin changes in dcSSc are characterized by oedema of the feet and hands. As the disease progresses, a deep fibrotic process occurs, causing the skin to become thicker and lose its natural lines and wrinkles. Skin lesions are usually found on the fingers (sclerodactyly) and then increase in proximal areas of the body, namely the trunk and face. In this patient, diffuse skin lesions were found in the thoracic, anterior, and posterior abdominal regions, palms, and soles. Based on the skin lesions, the scleroderma was classified as a diffuse type of scleroderma subtype (dcSSc).

Systemic sclerosis not only affects the skin but can also affect internal organs, such as cardio-pulmonary, gastrointestinal, renal, and musculoskeletal.[5] Most patients with scleroderma will develop pulmonary fibrosis, from mild and asymptomatic to fatal. The prevalence of pulmonary hypertension in scleroderma is estimated at 8%, with an incidence of 0.61 cases per 100 patients. Symptoms of pulmonary hypertension can be mild, such as shortness of breath, which can lead to right heart failure, which causes oedema of the lower extremities and increased jugular pressure.[6] Decreased isolated diffusive capacity for carbon monoxide on pulmonary function tests may indicate increased pulmonary pressure. Most of the systemic symptoms in systemic sclerosis patients were not specific; it is recommended for routine screening for interstitial lung disease.[7] In our case, the patient complained of intermittent shortness of breath. On chest X-ray examination, pulmonary congestion and cardiomegaly were found.

When the diagnosis of scleroderma is made in the elderly, recognizing the typical symptoms and course of the disease will help the clinician determine therapeutic strategies, choose the proper examination to make the diagnosis, and educate the patient and his family.[8] The diagnosis should be made as quickly as possible, and routine monitoring of pulmonary vascular disease should be carried out through pulmonary function tests and echocardiography. The diagnosis of systemic sclerosis was made based on the 2013 ACR/EULAR classification criteria. If sclerosis of the fingers of both hands is found in the proximal part of the metacarpophalangeal joints, then a score of 9 is sufficient to diagnose systemic sclerosis.[9] Other positive symptoms include sclerodactyly, lesions of the fingertips (ulcers, scars), telangiectasis, abnormal nail fold capillaries, pulmonary hypertension, Interstitial Lung Disease, and Raynaud's phenomenon, and positive systemic sclerosis-related antibodies will be added to the scoring system. In this case, the total score is 15.

Until now, no studies have precisely described the treatment of elderly patients with scleroderma. In patients with progressive lung disease, administering immunosuppressants such as cyclophosphamide is standard therapy. Cyclophosphamide can be given as a monthly infusion or daily oral therapy. This therapy is given at low doses for several months, followed by administering Mycophenolate or Azathioprine to reduce side effects. Immunosuppressive therapy should only be given for one year. However, relapses occur frequently, so long-term therapy is sometimes given.[7,8]

In patients with renal crises, the administration of ACE inhibitors has been shown to reduce disease progression.[6,7] Immunosuppressive agents can also be administered to treat skin lesions. Some new therapies currently being investigated are intravenous Gamma globulin, Rituximab, Tyrosine kinase inhibitors, and other biologic genes that block pro-fibrotic cytokines such as anti-IL 13 and anti-TGF beta. In addition to pharmacological therapy, non-pharmacological therapy is essential in treating skin lesions by

preventing sunburn, topical emollients, and routine wound care.[6]

Scleroderma mortality increases with age. Another study found that the in-hospital mortality rate in patients with scleroderma showed a 15% increase mortality every ten years.[8] In a 2020 American study found that the most common severe infection in patients with scleroderma was pneumonia (45%) and sepsis (32%), followed by soft tissue infections (19%), urinary tract infections (3%), and opportunistic infections (3%). The hospital mortality rate in scleroderma patients with severe infections is 9%.[9]

CONCLUSION

Systemic sclerosis mortality increases with age. To date, there are no specific studies describing the treatment of systemic sclerosis in geriatrics. Recognizing and understanding the specific symptoms will help determine the treatment for the patient. Most of the systemic symptoms in systemic sclerosis patients were not specific. Pulmonary complications such as pulmonary hypertension and pulmonary fibrosis are the main causes of death in scleroderma patients. It is recommended for routine screening for interstitial lung disease.

Acknowledgement: None

Conflict of Interest: None

Source of Funding: None

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How to cite this article: IGNG Bayu Prawira Putra, RA Tuty Kuswardhani. The challenges of diagnosis and management of geriatrics with systemic sclerosis: a case report. *International Journal of Research and Review*. 2022; 9(7): 1-5. DOI: *https://doi.org/10.52403/ijrr.20220701*

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