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RAYNAUD'S PHENOMENON IN A PATIENT WITH SCLERODERMA: A CASE REPORT

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ABSTRACT
Systemic sclerosis, or scleroderma, is a chronic autoimmune disorder characterized by fibrosis of the skin and internal organs, vasculopathy, and immune dysregulation. Raynaud's phenomenon (RP), marked by episodic vasospasm of the digital arteries in response to cold exposure or emotional stress, is often one of the earliest and most common sign of SSc. Early diagnosis plays a crucial role in preventing and managing complications of RP, such as digital ulcers and tissue damage due to chronic circulatory impairment, as well as in reducing long-term disability. This case report aims to highlight pedal ulcers as a complication of RP scleroderma and to emphasize the importance of early intervention in preventing progressive tissue damage. The data was conducted through a comprehensive approach including history assessment, physical examinations, electronic medical record reviews, laboratory diagnostics, and imaging studies to ensure accurate diagnosis and effective interventions. We report a 26-year-old woman who presented with a six-month history of progressive skin thickening and hardening involving almost all of her body. The patient also experienced finger stiffness, flexion contractures, and limited mouth opening. Dermatological examination revealed sclerotic skin with hypopigmented and hyperpigmented patches forming a "salt-and-pepper" appearance across various body areas, along with microstomia and a beaked nose. The Modified Rodnan Skin Score (mRSS) was 34. A diagnosis of systemic sclerosis was established based on the American College of Rheumatology in collaboration with the European League Against Rheumatism (ACR/EULAR) criteria. The patient was lost to follow-up after the initial visit and later returned with toe necrosis and ulceration over the ankle area. Treatment included systemic corticosteroids combined with methotrexate, along with counseling to avoid cold exposure and manage stress. Raynaud's phenomenon should be recognized as a critical early sign of scleroderma. Comprehensive early evaluation, including autoantibody testing, is essential for timely diagnosis and the prevention of severe vascular complications.

Keywords: autoimmune disease; raynaud's phenomenon; systemic sclerosis; vasculopathy

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INTRODUCTION

Raynaud's phenomenon is one of the earliest and most common sign of systemic sclerosis. Raynaud's phenomenon is an episodic vascular disorder characterized by vasospasm of small arteries, primarily in the distal extremities, leading to progressive skin color changes in response to cold temperature or emotional stress. This condition can occur idiopathically (primary) or be associated with systemic diseases such as scleroderma (secondary). In patients with scleroderma, Raynaud's can be more severe and persistent, potentially leading to digital ulcers and tissue damage due to chronic blood flow impairment. (Maciejewska et al., 2022). Scleroderma, or systemic sclerosis, is an autoimmune disease marked by tissue fibrosis, vascular endothelial dysfunction, and immune activation. Approximately 90% of patients with scleroderma experience Raynaud's phenomenon as an initial symptom. Therefore, this phenomenon serves as an important indicator for the early diagnosis of systemic connective tissue diseases (Cutolo et al., 2024).

The classification criteria for systemic sclerosis were recently updated by a joint committee of the American College of Rheumatology and the European League Against Rheumatism to provide more specific diagnostic guidelines, enabling timely treatment before irreversible organ damage occurs (Van den Hoogen et al., 2013). Histopathological examination of skin or affected tissue can provide confirmatory evidence by demonstrating hallmark features of scleroderma, reflecting chronic vascular endothelial damage and fibroblast activation, which underlie the pathophysiology of both scleroderma and its associated Raynaud's phenomenon (Domsic & Medsger, 2021).The diagnosis of Raynaud's phenomenon in scleroderma patients is established through a multimodal approach. Clinically, evaluation includes a history of vasospastic attacks and nailfold capillaroscopy, which reveals characteristic findings such as capillary enlargement, microhemorrhages, and avascular areas. Serological tests for antinuclear antibodies (ANA), anti-centromere, and anti-topoisomerase I (Scl-70) antibodies further support the diagnosis (Maciejewska et al., 2022). Management of Raynaud's phenomenon in scleroderma involves both non-pharmacological and pharmacological therapies. Non-drug strategies include lifestyle modifications, while medical interventions encompass the use of calcium channel blockers, prostacyclins, phosphodiesterase-5 inhibitors, and botulinum toxin, all of which have shown clinical benefits (Pang et al., 2025).This case report aims to describe the clinical manifestations, diagnostic process, and management of scleroderma, while also highlighting pedal ulcers and toe necrosis as vascular complications resulting from Raynaud's phenomenon associated with the disease.

METHOD

This article is a case report study that provides diagnosis, clinical management, and patient follow up care. Data from this case report were obtained through anamnesis, physical examination, and supporting examinations conducted at Adam Malik General Hospital, Medan. The data obtained were then analyzed qualitatively and presented in narrative form. This case report discusses a a 26-year-old woman who experienced schleroderma with Raynaud's phenomenon. This case report will analyze individual case, and is expected to provide insights into clinical practice, especially regarding the problem of scheroderma with Raynaud's phenomenon.

RESULT

A 26-year-old woman presented to the Dermatology and Venereology Outpatient Clinic at H. Adam Malik General Hospital, Medan, with complaints of skin stiffness and hardening accompanied by pain involving the face, neck, chest, abdomen, back, both arms, and lower limbs over the past six months. She also reported finger stiffness with flexion deformities and difficulty fully opening her mouth while eating, which interfered with daily activities. The symptoms began two years prior with the appearance of white patches and dry skin that gradually spread to nearly the entire body, although she had never sought medical treatment. Over time, the skin tightening and stiffness worsened, prompting her to seek medical attention. There was no family history of similar symptoms.On physical examination, the patient appeared moderately to severely ill, with full consciousness (compos mentis). Her vital signs were as follows: blood pressure 102/61 mmHg, pulse rate 92 beats per minute, respiratory rate 20 breaths per minute, body temperature 36.8°C, and oxygen saturation (SpO_2) 98%. She weighed 57 kg and was 156 cm tall. Dermatological examination revealed sclerotic skin with areas of hypopigmentation and hyperpigmentation forming a characteristic "salt and pepper" appearance on the facial region, posterior neck, bilateral forearms, bilateral hands, anterior and posterior trunk, thighs, shins, and feet (Figure 1). Additional findings included microstomia, indicated by radial furrowing around the mouth, and a beaked nose, referring to thinning and projection of the nasal tip resembling a bird's beak.

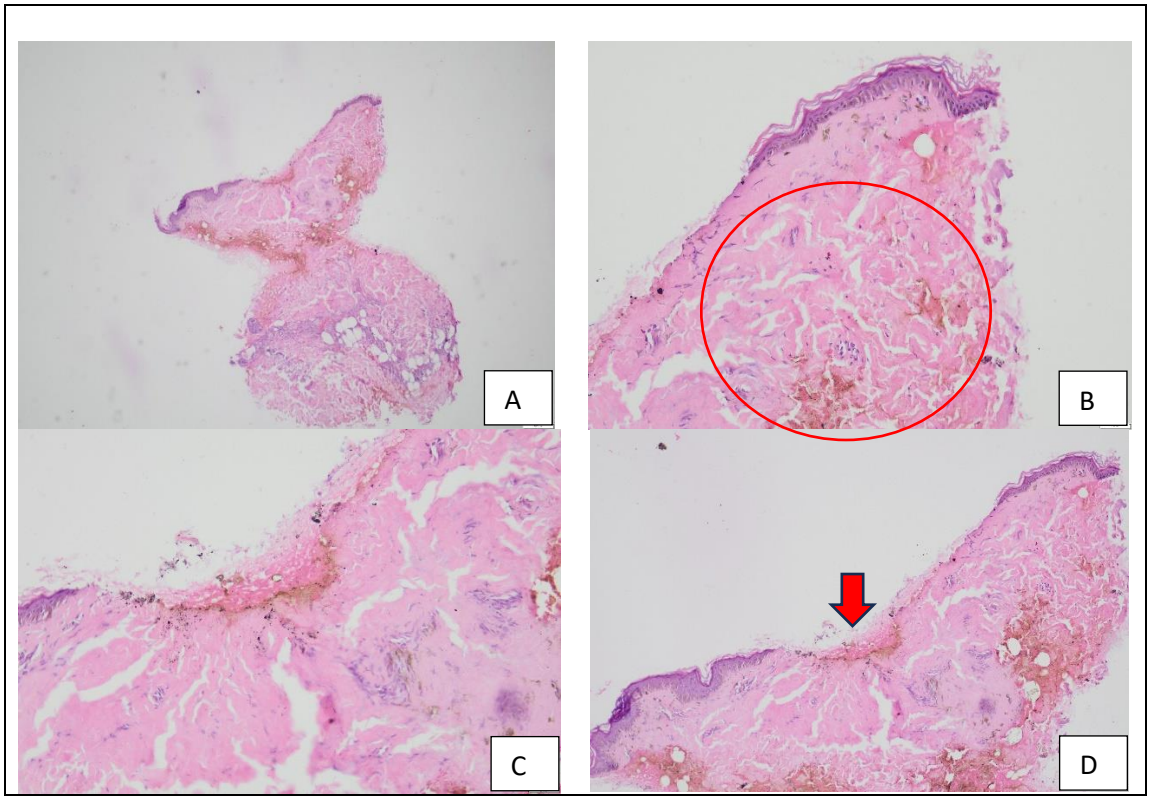

Figure 1. (A, B, C): "Salt and pepper" pigmentation on the facial region, beaked nose, microstomia, and mask-like facial stiffness.(D, E, F, G, H, I, J, K): Sclerotic skin and "salt and pepper" pigmentation on the posterior neck (colli posterior), bilateral forearms (antebrachii), bilateral hands (manus), anterior and posterior trunk, thighs (femoris), shins (tibialis), and feet (pedis). (L): Puffy hands and sclerodactyly.

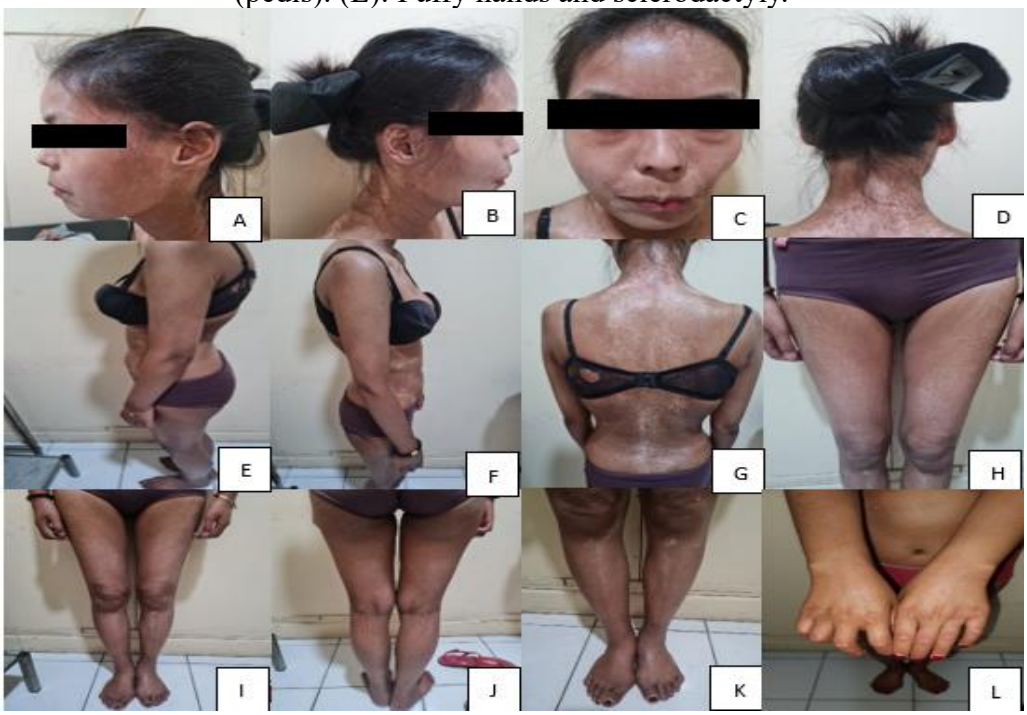

Figure 2. Histopatological examination microscopic examination of the biopsy specimen revealed thinned and discontinuous squamous epithelium, with preserved cellular morphology. (A) 40× magnification. (B) The subepidermis is dominated by fibrocollagenous connective tissue infiltrated by lymphocytic inflammatory cells. (C, D) Minimal interstitial hemorrhage in the erosive epithelial area.

The total mRSS for this patient was 34. Laboratory investigations were conducted to assess systemic involvement and support the diagnosis. Urinalysis revealed the presence of leukocytes and bacteria, indicating a possible urinary tract infection. Complete blood count, liver function tests, renal function tests, and random blood glucose levels were all within normal limits. Immunological testing showed a positive antinuclear antibody (ANA) result with a titer of 127.7 IU/mL (normal value: < 20 IU/mL), while anti-dsDNA was 24.8 IU/mL (normal range: 0–200 IU/mL). The Histopathological examination showed in figure 2.

Differential diagnoses for this patient was scleroderma include morphea and vitiligo. The diagnosis of systemic sclerosis is based on anamnesis, physical examination, and histopathological findings. According to the 2013 American College of Rheumatology/European League Against Rheumatism (ACR-EULAR) criteria, this patient scored 15.The patient was managed with education and pharmacological therapy. She was advised to reduce stress and avoid cold exposure. The prescribed treatment included oral methylprednisolone 12 mg/day that planned to tappering off based on patient's clinical improovement, omeprazole 20 mg twice daily, paracetamol 500 mg three times daily, and urea-based moisturizing cream applied to dry skin twice daily. The patient was scheduled for follow-up every week to check on patient clinical improovement.However, after 5th follow up, the patient did not attend the follow-up appointment again and reported self-medicating by purchasing drugs at a pharmacy. She returned four months later with complaints of hardened skin and purulent ulcers on the right ankle and foot, accompanied by pain that impaired walking and limited movement of the fingers.

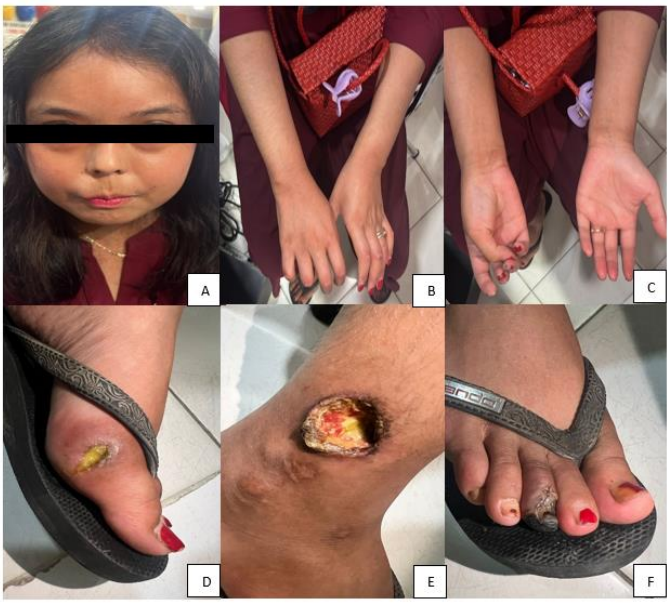

Figure 3. (A) Beaked nose, microstomia, and moon face. (B, C) Puffy hands, sclerodactyly, and multiple ulcerations on the right fourth finger (digit IV manus dextra). (D, E) Ulcers and pus associated with the right foot (pedis dextra) and lateral malleolus. (F) Necrosis caused by Raynaud's phenomenon on the right third toe (digit III pedis dextra).

On physical examination, the patient appeared moderately ill but was alert (compos mentis). Vital signs were: blood pressure 110/70 mmHg, pulse rate 96 beats per minute, respiratory rate 20 breaths per minute, body temperature 36.6°C, and oxygen saturation (SpO_2) 98%. Her weight was 59 kg with a height of 156 cm. Moon face was observed on the face.Dermatological examination revealed sclerotic skin with hypopigmentation and hyperpigmentation in a "salt and pepper" pattern on the facial region, posterior neck, bilateral

forearms, bilateral hands, anterior and posterior trunk, thighs, shins, and feet, which had decreased and thinned compared to previous examination. An ulcer was found on the fourth digit of the right hand. Ulcers with pus and necrosis were observed on the lateral malleolus and right foot, including necrosis on the third digit of the right foot (Figure 3). Microstomia, characterized by radial perioral furrowing, and a beaked nose were also present with mRSS score was 31.

The current diagnosis for the patient is systemic sclerosis with suspected pyoderma gangrenosum and necrosis caused by Raynaud's phenomenon affecting the right third toe (digit III pedis dextra). The patient was prescribed oral methotrexate 10 mg twice daily on Saturdays for one month and methylprednisolone 8 mg daily for two weeks. Pus culture results revealed *Pseudomonas aeruginosa*, with antibiotic susceptibility testing showing resistance to several antibiotics, including cefazolin, gentamicin, ciprofloxacin, and tigecycline. The patient remained sensitive to piperacillin, cefepime, ceftazidime, aztreonam, meropenem, and amikacin.Management included consu ltation with the vascular surgery department for further treatment of the necrosis on the right third toe. Wound care education was provided, recommending 0.9% NaCl compresses for 15 minutes every 4 hours.

DISCUSSION

Scleroderma is a multisystem disease characterized by autoimmune processes, vascular endothelial cell injury, inflammation, and extensive fibroblast activation. Epidemiologically, females are more commonly affected by systemic sclerosis, with a female-to-male ratio of approximately 3:1, and it frequently occurs between the ages of 30 and 50 (Gudjonsson JE et al., 2019).Systemic sclerosis is classified into limited cutaneous systemic sclerosis (lcSSc) and diffuse cutaneous systemic sclerosis (dcSSc). In dcSSc, skin thickening extends beyond the elbows and may involve the trunk, face, and extremities, typically within the first one to two years after the onset of Raynaud's phenomenon. The primary mechanisms in SSc include vasculopathy, inflammation, and fibrosis (Gudjonsson JE et al., 2019).

The pathogenesis of scleroderma involves complex interactions between endothelial cells, epithelial cells, fibroblasts, and lymphocytic cells, which are influenced by genetic, environmental, and chemical factors. These interactions modulate the expression of cytokine and growth factor genes that play crucial roles in immune activation and fibrotic processes. Systemic sclerosis results from interactions between innate and adaptive immune cells, blood vessels, and connective tissues. The interaction between cells and the extracellular matrix (ECM) plays a critical regulatory role in cellular function. In the vascular system, there is autonomous fibroblast activation and differentiation into myofibroblasts, which contract within soft tissues, accompanied by excessive accumulation of ECM proteins. These cells may originate from resident connective tissue fibroblasts, transdifferentiation of other cells, activation of pericytes, or recruitment of circulating progenitor cells (fibrocytes). Various cytokines and growth factors act as mediators in this process, creating a profibrotic microenvironment. The ECM also serves as a reservoir for inflammatory mediators (Gudjonsson JE et al., 2019).

The clinical diagnosis of SSc is established using the 2013 ACR-EULAR criteria, which include additional features such as abnormal nailfold capillaries, fingertip lesions, and the presence of specific autoantibodies. A total score ≥9 indicates classification as dcSSc. According to ACR criteria, diagnosis requires either one major criterion or at least two minor criteria. The major criterion is the presence of proximal joints scleroderma (metacarpophalangeal or metatarsophalangeal joints), while minor criteria include sclerodactyly, digital ulcers, and bilateral pulmonary fibrosis. The table 1 shows the ACR-

EULCAR scoring system. (Aringer, 2025; Hoogen F et al., 2013). In the presented case, the total ACR-EULAR score was 15.

Table 1.
The American College of Rheumatology/European League Against Rheumatism criteria for the classification of systemic sclerosis

Item	Sub-item	Weight/score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints (*sufficient criterion*)	-	9
Skin thickening of the fingers (*only count the higher score*)	• Puffy fingers • Sclerodactyly of the fingers (distal to the metacarpophalangeal joints but proximal to the proximal interphalangeal joints)	2 4
Fingertip lesions (*only count the higher score*)	• Digital tip ulcers • Finger tip pitting scars	2 3
Telangiectasia	-	2
Abnormal nailfold capillaries	-	2
Pulmonary arterial hypertension and/or interstitial lung disease (*maximum score is 2*)	• Pulmonary arterial hypertension • Interstitial lungdisease	2 2
Raynaud's phenomenon SSc-related autoantibodies (*maximum score is 3*)	- • Anticentromere • Anti–topoisomerase I • Anti–RNA polymerase III	3 3

Cutaneous symptoms in scleroderma include widespread skin thickening, cutaneous calcinosis due to calcium deposition in subcutaneous tissues, and areas of hypo- and hyperpigmentation (referred to as "*salt and pepper*" appearance), as well as loss of hair follicles and sweat glands (hypohidrosis/anhidrosis). Characteristic facial features include telangiectasias, a beaked-shaped nose, and a tight mouth (*microstomia*), evidenced by radial perioral wrinkles, expressionless facies, mask-like appearance, and sclerosis of the frenulum. Digital manifestations include digital ulcers, puffy fingers, and sclerodactyly. Raynaud's phenomenon is characterized by recurrent vasospastic attacks affecting the small arterioles/arteries of the fingers and toes, typically triggered by cold exposure or emotional stress. Clinically, it presents with sudden and well-demarcated episodes of pallor or ischemia with pain in one or more digits, followed by reactive hyperemia and cyanosis, forming the classic triphasic Raynaud's phenomenon (Gudjonsson JE et al., 2019).

Skin involvement should be assessed using the mRSS. Seventeen anatomical sites are evaluated: a score of 1 is given for skin thickening, 2 for thickened skin that cannot be pinched, and 3 for thickened, rigid skin. Skin thickness is a key parameter to assess disease activity, severity, and mortality risk (Gudjonsson JE et al., 2019; Avouac J et al., 2009; Johnson SR, 2015). Worsening mRSS is associated with higher mortality. In clinical practice, a meaningful change in mRSS (≥4 points) typically does not occur within less than three months. However, early effects of pharmacological intervention may be detected through mRSS, thus assessment is recommended at least every three months (Dinesh K et al., 2017). In this case, the patient had an initial mRSS of 34, which decreased to 31 after nine months.Histopathological examination of SSc shows fibrosis of the lower two-thirds of the dermis and subcutaneous fibrous trabeculae due to excessive ECM deposition, particularly type I and III collagen. In the early phase, subcutaneous fat is replaced by fibrous connective

tissue, panniculitis, and mucinous edema. Abnormal collagen bundles appear pale, homogenous, and parallel to the skin surface, with accompanying perivascular lymphocytic infiltration. Vascular changes include capillary dilation, endothelial proliferation, and eventual occlusion. The epidermis exhibits atrophy, followed by loss of the pilosebaceous units, eccrine glands, and rete ridges, especially in late-stage diffuse SSc (Gudjonsson JE et al., 2019).

Antinuclear antibodies (ANA) are commonly associated with connective tissue diseases, including scleroderma. ANA is positive in 60–90% of scleroderma patients, with a diagnostic sensitivity of approximately 85% and a specificity of 54%, based on indirect immunofluorescence (IIF). A typical scleroderma screening panel includes anti-Scl-70 antibodies (associated with diffuse SSc) and anti-centromere antibodies (associated with limited SSc). ANA prevalence in the general population may also be linked to immune dysregulation (Tsuji et al., 2025; Bogna G et al., 2018).Management of scleroderma requires a multidisciplinary approach involving specialized nurses, physiotherapists, occupational therapists, subspecialist physicians, and surgeons. The main therapeutic strategies include vascular, immunosuppressive, and antifibrotic approaches. Skin care is essential due to loss of eccrine gland function, leading to dryness. Moderate-to-potent topical corticosteroids are used during the active phase, typically limited to 3–4 months. Systemic corticosteroids (0.5–1.0 mg/kg/day) are first-line treatments and show good response. Adjunctive therapies for localized scleroderma include topical corticosteroids, vitamin D analogs, UV-A phototherapy, and methotrexate. Sildenafil and tadalafil are used for Raynaud's phenomenon and digital ulcers, although prospective clinical trial data remain limited. Surgical options such as digital microarteriolysis may be considered, while amputation should be avoided. Parenteral prostacyclin, phosphodiesterase-5 inhibitors, and potent analgesics are used in severe cases. Antiplatelet agents are indicated in digital ischemia, and patients should be educated to avoid nicotine, stress, cold exposure, and finger trauma (Gudjonsson JE et al., 2019; Odonwodo A et al., 2023).

Prognosis in scleroderma varies significantly depending on cutaneous manifestations and internal organ involvement. The overall survival rate in SSc is approximately 75–80% at 5 years, 55% at 10 years, 35–40% at 15 years, and 25–30% at 20 years. In localized scleroderma, skin thickening typically stabilizes within two years and does not progress, although some lesions may persist or worsen even after inflammation has resolved (Hughes M et al., 2020). The prognosis for this patient is quo ad vitam: bonam, quo ad functionam: dubia ad bonam, and quo ad sanationam: dubia ad malam

CONCLUSION

This case underscores the importance of recognizing Raynaud's phenomenon as an early sign of systemic sclerosis. Delayed diagnosis and follow-up led to severe complications, including pedal ulcers and toe necrosis. Early identification through clinical evaluation, autoantibody testing, and use of ACR/EULAR criteria is crucial for timely treatment. Immunosuppressive therapy and patient education can help prevent irreversible damage. Prompt intervention is key to improving outcomes and reducing long-term morbidity in systemic sclerosis.

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