



Massive multi-fingered gangrene in a case of systemic scleroderma: a rare and disabling complication

Gangrène massive à plusieurs doigts dans un cas de sclérodémie systémique : une complication rare et invalidante

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Résumé

Cet article présente un cas rare de nécrose digitale pluridigitale sévère chez une patiente noire africaine atteinte de sclérose systémique (SSc). Cette femme de 29 ans, suivie irrégulièrement depuis deux ans, a été admise pour dyspnée et nécrose des doigts et des orteils. Les lésions nécrotiques touchaient plusieurs doigts, affectant les articulations phalangiennes proximales. Les marqueurs inflammatoires étaient élevés et l'échocardiographie a révélé un thrombus intra-ventriculaire gauche et une hypertension artérielle pulmonaire (HTAP). Le diagnostic retenu était une poussée de sclérose systémique avec atteinte vasculaire et cardiaque. Le traitement comprenait des corticostéroïdes, de la Nifédipine, du Rivaroxaban et des antibiotiques. L'intervention chirurgicale a permis d'amputer les doigts nécrosés et le traitement de fond a été ajusté. Cette étude met en évidence la rareté et la gravité de la nécrose digitale pluridigitale dans la ScS, soulignant la nécessité d'une prise en charge médicale et chirurgicale méticuleuse. Après quatre mois, la cicatrisation du moignon a été observée.

Mots-clés : gangrène, digitale, raynaud, sclérodémie

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Summary

This article presents a rare case of severe pluridigital digital necrosis in a Black African patient with systemic sclerosis (SSc). The 29-year-old woman, irregularly monitored for two years, was admitted with dyspnea and necrosis of fingers and toes. The necrotic lesions involved multiple digits, affecting proximal phalangeal joints. Inflammatory markers were elevated, and echocardiography revealed intra-left ventricular thrombus and pulmonary arterial hypertension (PAH). The diagnosis indicated a systemic sclerosis flare with vascular and cardiac involvement. Treatment included corticosteroids, nifedipine, rivaroxaban, and antibiotics. Surgical intervention amputated necrotic digits, and the background treatment was adjusted. The study highlights the rarity and severity of pluridigital digital necrosis in SSc, emphasizing the need for meticulous medical and surgical management. After four months, stump healing was observed.

Keywords: gangrene, digital, Raynaud, scleroderma

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Introduction

Systemic sclerosis (SSc) is an autoimmune connective tissue disease characterized by excessive fibrosis of the skin and internal organs, leading to significant functional impairment. Its incidence ranges from 0.3 to 2.8 per 100,000 populations, indicating a relatively rare prevalence of this pathology within the general population (1). One of the severe and infrequently described complications of systemic sclerosis is digital gangrene, especially in its massive pluridigital form. This alarming clinical manifestation can significantly impact patients' quality of life. The Raynaud's phenomenon (RP), observed in 95% of individuals with systemic sclerosis, often precedes this devastating complication (2). However, its close association with systemic sclerosis underscores its crucial role in the development of serious ischemic complications, such as massive pluridigital digital gangrene. In this article, we present a rare clinical case of massive pluridigital digital gangrene occurring in the course of SSc in a Burkinabe patient.

Observation

Patient Information: Mrs. ZF is a 29-year-old woman, a non-smoker and mother of one child. She has been irregularly monitored for two years for systemic sclerosis, receiving methotrexate 15 mg per day and hydroxychloroquine 400 mg per day. She has no other significant medical history.

Timeline: She presented to the rheumatology department with dyspnea stage III associated with necrosis of the fingers of the right hand and toes, reportedly occurring after exposure to cold for about 3 weeks. This was accompanied by a general deterioration of health, apyrexia, and a history of treatment interruption for six months.

Clinical Findings: On physical examination, there was a general state of deterioration, a

respiratory rate of 24 cycles per minute, and other hemodynamic parameters were unremarkable. Skin and nail examination revealed scattered mottled hypochromia on the chest, abdomen, and thighs. The patient also exhibited a pointed nose, limited oral aperture with an inability to whistle and blow. Cutaneous aspects of the limbs showed disseminated sclerosis with a Rodnan score of 26. Ulcerative-necrotic lesions were noted on the fingers II, III, IV, and V of the right hand (fig 1a) and fingers II and III of the left hand (fig 1b), affecting the proximal interphalangeal joints (IPPs).



Figure 1(a): Necrosis of the fingers of the right hand



Figure 1 (b): Necrosis of the fingers

Similar lesions were found on the toes II, III of the right foot (fig 2), limited to the IPPs.





Figure 2: Necrosis of the toes of the right foot

The patient did not present with calcinosis, sclerodactyly, telangiectasia, or arthritis. Cardiovascular examination was normal, and peripheral pulses were normally perceived. Examination of other systems and organs revealed no particularities. Laboratory results showed a biological inflammatory syndrome with a C-reactive protein (CRP) of 76 mg/L and neutrophil-predominant leukocytosis of 12,000/ μ L. Hemostasis, renal, hepatic, and muscular function tests were unremarkable. Anti-Scl70 antibodies were strongly positive. Limb angioscanner was unremarkable. Transthoracic echocardiography (TTE) revealed apex left ventricular akinesia associated with a 19 x11 mm intra-left ventricular thrombus in the apical position, an ejection fraction of 55 %, and a systolic pulmonary artery pressure (PAPs) of 60 mmHg.

Diagnostic Assessment: Given this clinical picture, the diagnosis of a systemic sclerosis flare with vascular and cardiac involvement, presenting as dry gangrene of the fingers and toes, pulmonary arterial hypertension (PAH), and intra-left ventricular thrombus was established.

Therapeutic Intervention: The patient received corticosteroid therapy with prednisolone at a dosage of 1 mg/kg/day, nifedipine, rivaroxaban, and antibiotic therapy. Dermatological lesions were treated with acetylsalicylic acid. Surgical intervention, including metacarpophalangeal disarticulation of the last four fingers on the right hand and metatarsophalangeal disarticulation of the second and third toes on the right foot, was performed on the 6th day of hospitalization under local anesthesia (Figure 3).



Figure 3: Hand at D2 post-disarticulation

Follow-up and Outcomes: Postoperative recovery was uneventful except for mild reactive depression. At day 14, there was clinical and laboratory improvement with the resolution of dyspnea, PAH on TTE, and the biological inflammatory syndrome.

Patient Perspective: The patient's background treatment was adjusted by discontinuing methotrexate and adding D-penicillamine at a dose of 300 mg per day, which she continued along with hydroxychloroquine at home. Rivaroxaban was continued for six months, along with sildenafil. At 4 months, there was evidence of stump healing (Figure 4).



Figure 4 (a): Progression of the right hand at 4 months



Figure 4 (b): Evolution of the stump at 4 months

Discussion

Vascular symptoms are major prognostic factors for SSc, and overall outcomes depend on the extent and severity of vascular lesions (3). SSc and Raynaud's phenomenon (RP) can lead to the development of painful digital ulcers on fingers or toes, causing complications such as severe infection, gangrene, albeit rarely to the extent seen in our case (4). RP emerges as a determining factor in the development of digital necrosis. In the general population, RP is less frequent, with an estimated prevalence between 3 and 5 % (2). Its episodes of peripheral vasoconstriction contribute to impaired blood circulation, leading to severe ischemia of the fingers. Studies highlight the direct correlation between the frequency and severity of RP and the increased incidence of digital necrosis (4).

Nearly half of SSc patients develop a digital ulcer during their disease course, and 30% of these ulcers may progress to some degree of digital gangrene (2). The incidence of digital gangrene increases with a longer duration of digital ulcers, suggesting that inadequate healing heralds further adverse digital sequelae and warrants aggressive treatment and monitoring (1). The underlying mechanisms of digital necrosis in SSc are complex and multifactorial. Vascular involvement plays a central role, characterized by dysregulation of the vascular system and microvessels in particular. RP-induced vasoconstriction contributes to ischemia, compromising blood supply to the digital extremities (5). Additionally, excessive fibrosis in vessels and surrounding tissues hinders normal circulation (5). The chronic



inflammation and dysfunctional immune response characteristic of SSc also contribute to the progression of digital necrosis. Genetic and environmental factors may modulate these mechanisms, creating a complex interplay (6). Angiographic and pathological studies on RP associated with SSc also indicate that vascular disease is characterized by progressive obliteration of affected arteries, with defective angiogenesis and vasculogenesis, resulting in inadequate collateral circulation (5). Endothelial dysfunction, vascular smooth muscle cell activation, and intimal hyperplasia are hallmark features of SSc vasculopathy (5). Understanding these mechanisms is crucial for developing targeted therapeutic approaches aimed at mitigating vascular pathology and preventing digital necrosis in systemic sclerosis patients.

The time between the onset of RP and gangrene in our patient is 3 weeks, which is the shortest timeframe in the review by Takahashi et al., where the longest duration was 54 months (7). Amputation was extremely debilitating for our patient given its right-sided lateralization and her occupation as a cashier.

Pulmonary arterial hypertension (PAH) in the course of SSc significantly worsens the prognosis, leading to progressive deterioration of cardiac function. The exact mechanisms of PAH development in SSc are not fully elucidated, but pulmonary fibrosis and vasoconstriction contribute significantly (8).

Massive pluridigital digital necrosis in the context of systemic sclerosis poses a major clinical challenge requiring complex management strategies. A multidisciplinary approach is essential. Initially, correcting vascular risk factors, such as RP, is crucial to prevent worsening of digital necrosis. Vasodilators, such as calcium channel blockers, N-acetylcysteine (NAC), bosentan, and intravenous prostaglandins, may be used to improve peripheral blood circulation (3). Immunosuppressive treatments may also be considered to modulate the dysregulated immune response in SSc. In advanced cases, surgical management, including debridement of necrotic tissues and vascular reconstruction, may be necessary, even adventitiectomy (3).

Hyperbaric therapy may promote lesion healing and prevent further complications. The only alternative available to us was disarticulation to prevent the spread of gangrene.

It is crucial for clinicians to be highly aware of the distinction between digital necrosis stemming from systemic lupus erythematosus (SLE) as opposed to that seen with SSc because management differs. Digital ischemia related to SLE often necessitates the prompt initiation of corticosteroids, whereas SSc-related digital ischemia requires aggressive vasodilator therapy (9).

Research on predictive biomarkers for digital necrosis in the context of SSc is a promising and essential area of investigation to improve clinical management. Studies have identified biomarkers such as cytokines, growth factors, and autoantibodies associated with SSc (10). The discovery of predictive biomarkers could allow for early intervention, thus enhancing the prevention and management of digital necrosis.

The importance of patient awareness and education in preventing digital necrosis in the course of systemic sclerosis is crucial for improving clinical outcomes and quality of life. Informed patients are better equipped to recognize early signs of vascular complications, such as RP, which may predispose to digital necrosis. Patients should be encouraged to maintain rigorous digital hygiene, avoid excessive cold exposure, and adopt coping strategies to minimize physical stress on the fingers. Additionally, patient education on the importance of regular follow-up with their medical team and adherence to prescribed treatments contributes to a holistic approach to preventing digital necrosis.

Conclusion

In conclusion, massive pluridigital digital necrosis remains a rare but severe complication in the course of systemic sclerosis, requiring careful clinical attention. This case underscores the importance of close monitoring of SSc patients, highlighting the potential risk of serious complications such as digital gangrene. While current management focuses on the prevention and treatment of RP, thorough investigations are



needed to identify more effective therapeutic strategies.

Consent: Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy. The patient gives written consent.

Conflict of interest

We have no conflict of interest

We have obtained the informed consent of the patient and his family

We have obtained consent to publish this article

Authors' contributions

All the authors contributed to the drafting of this article.

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