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CaseReport

Case report of digital ulcers as a dermatological manifestation in systemic sclerosis

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ABSTRACT:
Systemic sclerosis is one of the most frequent causes of secondary Raynaud's phenomenon; its appearance may occur long before other signs and symptoms. Timely, accurate identification of secondary Raynaud's phenomenon may accelerate a final diagnosis and positively alter prognosis. Correct approach to the patient with DUs begins with a careful examination and evaluation of risk factors and comorbidities in order to cure and prevent complications and further lesions. In the present case report, we reported case of a 37 year old female patient reported with the chief complaint of Bluish discolouration of fingers from past 10 years along with Pitting over fingertips.
Key words: Digital Ulcers, Systemic sclerosis

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INTRODUCTION

Scleroderma is a rare connective tissue disorder with unknown and complex pathogenesis. Scleroderma can be divided into two forms, localized scleroderma or Systemic sclerosis, which can further be classified as either limited systemic sclerosis or diffuse systemic sclerosis based on clinical and serological criteria.[1, 2] Systemic sclerosis (SSc) is a chronic autoimmune disease which still poses a great challenge to clinicians. The most prominent feature of SSc is the process of progressive fibrosis resulting from the excessive deposition of extracellular matrix components in different tissues and organs.[3] Vascular damage, inflammation and the presence of specific autoantibodies are also characteristic for SSc. Systemic sclerosis affects skin and internal organs, such as lungs, heart, kidneys, musculoskeletal system and the gastrointestinal tract. Skin sclerosis is a main symptom of SSc, evaluated using the modified Rodnan skin score (mRss) and an easily detectable marker of disease activity.[4]

Digital ulcers (DUs) are a common visible manifestation of the progressive vascular disease that characterizes the SSc disease process. DUs not only impact significantly on patients' quality of life and hand function, but are also a biomarker of internal organ involvement and of disease severity. The aetiology of (digital) vascular disease in SSc is multifactorial, and many of these factors are potentially amenable to therapeutic intervention.[5, 6] Here we present a case of digital ulceration in a case of systemic sclerosis, managed at our tertiary hospital.

CASE REPORT

A 37 year old female patient reported with the chief complaint of Bluish discolouration of fingers from past 10 years along with Pitting over fingertips. Patient gave history of trauma to her right finger tip. The discoloration gradually increased in size. It was associated non healing for a period of 2 months with multiple dressings and with bluish discolouration of finger of both hands. The lesions aggravated on

exposure to cold environment and were relieved on immersing hands in warm water.

Patient also had a history of difficulty in swallowing, solid more than liquids. After one month, patient also developed pitting over her right third fingertip. She visited KEM hospital for the same complaints. Patients were prescribed some medications, she continued the same medications for 3 years till 2015. In 2015, patient developed digital ulcer over other fingertips. She consulted KEM again and diagnosed with MCTD/ILD/Digital gangrene and was started on InjEndoxan 500 mg; 7 cycles - once a month in 2015, 6 cycles – twice a month in 2016. Patient slowly started noticing tightening of skin around the face While assessing the past medical history of the patient, she gave h/o taking AKT in 2012 for a year after developing cough and white expectorant. Patient gave history of menstrual irregularities since 2012, P1 IUFD in 2009, P2 8th months, preterm delivery, neonatal death on day 2 and P3 abortion in 2015. She received bosentan 62.5 mg twice daily for 4 weeks and then 125 mg twice daily and iloprost 0.5–2 ng/kg/min for the duration of 6 h every 4 weeks for 6 months. Blood chemistry, including liver function and total blood count, was monitored every 2 weeks during the first month and every 4 weeks thereafter. At each visit, changes in digital ulcer characteristics were assessed, comparing the number, size, extent, and granulation, recording onset of new digital ulcers and scoring the intensity of pain reported. The treatment with bosentan in combination with iloprost is effective in determining the healing of digital ulcers in SSc.

DISCUSSION

Systemic sclerosis clinically characterized by thickening of the skin caused by the accumulation of collagen and by structural and functional abnormalities of visceral organs including the gastrointestinal tract, lungs, heart, and kidneys. Digital ulcers (DUs) is an external manifestation of vasculopathy in scleroderma and patients who experience a DU, more than half have persistent or recurrent DUs for at least 6 months.[7]

In the present case report, we reported case of a 37 year old female patient reported with the chief complaint of Bluish discolouration of fingers from past 10 years along with Pitting over fingertips. Patient gave history of trauma to her right finger tip. The discoloration gradually increased in size. She received bosentan 62.5 mg twice daily for 4 weeks and then 125 mg twice daily and iloprost 0.5–2 ng/kg/min for the duration of 6 h every 4 weeks for 6 months. Blood chemistry, including liver function and total blood count, was monitored every 2 weeks during the first month and every 4 weeks thereafter. Kato et al reported in a small study of 10 patients that a reduced ankle brachial index (reflecting macrovascular disease) is associated with DUs of the feet and with (lower) skin perfusion pressure. DUs may also develop in relation to s.c. calcinosis, not uncommonly associated with a local inflammatory response and the discharge of calcinotic material.[8]

In the present case report, at each visit, changes in digital ulcer characteristics were assessed, comparing the number, size, extent, and granulation, recording onset of new digital ulcers and scoring the intensity of pain reported. The treatment with bosentan in combination with iloprost is effective in determining the healing of digital ulcers in SSc. Digital ulcers occur in 30–58% of patients, and are more frequent in patients with diffuse SSc. Digital ulcers are painful and determine a functional impairment with a significant impact on the patient's quality of life. Furthermore, chronic ulcers can become infected, leading to gangrene and need of amputation. The healing is slow due to the atrophic, fibrotic, and avascular nature of the local tissue.[8- 10]

Currently, there is no official algorithm for diagnosis and therapy of digital ischemia leading to digital ulcers in SSc. A conventional therapeutic approach to digital lesions should include vasoactive medications, antiplatelet agents, antibiotics as needed, and analgesia. The response to vasodilators in patients with SSc is variable and often disappointing. There is a visible need for strategies to facilitate healing of the digital ulcers and to prevent occurrence of new lesions.

The development of (digital) vascular disease in SSc is believed to be multifactorial, and many of these factors are amenable to pharmacological intervention. These include: Vasoactive therapies, Calcium channel blockers, Angiotensin-converting enzyme inhibitors, Prostanoids, Endothelin receptor-1 antagonists, Phosphodiesterase type-5 inhibitors and Combination therapy.[9, 10]

De Cata A et al analyzed data regarding 34 patients with SSc and at least one active DU persisting despite 6 months of iloprost therapy, and treated for other 6 months with a combination therapy, i.e.iloprost plus bosentan. Overall, patients initially presented 69 DUs (58 on the fingers and 11 on the legs). At the end of the study 34 (49.3%) DUs were completely healed (responding, R), 18 (26.1%) started the healing process (partially responding, PR), and 17 (24.6%) did not respond (NR) to therapy. In the group with mild fibrosis, 83.4% of DUs resulted with showing complete healing while, in the group with severe fibrosis, only 18% of DUs were healed ($P = 0.024$). The treatment with iloprost plus bosentan is effective in determining healing of DUs in SSc patients with mild digital skin fibrosis.[10]

CONCLUSION

Systemic sclerosis is one of the most frequent causes of secondary Raynaud's phenomenon; its appearance may occur long before other signs and symptoms. Timely, accurate identification of secondary Raynaud's phenomenon may accelerate a final diagnosis and positively alter prognosis. Correct

approach to the patient with DUs begins with a careful examination and evaluation of risk factors and comorbidities in order to cure and prevent complications and further lesions. Then also a correct local treatment of the DU provides a better milieu to foster healing and prevent complications such as infection or gangrene. Future research is warranted to optimize the management of DUs, including combination therapies and the development of locally acting treatments.

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