



A 44-year-old male with *Raynaud's Phenomenon* and Systemic Sclerosis : CASE REPORT

Baiq Rulia Ashlihan¹, Maz Isa AA²

1. Young Doctor At West Nusa Tenggara General Hospital, Medical Faculty of Mataram University, Indonesia
2. Cardiothoracic Surgeon at Department of Thoracic and Cardiovascular Surgery, West Nusa Tenggara General Hospital

ABSTRACT

Raynaud's Phenomenon is a condition caused by vascularization disorders with episodic responses to cold temperatures or emotional stress that can cause changes in skin color to pale, cyanosis, and erythema and has symptoms such as numbness, pain, and tingling in the extremities. This case report discusses a 44-year-old male patient with a primary complaint of blackened left fingers and left feet. This complaint has been present for the past 6 months, accompanied by numbness. When the temperature is cold, the hands turn blue and return to red when the temperature normalizes. The patient has a smoking habit of 1 pack per day. There is a history of seizures felt on the left side of the body, and the patient is aware of the seizures.

Keywords: *Raynaud's Phenomenon*, vasospasm, skin color



INTRODUCTION

This condition can be idiopathic, known as primary Raynaud's phenomenon, or associated with underlying connective tissue diseases such as systemic sclerosis, referred to as secondary Raynaud's phenomenon. Some studies emphasize that although the primary type is more common (3-5% of the general population), secondary Raynaud's phenomenon is primarily associated with systemic sclerosis, which is rare (0.2% of the general population), but can exacerbate conditions such as digital ulcers.¹ Clinical manifestations of this condition are characterized by episodic spasm of the arterioles, typically affecting several fingers, which may present as pallor or cyanosis, and are triggered by exposure to cold, emotional stress, or medications. During the critical phase, patients may complain of pain, hypothermia, numbness, and paresthesia in the affected fingers. If these episodes occur frequently and intensely, they can lead to arterial blockage in the fingers and palms, resulting in ischemic lesions on the fingers, which are extremely painful and difficult to heal.²

Treatment for Raynaud's phenomenon is primarily non-surgical. Sympathectomy has

been used in certain patients who, despite adequate clinical treatment, continue to experience severe symptoms or difficult-to-heal trophic lesions.

CASE

A patient referred to Tripat General Hospital presented with blackening of the left fingers and toes for the past 6 months. The complaint was accompanied by numbness in the hands and feet. When exposed to cold, the hands turned blue and red when the hands and feet dried. The patient reported that these symptoms appeared suddenly. The complaints were not accompanied by pain, and the patient was able to perform activities within normal limits. The symptoms worsened when the patient was exposed to cold temperatures, causing the hands and feet to turn blue. When at normal temperatures, the symptoms improved. The patient also reported intermittent tingling sensations in the hands and feet and frequently felt easily fatigued when walking more than 5 meters. The patient also complains of a feeling of tightness throughout the body and difficulty walking for the past 5 years, as well as difficulty moving the mouth. The patient also reports weakness, nausea, and fever for the



past 5 days. Urination and bowel movements are normal. Physical examination of the upper extremities revealed warm extremities, CRT <2 seconds, and necrosis on the fourth digit of the left hand. The lower extremities also showed warm extremities, CRT <2 seconds, and necrosis on the second digit of the left foot (Figure 1)

DISCUSSION

The patient in this case is suspected of having Raynaud's phenomenon based on the clinical manifestations observed. Raynaud's phenomenon is a vascular disease characterized by intermittent reduced blood supply accompanied by vasospasm in the digital arteries of the extremities. The patient complained of the left ring finger and left index toe turning blue when exposed to cold air and reddening when the air temperature warmed. The symptoms improve when the ambient temperature is normal. The symptoms observed in Raynaud's phenomenon reflect complex vasospastic disorders that cause episodic color changes (pallor, cyanosis, and erythema) in the extremities when exposed to cold or emotional stimuli. This is due to increased vasoconstriction combined with impaired vasodilation.^{1,11}

In 10–20% of cases, Raynaud's phenomenon is an early manifestation of

an undiagnosed connective tissue disorder, such as scleroderma, dermatomyositis, systemic lupus erythematosus (SLE), Sjögren's syndrome, and rheumatoid arthritis (RA).¹⁰ If the history and physical examination point to Raynaud's phenomenon, further testing should include a complete laboratory workup, such as a complete blood count, erythrocyte sedimentation rate (ESR), inflammatory markers (C-reactive protein), and autoantibodies. Autoantibody testing is typically important for patients at risk of developing connective tissue diseases as it helps identify the target antigens of autoantibodies. Other routine tests should also include a biochemical profile, thyroid function tests, and chest radiography.^{12,13} Based on the patient's laboratory results, microcytic hypochromic anemia, prolonged ESR, positive ANA, and ANA profile analysis showing extremely high Scl-70 antigen were identified, pointing to underlying systemic sclerosis as the cause of the patient's symptoms.

In secondary Raynaud's phenomenon, abnormalities occur in the density and structure of arteries, arterioles, and capillaries associated with the loss of nutrient blood vessels in the fingers. Larger digital arteries also involve hyperplasia



intima and fibrosis caused by increased collagen deposition. A histological study of digital arteries showed a reduction in lumen diameter $>75\%$ in 79% of the vessels examined. These vascular changes are not limited to the skin or digital vessels but can occur throughout the systemic microcirculation, particularly in organs such as the heart, lungs, kidneys, and gastrointestinal tract. In addition to excessive vasospasm, the formation of microthrombi within blood vessels can cause tissue damage. This process is likely associated with platelet activation and impaired fibrinolysis. Platelet activation markers have been shown to be elevated in scleroderma, including thromboxane A2, β -thromboglobulin, serotonin, platelet-derived microparticles, and platelet-derived growth factor.¹⁰

Management of this condition begins with medical treatment; however, there is no

A definitive treatment method for curing the symptoms. Conservative management such as reducing emotional stress, protection from the cold, and quitting smoking, in addition to medical therapy, is known to have a small effect in minimizing the signs and symptoms. Surgical treatment options include balloon angioplasty, vein or artery grafting, and sympathectomy, which show

varying results. Sympathectomy is a surgical option for patients with persistent tissue ischemia despite optimal medical therapy. This procedure aims to relieve vasospasm by stopping sympathetic nerve signals to the digital arteries.

Video-Assisted Thoracoscopic Surgery (VATS) has evolved over the years to include medical management that cannot be addressed with standard medical therapy. The introduction of VATS sympathectomy in managing Raynaud's phenomenon has been highlighted in several cases and is a surgical technique that has been used for years to alleviate symptoms of Raynaud's phenomenon. However, in recent years, this procedure has been used to eliminate symptoms in cases of Raynaud's phenomenon, including frequent and severe episodic attacks, tissue damage such as ulcers and digital necrosis that do not heal, and degenerative changes in the extremities despite adequate medical treatment. VATS sympathectomy reduces peripheral vascular resistance, thereby increasing blood flow to the peripheral vascular system.

, thereby increasing blood flow to the peripheral vascular system.¹¹ During the procedure, the sympathetic nerve chain is visualized behind the parietal pleura, cauterized, and cut using an electrocautery, harmonic surgical knife, endoscopic



scissors, or other modern techniques. VATS sympathectomy has a good prognosis and provides a positive response with minimal scarring and no complications. A retrospective study of 34 patients treated with thoracoscopic sympathectomy showed that immediate effects were observed in 83% of patients, but symptoms recurred in 60% during a 40-month follow-up period.¹² Long-term effects of thoracic sympathectomy include better outcomes in patients operated on for secondary Raynaud's phenomenon, with 31% having excellent results and 58% having beneficial effects. These differences from the primary type can be explained by more objective outcomes for patients with the secondary type (tissue loss and ulcers) compared to the more subjective outcomes of pain and the three-color phenomenon (pallor, cyanosis, and erythema) in most patients with the primary type. This is because complete healing or significant improvement was achieved in 95% of patients undergoing treatment for digital ulcers. Another explanation is that sympathectomy for secondary Raynaud's phenomenon is often performed after tissue loss due to embolization or digital artery thrombosis, where temporary vasodilation (6 months) of skin arterioles appears to improve tissue oxygenation sufficiently for wound healing.

After healing, less oxygenation is required, and increased sympathetic tone has no clinical consequences on the healed wound.

CONCLUSION

A 44-year-old male patient presented with the main complaint of blackening of the left hand and left foot. The patient has experienced these symptoms for the past 6 months, accompanied by numbness, with the hands turning blue in cold temperatures and reddening when the temperature returns to normal. The patient has a smoking habit of 1 pack per day. The patient has a history of seizures affecting the left side of the body, with the patient remaining conscious during the seizures. The ANA test results were positive for systemic sclerosis, SLE, and ANA IF (+). The patient underwent amputation of the fourth digit of the left hand, amputation of the second digit of the left foot, and VATS sympathectomy.

REFERENCES

Yu, F., Liu, Y., Zhang, C. et al. (2023). Efficacy analysis of minimally invasive surgery for Raynaud's syndrome. *BMC Surg* 23, 313. [doi: 10.1186/s12893-023-02225-x](https://doi.org/10.1186/s12893-023-02225-x).

De Campos JRM, Kauffman P, Faustino CB, Wolosker N. (2018). Upper extremity



sympathectomy. *J Vis Surg* 4:180. doi: 10.21037/jovs.2018.08.04.

Pain, C. E., Constantin, T., Toplak, N., Moll, M., Iking-Konert, C., Piotto, D.

P., Aktay Ayaz, N., Nemcova, D., Hoeger, P. H., Cutolo, M., Smith, V., Foeldvari, I., & Paediatric Rheumatology European Society (PRES) Juvenile Scleroderma Working Group. (2016). Raynaud's syndrome in children: systematic review and development of recommendations for assessment and monitoring. *Clinical and Experimental Rheumatology*, 34 Suppl 100(5), 200–206.

Valdovinos, S. T., & Landry, G. J. (2014). Raynaud Syndrome. *Techniques in Vascular and Interventional Radiology*, 17(4), 241–246. doi:10.1053/j.tvir.2014.11.004

Pauling, J. D., Hughes, M., & Pope, J. E. (2019). Raynaud's phenomenon—an update on diagnosis, classification and management. *Clinical Rheumatology*. doi:10.1007/s10067-019-04745-5

Maundrell, A., & Proudman, S. M. (2014). Epidemiology of Raynaud's phenomenon. In *Raynaud's phenomenon: a guide to pathogenesis and treatment* (pp. 21-35). New York, NY: Springer New York.

Haque, A., & Hughes, M. (2020). Raynaud's phenomenon. *Clinical medicine* (London, England), 20(6), 580–587. <https://doi.org/10.7861/clinmed.2020-0754>.

Musa R, Qurie A. Raynaud Disease. [Updated 2023 Aug 8]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK499833/>

Herrick, A. L., & Wigley, F. M. (2020). Raynaud's phenomenon. *Best Practice & Research Clinical Rheumatology*, 101474. doi:10.1016/j.berh.2019.101474

Achurra-Godínez CP et al. (2023). Role of the vascular surgeon in the treatment of secondary Raynaud's phenomenon, literature review and endovascular management proposal. *Int Surg J*. 10(12):2025-203. doi: 10.18203/2349-2902.isj20233688.

Aljehani Y, Alhouri A, Turkistani A, ShahBahai R, AlQatari AA. (2020). Bilateral uniportal video-assisted thoracoscopic sympathectomy for managing secondary Raynaud's in CREST syndrome: A case report. *Int J Surg Case Rep*. 75:203-206. doi: 10.1016/j.ijscr.2020.08.041.

Marczyk, Aleksandra, et al. (2024). Review of Raynaud's phenomenon: pathomechanisms, diagnosis and treatment. *Journal of Education, Health and Sport*. 72:51508.

Lis-Święty A. (2019). Recent advances in the workup and management of Raynaud phenomenon. *Pol Arch Intern Med*. 129: 798-808. doi:10.20452/pamw.15008.

14. Coveliers HM, Hoexum F, Nederhoed JH, Wisselink W, Rauwerda JA. (2011). Thoracic sympathectomy for digital ischemia: a summary of evidence. *J Vasc Surg*. 54(1):273-7. doi: 10.1016/j.jvs.2011.01.069.