Beyond the Irregular Beat: An Unexpected Case of Raynaud's Phenomenon in a Patient with Atrial Fibrillation

Ni Putu Widyanti Suastiari[1], Rahmania[2,3], Riawati Utama[1]*

[1]Faculty of Medicine, Wijaya Kusuma Surabaya University, Surabaya, Indonesia.
[2]Department of Internal Medicine, Faculty of Medicine, Wijaya Kusuma Surabaya University - Dr. Mohamad Saleh General Hospital, Probolinggo, East Java, Indonesia.
[3]Division of Cardiology and Vascular Medicine, Department of Internal Medicine, Faculty of Medicine, Wijaya Kusuma Surabaya University - Dr. Mohamad Saleh General Hospital, Probolinggo, East Java, Indonesia.

Corresponding Author:
Ni Putu Widyanti Suastiari, MD. Faculty of Medicine, Wijaya Kusuma Surabaya University. Jl. Dukuh Kupang XXV No.54, Dukuh Kupang, Surabaya 60225, Indonesia. Email: putuwidyanti03@gmail.com.

ABSTRACT

Raynaud's phenomenon is characterized by episodic vasospasm of the extremities, often triggered by cold or stress. It is a hallmark of systemic sclerosis (SSc) and is frequently associated with pulmonary arterial hypertension (PAH). PAH may cause right atrial dilatation, predisposing to atrial fibrillation (AF). While Raynaud's and PAH are well-documented, their coexistence with AF is rarely reported. A 71-year-old male presented with rapid AF and pulmonary oedema. On admission, vital signs were stable, but he developed symmetrical discoloration of the fingers, palms, and feet after cold exposure in the ICU, accompanied by numbness and pain. Laboratory tests revealed prolonged PT, high ESR (65/121 mm/hr), and positive ANA (44.8 IU/mL). ECG showed rapid AF with left ventricular hypertrophy. Chest X-ray demonstrated pulmonary artery dilatation and cardiomegaly. Echocardiography revealed moderate tricuspid regurgitation and severe PAH. This case illustrates a rare combination of Raynaud's phenomenon, PAH, and AF, likely secondary to autoimmune vascular dysfunction. PAH-induced atrial dilatation may trigger AF, while endothelial dysfunction may worsen Raynaud's symptoms. A positive ANA suggests underlying SSc or SLE. Early recognition and multidisciplinary management targeting pulmonary pressure, arrhythmia control, and vasospasm are crucial to prevent complications. Long-term outcome was not assessed due to loss to follow-up.

Keywords: *Raynaud's phenomenon, atrial fibrillation, pulmonary arterial hypertension, systemic sclerosis.*

INTRODUCTION

Raynaud's phenomenon is characterized by episodic vasospasm of the extremities, typically triggered by cold exposure or emotional stress.[1] It affects approximately 3-5% of the general population, with a higher prevalence in women and those living in colder climates.[2] Clinically, it presents with sequential color changes in the fingers: pallor from arterial spasm, cyanosis from blood deoxygenation, and erythema during vascular reperfusion.[1]

Primary Raynaud's phenomenon occurs in isolation, while secondary Raynaud's phenomenon is often a manifestation of underlying autoimmune diseases, particularly systemic sclerosis (SSc).[3] SSc affects approximately 240 cases per million people worldwide and is frequently complicated by pulmonary arterial hypertension (PAH), which occurs in 8-12% of patients with SSc.[3] PAH represents

a progressive vasculopathy characterized by elevated pulmonary vascular resistance and increased pulmonary arterial pressure, leading to right heart failure if left untreated.[4]

The pathophysiological connection between PAH and cardiac arrhythmias, particularly atrial fibrillation (AF), has been increasingly recognized.[4] PAH can lead to right atrial enlargement and increased wall stress, creating a substrate for atrial remodeling and subsequent development of AF.[4] Atrial arrhythmias develop in 10-25% of patients with severe PAH over five years and are linked to increased morbidity and mortality.[5]

Although Raynaud's phenomenon and PAH are well-documented complications of connective tissue diseases, their coexistence with AF in a single patient is rarely reported in the literature.[3,4] The simultaneous occurrence of this triad may represent a complex manifestation of underlying systemic autoimmune vasculopathy. This case underscores the importance of recognizing this rare but clinically significant constellation of findings and understanding its diagnostic and therapeutic implications.

CASE ILLUSTRATION

A 71-year-old man with a history of hypertension presented to the intensive cardiac care unit (ICCU) with acute shortness of breath. On arrival, vital signs showed blood pressure 110/78 mmHg, heart rate 100 beats per minute, temperature 36.7°C, and oxygen saturation 98% on room air. He exhibited rapid atrial fibrillation and signs of pulmonary oedema. Shortly after exposure to the cold ICCU environment, he developed bluish discoloration of the fingertips, which symmetrically progressed to the palms and feet. He also reported numbness, tingling, and pain. Laboratory investigations revealed prolonged prothrombin time (14.5 seconds), markedly elevated erythrocyte sedimentation rate (65/121 mm/hr), and positive antinuclear antibody (44.8 IU/mL). Electrocardiography (ECG) showed an irregular rhythm consistent with rapid AF and evidence of left ventricular hypertrophy. Chest radiography demonstrated cardiomegaly, pulmonary oedema, and a prominent pulmonary artery with an inverted coma sign. Echocardiography revealed left atrial dilatation (4.3-4.4 cm), a small left ventricle (LVIDd 3.8 cm), preserved LVEF 56%, but reduced cardiac output (3.4 L/min). Right heart assessment showed moderate tricuspid regurgitation with peak velocity >3.4 m/s, moderate pulmonary regurgitation, and severe pulmonary hypertension (RVSP >56 mmHg).[4,5] The diagnosis included rapid AF and suspected connective tissue disease, manifesting as Raynaud's phenomenon and PAH. The patient

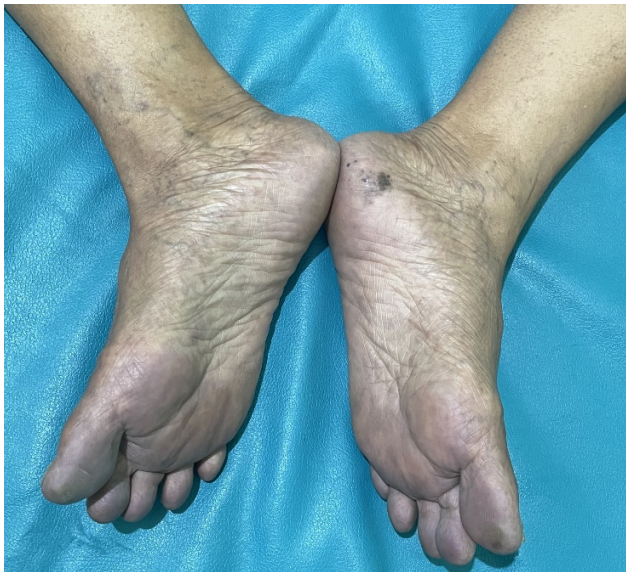

Figure 1. Fingers showing the ischaemic (pallor) phase of Raynaud's phenomenon, triggered by cold exposure.

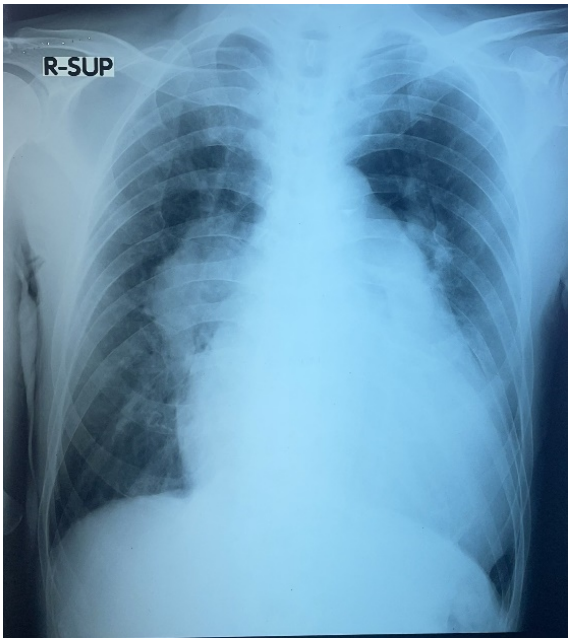

Figure 2. Chest radiograph demonstrating cardiomegaly and enlarged pulmonary arteries with an inverted coma sign, suggestive of pulmonary arterial hypertension.

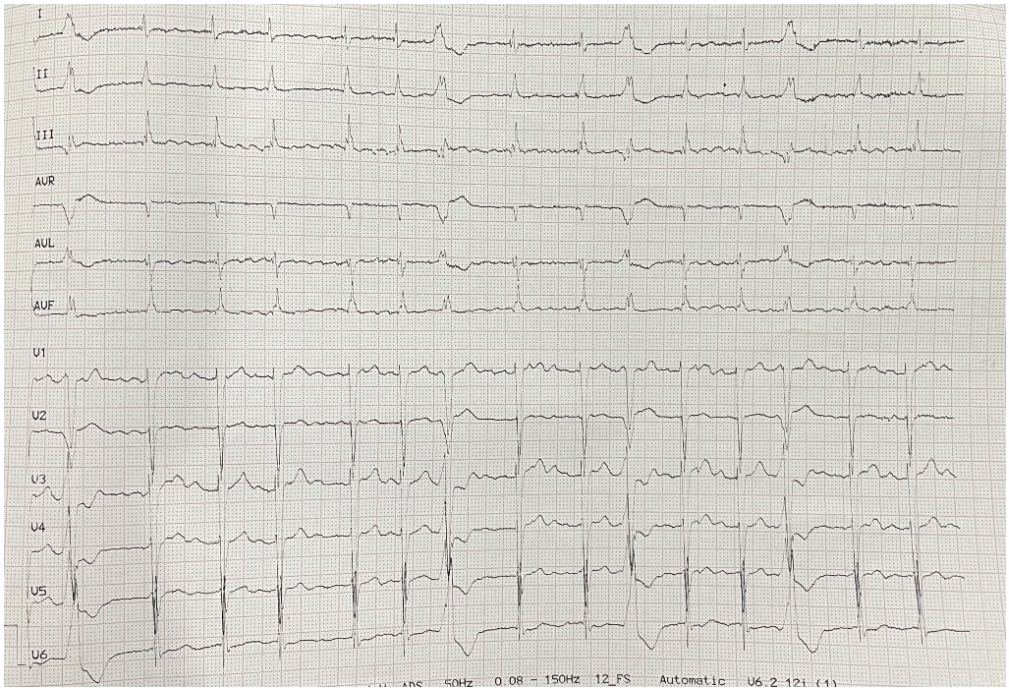

Figure 3. **Electrocardiogram showing atrial fibrillation with a rapid ventricular response and an irregular** rhythm.

received supportive therapy and was referred for rheumatological evaluation. Despite the inability to obtain long-term follow-up, the patient exhibited satisfactory short-term clinical improvement with stabilized cardiac rhythm and resolution of vasospastic manifestations at discharge. A comprehensive follow-up plan encompassing rheumatology evaluation and cardiac monitoring was recommended.

DISCUSSION

This case illustrates the complex interaction among autoimmune pathology, pulmonary vascular disease, and arrhythmias. PAH elevates right-sided cardiac pressures, promoting atrial remodelling and dilatation, which increase the risk of AF.[4] Vascular dysfunction, a hallmark of autoimmune connective tissue disorders including SSc and systemic lupus erythematosus, plays a central role in the pathogenesis of Raynaud's phenomenon.[3,6]

Although the link between connective tissue disease and PAH is well established, concurrent AF adds to the diagnostic and therapeutic complexity.[3,4] The patient's ANA positivity, digital vasospasm, and pulmonary hypertension strongly suggest an autoimmune etiology, most likely systemic sclerosis.[3]

Optimal management requires a multidisciplinary approach involving rheumatology, cardiology, and pulmonology.[7] Early diagnosis and targeted interventions, aimed at modulating immune responses, managing pulmonary pressures, and stabilizing cardiac rhythm, are essential for improving prognosis and quality of life.[4]

CONCLUSION

This case demonstrates the importance of recognizing rare manifestations and implementing comprehensive management strategies, even when long-term follow-up is not feasible. This case highlights the interplay between Raynaud's phenomenon, PAH, and AF, likely driven by vascular dysfunction secondary to an underlying autoimmune disorder.[3] PAH contributes to right atrial dilatation and myocardial stress, precipitating AF,[4] while endothelial dysfunction exacerbates Raynaud's symptoms.[3] A positive ANA suggests SSc or systemic lupus erythematosus as possible underlying conditions.[3] Given the strong association between connective tissue disease, PAH, and arrhythmias,[3] early identification and a multidisciplinary approach targeting pulmonary pressure, cardiac rhythm, and vasospasm control are essential to prevent complications and improve outcomes.[4,7] Despite the inability to obtain long-term follow-up, the patient exhibited satisfactory short-term clinical improvement with stabilized cardiac rhythm and resolution of vasospastic manifestations at discharge.

CONFLICT OF INTERESTS

The authors declare no conflicts of interest.

ACKNOWLEDGMENTS

The authors thank the healthcare staff of Dr. Mohamad Saleh General Hospital for their support and assistance. Gratitude is also extended to the patient and his family.

FUNDING

This research received no external funding.

REFERENCES

1. Curtiss P, Svigos K, Schwager Z, Lo Sicco K, Franks AG. Part I: Epidemiology, pathophysiology, and clinical considerations of primary and secondary Raynaud's phenomenon. J Am Acad Dermatol. 2024 Feb;90(2):223–34.
2. Wigley FM, Flavahan NA. Raynaud's phenomenon. New England Journal of Medicine. 2016 Aug 11;375(6):556–65.
3. Maciejewska M, Sikora M, Maciejewski C, Alda-Malicka R, Czuwara J, Rudnicka L. Raynaud's phenomenon with focus on systemic sclerosis. J Clin Med. 2022 Apr 28;11(9):2490.
4. Humbert M, Kovacs G, Hoeper MM, et al. 2022 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension. Eur Heart J. 2022 Oct 11;43(38):3618–731.
5. Anand S, Cronin EM. Arrhythmias in patients with pulmonary hypertension and right ventricular failure: Importance of rhythm control strategies. J Clin Med. 2024 Mar 24;13(7):1866.
6. Patnaik E, Lyons M, Tran K, Pattanaik D. Endothelial dysfunction in systemic sclerosis. Int J Mol Sci. 2023 Sep 21;24(18):14385.
7. Belch J, Carlizza A, Carpentier PH, et al. ESVM guidelines – the diagnosis and management of Raynaud's phenomenon. Vasa. 2017 Oct 1;46(6):413–23.