An Interesting Case of Raynaud's Phenomenon- A Question of Primary or Secondary Raynaud's Phenomenon

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Received: December 2023. Accepted: January 2024; Published: February 1, 2024.
Citation: Mohammad Irfan Zahid, Tehsin Kauser Ali. An Interesting Case of Raynaud's Phenomenon- A Question of Primary or Secondary Raynaud's Phenomenon. World Family Medicine. February 2024; 22(2): 80-82
DOI: 10.5742/MEWFM.2024.95257022

Abstract

Raynaud's Phenomenon (RP) is a common condition seen in family medicine clinics. Patients with RP are described as having either a primary or secondary process. Primary RP is a benign condition and is usually not associated with underlying disease, whereas secondary RP is associated with underlying disease and can be associated with significant complications such as digital ischemia, digital ulceration, necrosis, scarring and secondary infection. Thus, it is important to differentiate between the two types of RP.

Although primary RP is not caused by an underlying condition it can be aggravated by certain conditions such as diabetes and hypertension. This case report highlights a case where a patient presents with RP and was subsequently diagnosed with Latent Onset Type 1 Diabetes Mellitus after investigations. He was started on insulin treatment and the subsequent improvement in Hba1c resulted in significant improvement in his RP symptoms.

Keywords: Raynaud's phenomenon, case report

Introduction

Raynaud's phenomenon (RP) is episodic vasospasm of the arteries or arterioles in the extremities (usually the digits) which leads to colour change including - pallor (due to decreased blood flow), followed by cyanosis (due to deoxygenation), and/or rubor (due to reactive hyperaemia) which may be associated with significant discomfort (1). The most common triggers for RP are exposure to cold temperatures and to emotional stress (1).

Patients with RP are described as having either a primary or secondary process (2). Primary RP (80-90% of cases) refers to reversible vasospasm of the peripheral arteries in the absence of underlying disease (3). Secondary RP (10-20% of cases) is associated with an underlying cause (2). The number of diseases and exposures associated with secondary RP is extensive (1,2).

Primary RP usually does not progress to tissue damage but in people who are severely affected it can cause significant discomfort and reduce quality of life. Secondary RP is associated with complications such as digital ischaemia, digital ulceration, necrosis, scarring and secondary infection (4). Thus, it is important when patients present with RP, we perform a thorough assessment and investigate appropriately to ensure no underlying cause which can be treated to avoid any long-standing complications.

Case

A 38 year old Caucasian male came for a consultation to a General Practice (GP) surgery in the UK. He had no significant past medical history and had not seen a doctor for a few years. He worked as a manual laborer and was a smoker of 15 cigarettes a day since his late teens. He presented with classical RP symptoms over the last 6 months. He reported distal ends of all his fingers on both hands were going pale and a white-like color which was associated with pain at work when it was cold. He reported when he warmed up his hands his symptoms would resolve. He states this never happened prior to 6 months ago and had been working in the same environment for a while. Other than an unexplained 10kg weight loss over the last 12 months the patient was systemically well and had no other symptoms when questioned. There was no significant family history of RP or any other significant conditions. Physical examination was normal, and fingers and toes looked normal. He had a BMI of 28.

Presentation of new RP in a male patient, the fact that he was in his 30's and he also reported weight loss suggested the need for further investigation. Routine blood work was requested including full blood count, Hba1c, ESR, CRP, thyroid function, liver function, renal profile, calcium, lipid profile and auto-Immune screen (ANA, Anti-dsNA, Anti-Rheumatoid factor). His Hba1c came back significantly high, 12.9% (118 mmol/mol). His other blood results did not show anything significant. His previous Hba1c from 5 years ago was in the normal range and he was not a known diabetic. Thus, the patient was brought in for another consultation, urine was checked and ketones were negative, also Hba1c was repeated and was confirmed again as very high. He was referred urgently to the local diabetes clinic and was seen within 24 hours. He was found to have highly elevated Anti-GAD antibodies, thus it was presumed he had Latent Onset Type 1 Diabetes Mellitus. He was started on insulin. He was followed up at the surgery; his Hba1c improved significantly with insulin and he reported his RP symptoms had markedly improved since starting insulin treatment.

Discussion

The key discussion point from this case is whether this patient had case of primary or secondary RP. Since primary RP is common in the general population, some of the reported associations of RP have remained unproven (5). There is no significant evidence to suggest Diabetes Mellitus as a known or common cause of secondary RP. The disease processes which can be attributed to secondary RP can involve a variety of mechanisms which can disrupt the normal regulation of regional blood flow to the digits and skin. Thus, the number of diseases and exposures associated with secondary RP is extensive (1,5).

Diseases commonly associated with RP include autoimmune rheumatic diseases such as systemic sclerosis, systemic lupus erythematosus (SLE), mixed connective tissue disease (MCTD), Sjögren's syndrome, and dermatomyositis/polymyositis. Various drugs or toxins can also precipitate or exacerbate RP such as amphetamines and chemotherapeutic agents. Hematologic conditions associated with RP include cryoglobulinemia, cold agglutinin disease, paraproteinemia, POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal protein, Skin changes) syndrome, and cryofibrinogenemia. Occupational and environmental causes of RP include vascular trauma (injury to the distal ulnar artery), the use of tools that vibrate, exposure to vinyl chloride, frostbite, and carpal tunnel syndrome. Endocrine conditions such as hypothyroidism, phaeochromocytoma and carcinoid syndrome are also associated with secondary RP (1,5).

Secondary RP should be considered strongly suspected in patients if any of the following features are present:

- Digital ulcers, gangrene, severe ischemia of one or more digits.
- Onset over the age of 30 years.
- Episodes that are intense, painful, or asymmetrical.
- History of, or clinical features suggestive of, a connective tissue disorder (such as sclerodactyly or pitting scars on the fingertips)
- Positive ANA blood tests.
- Abnormal nail-fold capillaries. (2)

Although primary RP usually occurs in patients who are generally well some comorbid conditions can occur which may aggravate attacks. These include hypertension, atherosclerosis, cardiovascular disease, and diabetes mellitus. A meta-analysis of observational studies by Garner et al. found that risk factors for primary RP included female sex, family history, smoking, manual occupation, migraine, and cardiovascular disease (3).

Diabetes is well known for its association with microvascular disease. Thus, it would be understandable to expect patients with primary RP and poor controlled diabetes to be prone to more frequent and severe attacks of RP (3,5). Thus, in theory if there was an improvement in diabetes control, as seen in this case, you would expect improvement in RP episodes as we saw in this a case, as significant improvement in Hba1c resulted in RP episodes becoming less frequent for the patient.

Conclusion

We concluded based on the above that it would be understandable to explain that this was likely a case of primary RP aggravated by undiagnosed Diabetes Mellitus.

Although primary RP usually occurs in healthy people, we feel it is important to consider any potential aggravating conditions when diagnosing patients with primary RP such as underlying diabetes, hypertension, and cardiovascular disease, particularly in patients with risk factors.

From this case we recommend that is important to take a detailed history and perform a thorough examination when a patient presents with RP symptoms to help establish any aggravating factors or potential secondary causes.

As part of initial work up we suggest requesting Hba1c blood test should be strongly considered when performing initial blood investigations for patients who present with symptoms suggestive of RP.

References

1. BMJ (2019) Raynaud's phenomenon. BMJ. https://bestpractice.bmj.com/topics/en-gb/1932
2. Belch J, Carlizza A, Carpentier PH, Constans J, Khan F, Wautrecht JC et al. (2017) ESVM guidelines - the diagnosis and management of Raynaud's phenomenon. Vasa 46(6), 413-423.
3. Garner R, Kumari R, Lanyon P, Doherty M, Zhang W (2015) Prevalence, risk factors and associations of primary Raynaud's phenomenon: systematic review and meta-analysis of observational studies. BMJ Open 5(3).
4. Stringer T, Femia AN (2018) Raynaud's phenomenon: Current concepts. Clin Dermatol. 36(4), 498-507.
5. UpToDate (2023) Clinical manifestations and diagnosis of Raynaud phenomenon. UpToDate. https://www.uptodate.com/contents/clinical-manifestations-and-diagnosis-of-raynaud-phenomenon