Case Report: Treating Erythromelalgia in a Healthy Teenage Male

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ASRA PAIN MEDICINE
25th Annual Pain Medicine Meeting
November 5-7, 2026 | Tampa, Florida
#ASRAFALL26

INTRODUCTION

- Erythromelalgia is a rare neurovascular disorder.
- Symptoms include intermittent burning pain, erythema, and warmth of the distal extremities. They are triggered by warmth and activity and are alleviated by cooling.
- Primary erythromelalgia usually occurs in the first decade of life and is often linked to mutations in SCN9A.
- Secondary erythromelalgia often has autoimmune, hematologic or iatrogenic etiology.
- Understanding pathophysiology and management is crucial, as its symptoms are debilitating and challenging to treat.

MATERIAL AND METHODS

- This case report is devoid of patient identifiable information. Thus, it is exempt from IRB review requirements as per Montefiore policy.

CASE REPORT

- A healthy teenage male was admitted for an acute onset rash with burning pain in his hands and feet. Symptoms were exacerbated by heat and alleviated by cold.
- Autoimmune, infectious, hematologic, and genetic work-up were all unremarkable except for positive influenza. MRI brain was negative except for posterior reversible encephalopathy syndrome (PRES).
- During hospitalization, he was closely followed by a multidisciplinary team for erythromelalgia.
- A multimodal pain regimen was initiated including tylenol, carbamazepine, gabapentin, lidocaine patches, EMLA cream, tizanidine, and IV morphine.
- He continued to have significant pain despite uptitration of medications. Gabapentin was transitioned to pregabalin. Carbamazepine was changed to amitriptyline, then to duloxetine secondary to a new mood disorder.
- Desensitization techniques were also utilized.
- The patient's pain gradually improved and he was discharged home on pregabalin and duloxetine after 6 weeks of hospitalization.
- Outpatient neurology visits demonstrated stable symptoms on duloxetine. Pregabalin was gradually discontinued due to marked sleep disturbance. Repeat MRI revealed resolution of PRES. PT, OT, and stress relief strategies were all continued.
- At 3-months post-discharge, he achieved complete symptom resolution.

DISCUSSION

- Erythromelalgia treatment often requires individualized trial-and-error (Ma et al., 2022).
- Patient education, lifestyle modification, safe cooling counseling, and identifying and treating secondary causes are critical.
- Multimodal pain regimens are often necessary, including topical analgesics and oral medications such as carbamazepine and mexiletine for SCN9A variants.
- Epidurals, sympathetic nerve blocks and SCS are considered for refractory cases.
- Multidisciplinary rehabilitation should be integrated early, including psychotherapy.

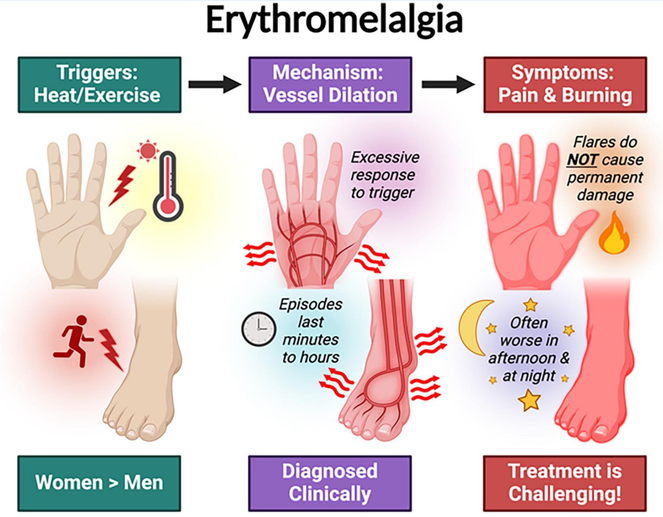


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