Erythromelalgia — A Life Of Fire And Pain

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By Elisabeth Antoine Crawford

Imagine holding your feet over a burning flame—and having to live with them there 24 hours a day. This is the pain of erythromelalgia (EM), a rare neurovascular disease affecting an estimated 1.3 people per 100,000 each year in the United States. The hallmark of EM is red, hot, burning feet, although many people experience symptoms in the hands and/or face as well. Flare-ups are essentially triggered by one thing: warmth. Because the heat threshold of the affected area becomes lowered, warm ambient temperatures that most people find comfortable are now perceived as excruciatingly hot. In addition, heat generated by such normally benign things as exercise, showers, and wearing socks is intolerable.

Relief is typically found by cooling the affected area, and many EM sufferers resort to soaking their feet in buckets of ice water. This is a dangerous practice; not only can it lead to breakdown of the skin, but it perpetuates a cycle of rebound flaring.

In a small percentage of sufferers, the condition is inherited, caused by a mutation of the SCN9A gene. While most cases are thought to be idiopathic, some are secondary to causes such as blood disorders, peripheral neuropathy, autoimmune diseases, and sports injuries. Many cases are severe, with constant, unrelenting pain; even the mild cases can be extremely debilitating. Remissions are infrequent, and because there is no single cause, there seems to be no single effective treatment.

In cases caused by myeloproliferative disease (e.g. polycythemia vera), EM seems to respond well to aspirin. Other patients have had varied success with antidepressants (namely, the SNRIs venlafaxine and duloxetine, as well as tricyclics such as nortriptyline), anticonvulsants (e.g. gabapentin, pregabalin), calcium channel blockers (e.g. diltiazem), beta blockers (e.g. propranolol), corticosteroids (e.g. prednisone), antihistamines (e.g. diphenhydramine, cyproheptadine), and opioids (e.g. morphine, oxycodone). The problem with these drug therapies is their inconsistency in effectiveness; a medication may provide relief for one person while making another worse.

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Pharmaceutical companies are currently focused on developing a drug to selectively block the sodium channel Nav1.7, which is encoded in the SCN9A gene; several clinical trials are currently in the works. In the meantime, particularly for those with inherited EM, the sodium channel blocker lidocaine remains a potential treatment in the form of topical patches, oral route (i.e. mexiletine), or

IV infusions. The drug carbamazepine, an anticonvulsant that acts as a sodium channel blocker, should also be considered.
Another promising treatment that has provided significant relief for some is the 8% capsaicin patch Qutenza. It has been theorized that one possible cause of EM may be the over-sensitization of the skin's heat-sensing capsaicin receptors, especially in cases preceded by nerve or tissue injury. Exposure to a high dose of capsaicin will paradoxically desensitize those receptors and may provide at least several months of pain relief.

Despite recent advances, EM still remains much of a mystery to the medical profession. The best step a patient can take for himself is to join a group like The Erythromelalgia Association (TEA) and bring all pertinent research to his doctor. Stumbling upon a successful treatment takes trial and error, but for many patients, it is possible to find relief and quell the burning fire.

— Elisabeth Antoine Crawford is the author of "Flavors of Friuli: A Culinary Journey through Northeastern Italy" and "Balance on the Ball: Exercises Inspired by the Teachings of Joseph Pilates." For more information on erythromelalgia, visit TEA's site: www.burningfeet.org.