Treatment of Erythromelalgia and Frostbite: A Case Report

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2:30 – 2:45 pm

Author and Co-authors:

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Objective:

Upon completion of the lecture, attendees should be better prepared to:
- Discuss erythromelalgia and complications as they relate to burn care.

Abstract:

Introduction: Erythromelalgia is an exceedingly rare disorder, with less than 30 primary cases reported in the United States. Individuals present with burning, erythema, and severe pain to the extremities. The lower extremities are most commonly affected with cases of severe wounds and even amputation. The disease presents either as a primary disorder or secondary with an associated myeloproliferative disorder.

Methods: Patient JK is a 17-year-old male who had been suffering from primary erythromelalgia for greater than ten years. He had been treated by multiple medical professionals with a variety of medications resulting in limited relief of symptoms. Ultimately his only source of control for the burning pain was to soak his feet in ice water for up to eighteen hours daily. This caused him to develop frostbite which became a necrotizing soft tissue infection complicated by severe chronic pain, deconditioning and impaired mobility, 60-pound weight loss, and psychosocial disorders.

Results: Patient JK was treated using the burn center's multidisciplinary approach with experts from burn care, dermatology, acute pain, chronic pain, physiatry, plastic surgery, and therapy. He underwent excision of the infected tissue and placement of allograft. Initially his pain was not well controlled despite a multimodal pain regimen with 10 different agents including ketamine infusions, so the patient cooled his feet resulting in a mycotic infection. This required revision excision and negative pressure wound therapy with intermittent instillations of amphotericin and mafenide acetate and subsequently had a 1:1 split-thickness autograft. The remarkable part of his care was the resolution of his erythema, all manifestations of pain, and any evidence of erythromelalgia following epidural placement and transition to methadone and medications that altered the sodium channels. At the time of discharge, he was ambulating over 350 feet, able to complete self-care with minimal assist, 100% graft take, and pain scores <3.

Discussion: While source control of a necrotizing soft tissue infection is a well understood principle, this case demonstrates two further points which may be applied

	to similarly complex patients in the future. First, patients with erythromelalgia have challenging wounds frequently managed with amputation, referral to vascular surgery, or chronic wound care centers when disease management might best be served holistically at a burn center. Secondly, placement of an epidural catheter resulted in complete resolution of symptoms and possible remission of a disease with cutaneous manifestations. The use of a long-term epidural with an implantable pain pump or spinal cord stimulator may serve as a viable means to control the disease process and pain associated with erythromelalgia.
Disclosure:	Joseph D. Giaimo – No Relevant Financial Relationships to Disclose Kelly Paulk – No Relevant Financial Relationships to Disclose Frank H. Lau – No Relevant Financial Relationships to Disclose Elizabeth H. Grieshaber – No Relevant Financial Relationships to Disclose Jeffrey E. Carter – Stock: PermeaDerm