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Lumbar Sympathetic Block for the Treatment of Intractable Erythromelalgia: A Case Report

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Background: Erythromelalgia is a rare pain disorder, which causes hyperperfused, red, and painful extremities. Symptoms can be so extreme that patients often resort to soaking their limbs in ice-cold water for hours on end and socially isolating themselves to avoid triggering an episode. Treatment is challenging, as many cases are refractory to medical management, which is first line. Invasive procedures are typically trialed next, some of which are still considered experimental.

Case Report: We describe a case of a 26-year-old woman with erythromelalgia, who remained symptomatic despite extensive pharmacologic therapy. Long-term relief was achieved following a lumbar sympathetic block.

Conclusions: Our case highlights the lumbar sympathetic block as a potentially effective treatment for erythromelalgia, especially in cases that are refractory to standard therapy.

Key words: Erythromelalgia, lumbar sympathetic block, case report

BACKGROUND

Erythromelalgia is a rare pain syndrome characterized by red, hot, hyperperfused, and painful extremities (1). Termed by Mitchell in 1878, erythromelalgia came from the Greek words erythros (red), melos (extremities), and algos (pain) (2). To further emphasize its distinguishing feature of warmth, the term "erythermalgia" was coined in 1938 by Smith and Allen. It is not until 1990 that the term erythromelalgia became commonplace (3).

Symptoms typically occur in the distal extremities (predominantly the feet and less often the hands), but rarely also in the neck, face, and groin (1,4,5). Episodes are typically triggered by elevated ambient temperatures or physical activity and can last from a few minutes to many hours. This often leads patients to cool the affected areas using various methods, including immersing them in ice-cold water (1,4,5). Some patients resort to soaking their extremities in ice water several hours a day (6), which leads to maceration of the skin, ulcers, necrosis, and secondary skin infections (5,7,8).

According to the Rochester Epidemiology Project, the incidence of erythromelalgia is 1.3/100,000, with a higher prevalence in women (3,9). Erythromelalgia is associated with a significantly decreased quality of life, as patients often experience severe anxiety related to trigger avoidance and may socially isolate themselves (1,8). Furthermore, patients with erythromelalgia have a higher mortality rate and an increased risk of suicide (4).

Treatment of erythromelalgia can be very challenging and often requires a multidisciplinary team (5,7,10). Pharmaceuticals are the first-line treatment, although many cases prove refractory to medications alone (10). These cases are often approached with invasive procedures, which vary in efficacy (10), and some of which are still considered experimental.

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We describe a case of erythromelalgia unresponsive to medications that successfully responded to lumbar sympathetic nerve blocks using bupivacaine and triamcinolone.

CASE REPORT

Written informed consent was obtained from the patient in compliance with the hospital's institutional review board.

Our patient was a 26-year-old woman with a medical history significant for Crohn's disease. She presented to our institution with pain and erythema involving both lower extremities. Symptoms began in April 2022, first in the toes, and then spread slowly to involve both feet. The pain was described as burning, shooting, and tingling in nature, and worse at night. The patient had been initially diagnosed with neuropathy and started on gabapentin 600 mg tid prior to presenting to our clinic, which provided minimal improvement. Her pain was tolerated only when she submerged both feet in ice water, initially for a few hours at a time, but then eventually progressing to nearly continuous immersion throughout the day.

Two months later, she developed bullous skin lesions from a cold-induced injury. This led to skin breakdown and superimposed bacterial infections (Fig. 1). A skin biopsy done by dermatology revealed thrombocytosis and an elevated white blood count containing multiple atypical cells, including myelocytes and metamyelocytes. Electromyography showed sensory peripheral polyneuropathy.

During her first visit to the pain clinic, the patient was already being treated with aspirin, long-term antibiotics for the superimposed bacterial infection, gabapentin 600 mg tid, and hydromorphone 4 mg tid. Her pain was minimally controlled on this regimen, so diclofenac 75 mg bid and amitriptyline 50 mg daily were added to her medications, and gabapentin was increased to 800 mg tid. On physical exam, her bilateral lower extremities exhibited some allodynia and sudomotor symptoms. Medical management did not help with her pain significantly, and she had tried hydromorphone up to 48 morphine milligram equivalents daily, which only slightly reduced her pain.

A bilateral lumbar sympathetic block was planned; however, it was initially denied by insurance. After multiple appeals and subsequent approval, the patient underwent a bilateral lumbar sympathetic block at the L3 level with bupivacaine and triamcinolone. She reported immediate pain relief after the procedure.

Approximately 4 months later, the pain started to return, prompting a second bilateral lumbar sympathetic block at the same level, again with excellent pain relief. She was able to discontinue hydromorphone and reduced her gabapentin dosage to 300 mg tid. The patient has not required a subsequent lumbar sympathetic block since and has not required follow-up in the pain clinic for over 3 years.

DISCUSSION

Our case report demonstrates the potential efficacy

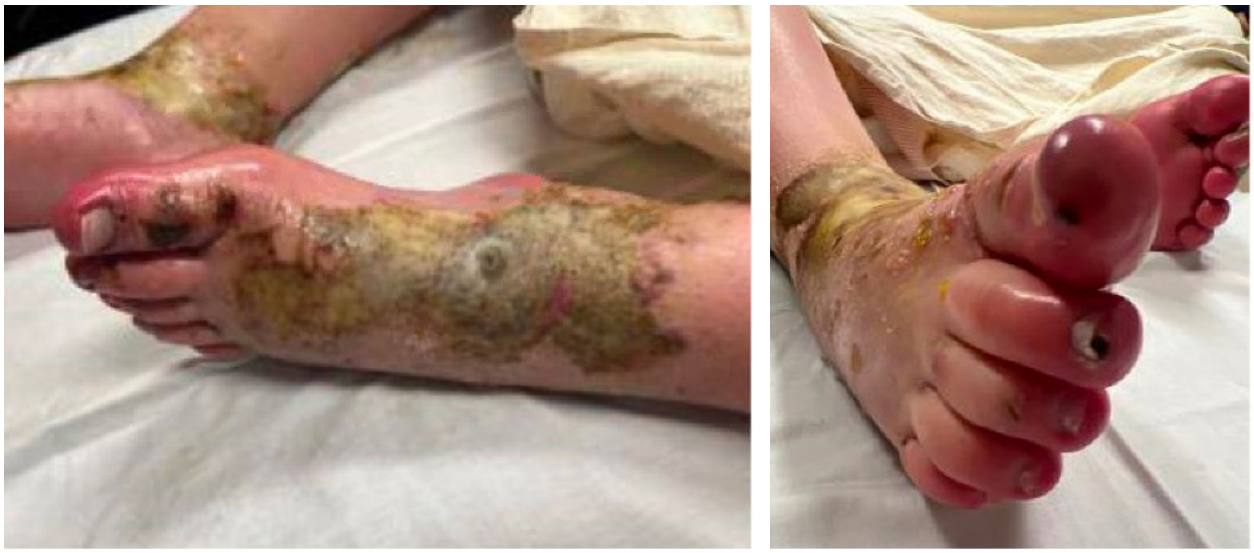

Fig. 1. Skin breakdown resulting from cold-induced injury, with superimposed bacterial infection.

of lumbar sympathetic blocks as a treatment option for patients with erythromelalgia. To our knowledge, this represents one of the few reported cases describing the success of lumbar sympathetic blocks for the symptomatic control of erythromelalgia, a condition that has historically been difficult to treat.

Erythromelalgia can be categorized as primary, secondary, or idiopathic (2,11). Primary, or inherited, erythromelalgia is due to a genetic mutation, and, according to one study (12), accounts for approximately two-thirds of all cases. An autosomal dominant mutation in the sodium voltage-gated channel alpha subunit 9 (SCN9A) gene that codes for the voltage-gated sodium channel, Nav1.7, has been identified as a cause of primary erythromelalgia (2). These sodium channels are found in sympathetic and nociceptive sensory neurons of the dorsal root ganglion, and it is thought that this mutation leads to hyperexcitability of these neurons and causes disproportionate pain (5,10,13).

Secondary erythromelalgia is related to an underlying condition, such as certain autoimmune and myeloproliferative disorders, infection, medications, neuropathies, and metabolic disorders, with a less well-understood pathophysiology, though it is believed to involve both vascular and neuropathic mechanisms (10,13,14). Neurophysiologic studies (5,14) involving nerve conduction studies and needle electromyography suggest small fiber neuropathy, with large fiber involvement observed in approximately 50% of cases. Around 90% of patients have abnormal thermoregulatory sweat testing, indicating small fiber neuropathy (5). It remains unclear whether the small fiber neuropathy is a cause or consequence of erythromelalgia (5).

It is also believed that patients with erythromelalgia have vascular abnormalities, including increased total perfusion via abnormally open arteriovenous shunts, with concomitant reduced nutritive perfusion, due to abnormal capillary flow. This paradoxically results in tissue hypoxia and ischemic pain despite elevated overall blood flow and increased perfusion (8,14,15).

The diagnosis of erythromelalgia is often made based on the classic triad of burning pain, erythema, and hyperperfusion (1), as well as the hallmark features of heat intolerance and symptomatic relief with cooling (2,8). Since symptoms are usually intermittent, some have proposed provocation testing with warming (32°C to 36°C), with a positive test if symptoms are elicited at this temperature (12). Skin biopsies are not typically diagnostic (2) and often reveal nonspecific or secondary changes due to chronic cold exposure (1). Ruling out underlying conditions is essential to differentiate primary from secondary erythromelalgia (2). Genetic testing for SCN9A variants may confirm primary erythromelalgia (2), though some inherited cases lack identifiable SCN9A mutations (5).

The differential diagnosis for erythromelalgia includes complex regional pain syndrome (CRPS). Although both may present with burning pain, erythema, and swelling, erythromelalgia is typically bilateral and features symptom-free intervals between episodes. Additionally, erythromelalgia usually occurs spontaneously, instead of after an injury (8,14). Furthermore, CRPS is typically aggravated by cold exposure, whereas erythromelalgia is alleviated by cooling (14). Other differential considerations include peripheral neuropathies due to various causes (12), especially given that some patients report burning pain for months prior to visible erythema (8).

Successful management of erythromelalgia is often difficult, with treatment outcomes depending on erythromelalgia subtype and specific NaV1.7 mutations (13). There are only rare cases of reported complete resolution of symptoms (10). Initial treatment of secondary erythromelalgia involves management of the underlying condition (2). For both primary and secondary cases refractory to underlying treatment, medical therapy is first line (10).

Initial therapy often involves topical agents, including lidocaine, amitriptyline with ketamine, midodrine, and gabapentin creams. These should be used for a minimum of 4 weeks before escalating to systemic therapy (5). Systemic therapy should address both neuropathic and vascular components, given the dual-pathway hypothesis of the condition (14). Aspirin is often trialed early due to its safety and reported efficacy in erythromelalgia associated with myeloproliferative disorders (2,5,8). Other systemic medications include nonsteroidal anti-inflammatory drugs (especially piroxicam), gabapentin, serotonin noradrenaline reuptake inhibitors, tricyclic antidepressants, antihistamines, beta-blockers, calcium channel blockers, magnesium, and prostaglandins (2,5,8,14). Sodium channel blockers, such as carbamazepine and mexiletine, have shown benefit in patients with primary erythromelalgia who have certain genetic mutations (13).

Some patients respond favorably to systemic steroids, and it is recommended in patients who have had a sudden onset of erythromelalgia or who have had symptoms for less than a year (5,12). Early administra-

tion may help prevent nociceptive remodeling and mitigate inflammatory neuropathy, especially in patients with erythromelalgia secondary to traumatic, surgical, or infectious causes (5). Patients are also advised to minimize exposure to known triggers while maintaining functional activity when possible—for example, substituting swimming for running. Cold immersion should be limited to fewer than 4 sessions per day and under 10 minutes each time (5).

Despite these approaches, many cases remain refractory to pharmacologic treatment, prompting consideration of interventional therapies. In such cases, interventional procedures are often considered (10). Invasive modalities include sympathetic blocks, epidurals, peripheral nerve blocks, spinal cord stimulators, and neurosurgical interventions, such as thalamic electrode placement (10).

A small number of case reports (6,7,16) demonstrate the potential benefit of lumbar sympathetic block in refractory erythromelalgia. In one report (6), a patient failed multiple treatments—including corticosteroids, aspirin, antiserotonergic agents, intravenous phentolamine, and epidural blocks—before achieving immediate and durable pain relief following lumbar sympathetic block with anhydrous ethanol. Another patient (16) experienced partial benefit from lumbar sympathetic ganglion blocks and was later successfully treated with pulsed radiofrequency ablation of the lumbar sympathetic ganglion, reducing both pain and medication requirements.

These cases reinforce the potential role of lumbar sympathetic blocks in treating refractory erythromelalgia and underscore the need for further research into sustained treatments, such as pulsed radiofrequency of the lumbar sympathetic ganglion. Furthermore, in our case, the patient's commercial insurance initially denied the procedure due to its classification as investigational. While eventual approval was obtained, the procedure remains classified as "experimental." Future studies could establish lumbar sympathetic blocks as a validated treatment for erythromelalgia and reduce associated insurance and cost-related barriers.

Lumbar sympathetic blocks induce vasodilation and increase regional blood flow, likely contributing to the observed symptom relief in our patient (10,16). Additionally, given the theorized autonomic dysfunction in erythromelalgia, this further supports the mechanistic rationale for lumbar sympathetic blockade (8).

Physicians should remain aware of available treatment modalities for erythromelalgia and consider lumbar sympathetic blocks for patients unresponsive to medical management.

CONCLUSIONS

Erythromelalgia is a rare yet frequently debilitating chronic pain condition. Achieving symptomatic control can be challenging and often requires more than conservative medical therapy. Interventional procedures are frequently pursued in refractory cases, although several remain classified as experimental. We illustrate a case of erythromelalgia successfully treated with a lumbar sympathetic block, despite initial denial by the patient's insurer due to its investigational status. Lumbar sympathetic blocks should be considered as a viable option in cases of intractable erythromelalgia.

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