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Successful Novel Treatment of Primary Erythromelalgia with Popliteal Sciatic Nerve Block and Peripheral Nerve Stimulation: A Case Report

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Background: Primary erythromelalgia (PE) is a rare, hereditary disorder characterized by chronic burning pain in the extremities and commonly linked to mutations in the SCN9A gene. The management of erythromelalgia-associated pain remains challenging, since there are currently no standardized guidelines for the treatment thereof due to the rare and refractory nature of the disorder.

Case Report: A 17-year-old boy with SCN9A-related PE presented with intractable bilateral lower extremity pain complicated by ulcerations and osteomyelitis. After not responding to multiple medications and interventions, he experienced significant relief upon receiving a popliteal sciatic nerve block effected with lidocaine and triamcinolone. Afterward, peripheral nerve stimulation (PNS) was implanted, leading to lasting pain relief, improved functionality, and wound healing for the patient.

Conclusion: PNS offers a promising treatment for refractory PE, warranting further investigation to establish the role of this technique in managing this challenging condition.

Key words: Primary erythromelalgia, peripheral nerve stimulation, sciatic nerve block, case report

BACKGROUND

Primary erythromelalgia (PE) is a rare hereditary disorder characterized by recurrent intense burning pain, erythema, and warmth of the distal extremities, provoked by warm temperatures and exercise, and relieved by cool temperatures and elevation (1). Classically, PE is caused by mutations in the SCN9A gene, leading to hyperexcitability of peripheral nociceptive neurons due to sodium-channel subunit Na(v)1.7 dysfunction in the nociceptive pathway (1). Secondary causes include myeloproliferative disease, neoplasms, adverse medication reactions (bromocriptine and calcium-channel blockers), or rare environmental toxins (2). In the United States, erythromelalgia has been found to occur at a mean age of 57 years, with a higher prevalence overall in ages over 65, and no specific gender preference (2,3). Not following any explicit distribution, there is also a peak in incidence at 14 years, with majority pediatric onset during adolescence (4). PE has been shown to occur approximately 5 times more frequently than secondary erythromelalgia, based on the only population study of PE that has been performed (5).

Erythromelalgia is considered a clinical diagnosis of exclusion based on the aforementioned criteria and requires ruling out of a broad differential diagnosis. Vasculopathy, Fabry disease, Raynaud's phenomenon, systemic lupus erythematosus, and arteriovenous insufficiency are all critical considerations to rule out

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on a case-by-case basis. Ultimately, a combination of clinical correlation and genetic testing involving next-generation or Sanger sequencing of the SCN9A gene is the only available confirmatory test for PE (2).

Initial conservative treatment consists of symptomatic management, most frequently cooling of affected extremities as needed, and pharmacological management. Sodium-channel blockers are commonly utilized, including topical and intravenous lidocaine, though these have been shown to relieve pain in up to only 55% of patients (6). Few case reports show augmented or improved pain control ensuing from the administration of oral mexiletine (2). Similarly, carbamazepine has demonstrated efficacy in relieving painful episodes and the frequency thereof in case reports and case series, but there are no large-scale studies demonstrating this effect (7). Gabapentinoids have also shown efficacy in case reports and series and may be likely to be trialed by physicians; however, the literature of longitudinal or retrospective studies concerning these medications' use as a treatment for PE is likewise rare (8-10).

Ascending the ladder to procedural intervention, sympathetic and peripheral blocks, epidural injections, dorsal root ganglion (DRG) stimulation, and spinal cord stimulation (SCS) have been utilized to mixed success in managing the disease (10,11). Notably, there are no described cases of treatment with peripheral nerve stimulation (PNS) in the literature. While the full mechanism of action of PNS is not entirely clear, studies have supported that its effects work through both central and peripheral pathways. It has been suggested that PNS engages the gate control pain theory by activating the non-nociceptive large-diameter Aβ fibers, which then leads to the activation of inhibitory dorsal horn interneurons on the nociceptive Aδ and C fiber activity (12). Additionally, PNS has been proposed to obstruct the transmission of peripheral nociceptive signals by interrupting both afferent and efferent fibers, downregulating excitatory neurotransmitters, and reducing both peripheral and central sensitization (13). Ultimately, though the American Society of Interventional Pain Physicians (ASIPP) guidelines highlight the use of PNS for neuropathic conditions similar to erythromelalgia, evidence for the treatment's use for neuropathic pain remains limited to case reports only (14).

Due to the rarity of this condition and the challenges associated with its refractory pain, there are currently no established guidelines for the treatment of PE. Here, we describe a novel case of using a sciatic nerve block and PNS to treat erythromelalgia pain.

CASE PRESENTATION

A 17-year-old boy with SCN9A-related inherited PE presented with chronic, debilitating, intractable burning pain in bilateral lower extremities. This pain had progressively worsened since age 7. His condition was complicated by a recent hospitalization for foot ulceration and fungal osteomyelitis from the prolonged direct exposure of his feet to air conditioners for pain relief. On examination, he was found to have bilateral leg erythema with ulceration and healing wounds (Fig. 1).

Prior to his initial presentation, he had already been unresponsive to numerous medication trials intended to alleviate his pain, including topical analgesics, anti-inflammatories (aspirin, meloxicam, nabumetone), antidepressants (duloxetine), gabapentinoids (gabapentin), anticonvulsants (carbamazepine), sodium-channel blocker (mexiletine), and opioids (morphine, methadone, oxycodone, tapentadol). Further treatment with lumbar sympathetic blocks and epidural infusions provided inconsistent to no relief.

He subsequently underwent a bilateral popliteal sciatic nerve block using 5 mL of 1% lidocaine mixed with 20 mg of triamcinolone under ultrasound guidance. This treatment provided him with 70% pain relief for 4 weeks. The patient noted that the treatment was the most successful one to date. A repeat sciatic nerve block was equally efficacious. Given the success of these blocks, a decision was made to trial PNS targeting the patient's bilateral popliteal sciatic nerves for a 60-day period, with the goals of reducing his pain by over 50%, allowing his wounds to heal, and improving his functionality.

The SPR® SPRINT® PNS system and its standard Micro-Lead™ were used. The procedure was performed with the patient in prone position. The patient's painful region was outlined with a marker, and the popliteal fossa area was prepared in a sterile fashion. Ultrasound was used to identify the sciatic nerve proximal to its bifurcation into the tibial and common peroneal nerve at the proximal popliteal fossa, using landmarks of the tibial artery and vein between the biceps femoris, semimembranosus, and semitendinosus. Using a single-incision technique, the percutaneous sleeve and stimulating probe system was assembled and inserted through an introducer needle under ultrasound guidance to a location proximal to the sciatic nerve. Multiple stimulation testing with repositioning of the stimulation probe was then performed until the patient experienced paresthesia over the distribution of his pain in the lower

extremities. The stimulating probe was then removed, and a percutaneous lead was guided through the needle to its location. Repeat stimulation testing was performed there to ensure appropriate delivery of the lead. The introducer needle was then removed, and the exposed end of the percutaneous lead was attached to the external stimulator unit with repeated testing. After proper placement was confirmed, the lead was anchored to the skin and attached to the connector block and external unit, with one final stimulation testing to confirm the desired response.

Prior to the implantation, the patient reported 9/10 pain on a numeric pain rating scale and was taking 150 mg of mexiletine 3 times daily, 1,200 mg of gabapentin 3 times daily, 60 mg of duloxetine daily, and 50 mg of tapentadol daily as needed. Two weeks into the trial period, he reported that his pain had decreased to 2-3/10 without requiring him to place his legs on the air conditioner or use his as-needed tapentadol. The initial PNS settings were frequencies of 40 Hz on the left and 80 Hz on the right, with the pulse width (30 to 200 μs) and amplitude (0 to 30 mA) adjusted to the lowest effective settings by the vendor representative. The intensity level was set to 40 (on a scale of 0 to 100). His trial period was unremarkable other than an accidental lead dislodgement one month after the implantation, leading to a recurrence of pain up to 7/10 in severity, which improved back to 2-3/10 after lead replacements. The pain fluctuated during the trial period and improved after programming adjustments with an increase of frequency settings to 120 Hz bilaterally and intensity level to 60. Ultimately, by the conclusion of the 60-day treatment period, PNS was found to successfully alleviate his pain by providing him overall 70% pain relief with average pain scores of 2-3/10, and he was able to be weaned off tapentadol completely. The further success of PNS was demonstrated by improvement in the patient's disability, since he was able to return to school, sleep through the night without requiring his usual quetiapine, renew his hobby of drawing, and walk outside more frequently, all of which were monumental to his quality of life. Additionally, he stopped directly exposing his feet to air conditioning, which allowed his existing wounds to better heal and close at both his 3- and 6-month visit

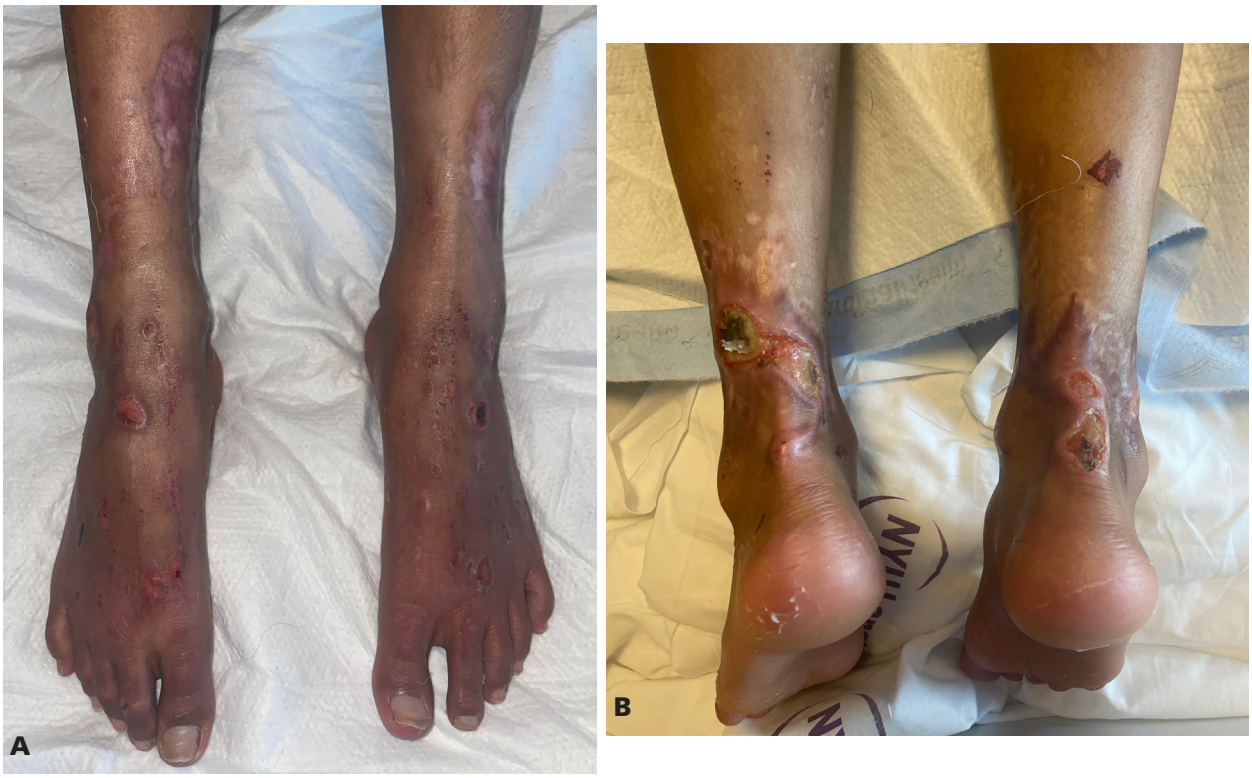

Fig. 1. Pre-procedural bilateral lower extremity wounds.
Anterior and posterior view of the patient's bilateral lower extremity wounds prior to PNS treatment. There are multiple locations of ulcerations and open wounds caused by excessive direct cold exposure, pressure, scratching, and immobility.

to the dermatologist (Fig. 2). The patient's symptoms fluctuated after the PNS system was removed, but he noted that the treatment was his most successful, and he reported an overall improvement in his pain and burning by 50-70% at 2- and 6-month follow-up.

DISCUSSION

This case presents a novel treatment for intractable PE-caused pain that has failed multiple courses of conservative and interventional treatments. PE is a difficult-to-treat condition that is often accompanied by complications of ulcerations, gangrene, and disability due to refractory pain that patients manage by immersing their affected limbs in ice water or with cold exposure. Conservative and pharmacological forms of management are often not sufficient to control patients' pain and symptoms adequately, and the evidence backing the typical options of sodium-channel blockers and gabapentinoids are based on case reports only. The difficulty of treatment may be linked to the etiology of the disease (10,15-16). Sodium-channel blockers are the most commonly utilized pharmacological option, though their utility may be limited by heterogeneity in the SCN9A mutation in PE. Only a minority of PE cases carry this specific mutation at 15%, with the remainder of primary cases having an otherwise idiopathic cause (9).

Table 1 shows a list of case reports published to date regarding percutaneous neuromodulation for management of PE. Three case reports have been published

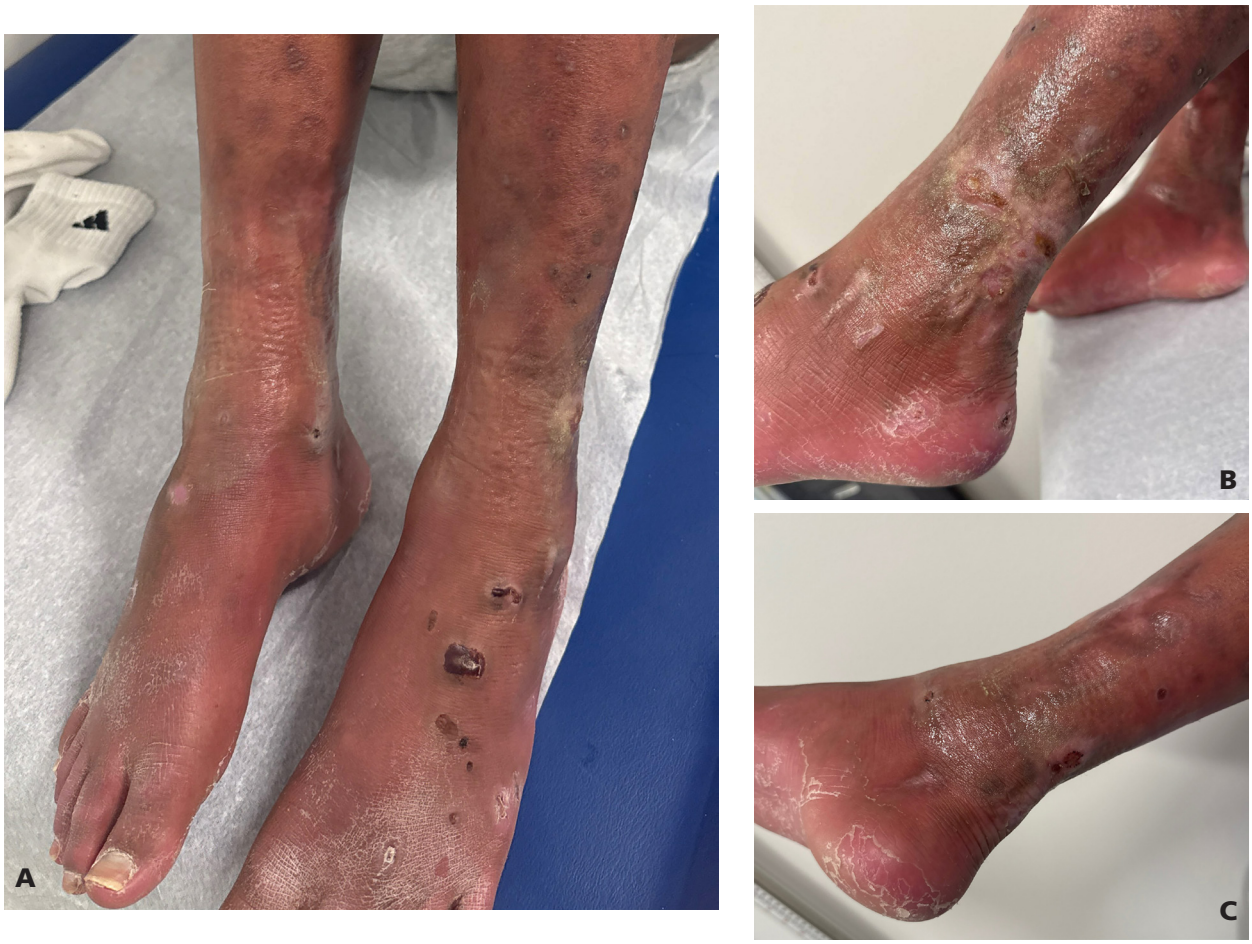

Fig. 2. Post-procedural bilateral lower extremity wounds.
Anterior and posterior view of the patient's bilateral lower extremity wounds on subsequent follow-up after treatment course of peripheral nerve blocks and PNS. These modalities provided significant pain relief for the patient, which allowed the wounds to heal and close.

Table 1. List of currently published cases that describe neuromodulation treatment for primary erythromelalgia (PE) pain.

Authors/ Reference	Age/ Gender	Modality	Follow-up (Months)	Outcomes
Maung et al (current case)	17, M	PNS—sciatic nerve (popliteal fossa)	6	Bilateral ankle and feet pain relief
Lam et al (16)	21, M	DRG—C2 and T9	12	Bilateral hand and feet pain relief
Fan et al (17)	12, F	SCS—T11	12	Bilateral feet pain relief with cessation of pain medications
Eckmann et al (18)	20, M	SCS—C3 and T8	6	Bilateral upper and lower extremity pain relief with cessation of pain medications
Patel et al (19)	15, F	SCS—T12	24	Bilateral feet pain relief
Hagedorn et al (10)	70, F	DRG—S1	3	Bilateral feet pain relief (80% improvement)

that show the successful management of intractable PE-caused pain with SCS. Each case report mentions at least 6 months of significant pain relief (17-19). In 2 cases, one from Hagedorn et al and the other from Lam et al, bilateral DRG stimulation provided relief from intractable pain at the 3-month and 12-month follow-ups (10,16).

With these considerations in mind, one of the challenges to the treatment of erythromelalgia is inconsistent response to therapeutic agents across patients. In addition to the chronic pain, disability, and consequent psychological impact that PE patients are likely to endure, the disease carries the risk of causing further medical complications. Patients are likely to self-administer cooling techniques, such as exposing the affected extremities to cold for prolonged amounts of time, which often leads to skin maceration, infection, and even necrosis. These difficulties can further progress to peripheral ischemia, gangrene, and even limb amputation (1,9). Such complications frequently require escalation in medical care and limit patients and physicians from performing further pain management interventions.

Among the advantages PNS offers over pharmacological treatment, genotypic variation among patients with PE should not impact efficacy. Because the patient in the present study showed relief with sciatic nerve blocks and was at risk of infection due to a history of osteomyelitis, a PNS was chosen rather than SCS implantation to treat his PE. Our patient experienced significant improvement in subjective pain and quality of life during the 2-month period of temporary PNS placement and on subsequent follow-ups. However, several limitations should be acknowledged, including the single-case nature of this report, the relatively short follow-up duration, and the lack of objective outcome measures.

A particular advantage PNS poses over SCS is reduced invasiveness and thereby fewer implications for outcomes and complications. Whereas SCS involves surgical placement of a lead within the epidural space, PNS are placed percutaneously near the target nerve, often with ultrasound guidance. The anatomy of PNS placement thereby eliminates the risk of epidural hematoma formation, rarely associated with SCS. Neurologic injury is similarly rare in SCS (occurring at a rate no greater than 0.25%), whereas ASPN guidelines and studies therein report no serious adverse events related to PNS placement (20). A literature review has found that the infection rate in PNS placement is between 0.03 and 0.83 per 1,000 indwelling days, which translates to 0.01-0.30% over a 12-month period (21). A separate US-based study reported a 12-month SCS device-related infection rate of 3.11% in a cohort of 6,615 patients (22). Though there are no direct studies comparing the 2 methodologies, PNS may offer an enhanced safety profile for patients who have an increased infection risk, such as this case.

CONCLUSIONS

We would like to highlight regional blocks and PNS as possible treatment options for erythromelalgia, as successfully demonstrated in this patient, who had not previously responded to the aforementioned treatments discussed in current literature. Though some literature reviews discuss interventional modalities in erythromelalgia, none at present discuss the utility of PNS for managing this condition. Neither do longitudinal studies. The PNS modality may play a greater role for PE patients than is currently utilized and necessitates further research.

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Author Contributions

CM, RD, AD, SS, and OA all made contributions in the writing, editing, and research for this article. The principal investigator, JL, oversaw each step of the writing process.

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