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Stellate Ganglion Block for the Treatment of Facial Erythromelalgia: A Case Report

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Background: Erythromelalgia (EM) is a rare disease characterized by recurrent episodes of erythema, pain, and burning dysesthesia, most often involving the extremities. A rare, atypical presentation involves the face. There are no current guidelines, with overall lack of consensus for the treatment of EM, and therapies are often formulated on a case-by-case basis.

Case Report: We describe the case of a 48-year-old woman who presented with facial EM causing erythema and burning pain with significant impact on quality of life, and was successfully treated with stellate ganglion block.

Conclusion: This case highlights that stellate ganglion blocks can be an effective treatment option for patients with facial EM, alongside other conservative therapies that may include medical management.

Key words: Erythromelalgia, facial erythromelalgia, stellate ganglion block, ultrasound guidance

BACKGROUND

Erythromelalgia (EM) is a rare disease characterized by recurrent episodes of erythema, pain, and burning dysesthesia, most often involving the extremities (1). The disease was first described in the 1870s by Mitchel (2), labelling it "erythromelalgia" using Greek nomenclature to represent the various components of symptoms associated with the disease, namely, erythros (redness), melos (extremity), and algos (pain). The disease is often classified as primary (idiopathic) or secondary when associated with myeloproliferative disorders (3). However, there is a lack of agreement regarding classification in the literature; Drenth et al (4) further classify EM based on responsiveness to treatment and associated diseases: EM (associated with thrombocythemia and responsive to aspirin), primary EM (idiopathic or inherited), and secondary EM (aspirin-resistant). EM most often involves the lower extremities, followed by the upper extremities, and finally the head and neck (5). An extremely rare, atypical presentation of EM involves isolated facial involvement (1).

To our knowledge, there exists only a handful of publications describing isolated facial EM. Furthermore, only 2 case reports describe the use of a stellate ganglion block (SGB) to treat EM pain (11,12). In this case report, we discuss the case of a 48-year-old woman with facial EM who was successfully treated with SGB, in addition to medication therapies.

CASE

The patient is a 48-year-old woman with a past medical history relevant for mitral valve prolapse, chronic lower back pain (secondary to lumbar facet arthropathy), Raynaud's disease, bipolar 1 disorder, and anxiety. She presented to the pain management center complaining of painful facial flushing and burning sensation for the previous 18 months. She described her pain as intense and episodic, lasting 5+ hours dur-

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ing exacerbations, rated as 10 of 10 on the Numeric Pain Intensity Scale, and localized to the malar regions primarily, rarely including the bilateral upper extremities (Fig. 1). Her symptoms were aggravated by warm conditions and the wearing of face masks. Her pain was relieved with cooling packs although this would often cause rebound flushing. She reported a significant impact on her quality of life, including a complete aversion to leaving her home during warm weather and no longer being able to prepare meals using the stove or oven due to exposure to heat. In addition to avoiding heat exposure, she would maintain a temperature of 60 degrees Fahrenheit in her home even during the winter season. Physical exam between episodes was unremarkable, with no erythema, warmth, or skin lesions noted. However, she provided photographs of herself during these episodes (Fig. 1) that demonstrated erythema, and during which she described flushing and warmth. She underwent an extensive workup, which included complete blood count, metabolic panel, thyroid stimulating hormone, rheumatoid factor, cyclic citrullinated protein antibody, urine and serum protein electrophoresis, urine metanephrines, cortisol level, complement level, anticardiolipin, cryoglobulin, chromogranin A, inflammatory/vasculitic markers, radioallergosorbent testing, and beta-2 glycoprotein, all of which were normal. Imaging included computed tomography (CT) of the abdomen and pelvis, which also was unremarkable. After eliminating medications with risk for photosensitivity and exclusion of possible causes (including rosacea, systemic lupus erythematosus, carcinoid syndrome, pheochromocytoma, etc.), a diagnosis of facial EM was made.

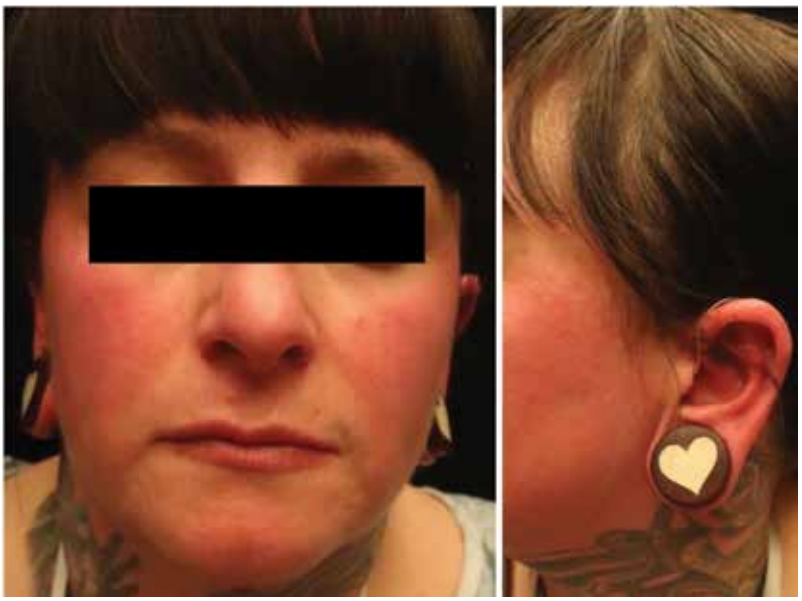

Fig. 1. Photographs demonstrating facial flushing during clinic visit.

Multiple medication classes were trialed to control her symptoms and eventually she was maintained on a combination of oral medications, including a beta-blocker (carvedilol), hemorheologic agent (pentoxifylline), $\alpha 2\delta$ agonist (gabapentin), and antiplatelet (aspirin). Topical agents, such as local anesthetic (lidocaine 5%) creams and compounded formulas containing a N-methyl-D-aspartate (NMDA) antagonist (ketamine) and tricyclic antidepressant (amitriptyline) were trialed to help alleviate symptoms during flare-ups with minimal relief. Ineffective classes of medications had included an alpha-adrenergic agonist (clonidine) and serotonin norepinephrine reuptake inhibitor (duloxetine).

In addition to medication management, she was offered a SGB. After obtaining informed consent, she was placed supine with her head raised to 30 degrees. In a sterile manner, a linear ultrasound probe was used to identify the relevant landmarks, including the transverse process of C6 (Chassaignac's tubercle), the sternocleidomastoid muscle, the thyroid gland, the carotid artery, the internal jugular vein, and the longus colli muscle. A 20-gauge, 50-mm echogenic needle (Pajunk®, Alpharetta, GA) was advanced in plane to rest posterior to the carotid artery and anterior to the longus colli muscle (Fig. 2). With intermittent aspiration that was negative for heme, air, or cerebrospinal fluid, 9 mL of 0.125% bupivacaine with 10 mg of dexamethasone was injected with satisfactory spread in the desired prefascial plane. She had this procedure performed bilaterally, at 3 separate appointments.

She reported significant symptomatic relief, with > 95% improvement reported on pain and symptom assessment at the 2-month follow-up. Prior to the SGB, her pain during EM flares was rated as 10 of 10 and would remain constant for 5+ hours. Post SGB, her pain was reduced to a range of 0 to 3 of 10, and the frequency of flares was reduced along with shorter duration, decreased to 2 to 3 hours, sometimes with no associated pain at all. She also stated that she had sig-

nificant improvement in her quality of life. She ultimately had improvement in her symptoms for approximately 4 months post SGB. She was advised that SGB may be repeated as necessary for symptomatic relief.

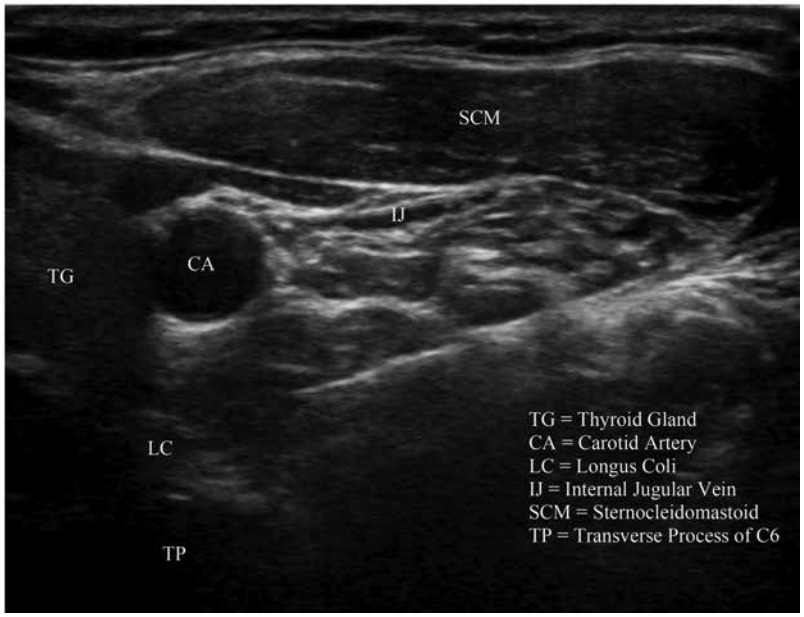

Fig. 2. Ultrasound demonstrating the performed stellate ganglion block.

DISCUSSION

The pathophysiology of EM is not fully understood; however, 2 pathophysiologic processes have provided plausible explanations for EM. First, a genetic gain-of-function mutation in the gene SCN9A that encodes voltage-gated sodium channel proteins in sensory neurons has been linked to primary EM (6). This results in hyperexcitability via a reduction in action potential threshold and an increase in frequency of firing of sodium channels in the dorsal root ganglion (7). Second, a component of vascular dysfunction has been proposed as a cause for EM, where vasodilation and resultant shunting in affected areas leads to tissue hypoxia (8). In a study of 61 patients with EM, in which blood flow to the skin and microvasculature were assessed with patients being placed in hot conditions, Littleford et al (14) suggested that the vascular component of EM is more likely to be due to vasoconstriction causing the initial insult and a subsequent reactive vasodilation causing the typical symptoms. This may be corroborated by the relatively high incidence of patients with EM suffering from concomitant Raynaud's disease.

The diagnosis of EM is primarily a clinical diagnosis of exclusion, as there is a lack of confirmatory diagnostic tests. EM results in high morbidity and mortality; a follow-up of 168 patients diagnosed with erythromelalgia reported an inability to walk/stand for prolonged times in 50% of patients, with 12.5% of patients having to give up jobs. The same study reported suicide as a common cause of death secondary to intractable pain (10).

There are no current guidelines for the treatment of EM and therapies are often formulated on a case-by-case basis. Commonly used medication classes include antiplatelets, nonsteroidal anti-inflammatory drugs, prostaglandins, anticonvulsants, sodium channel blockers, beta-blockers, antidepressants, antihistamines, and topical agents (9), with responsiveness to therapy being highly variable between patients. In cases that are refractory to medical treatment, interventional procedures revolve around sympathetic blockade, spinal cord stimulation (SCS), and in rare occasions neurosurgical interventions have been reported, including deep brain stimulation and ablation of the ventroposterolateral/centromedian nuclei. A literature review published by Chinn et al (13) suggests that epidural infusions may be an appropriate treatment for patients who have failed medical therapy. The review reported varying degrees of symptomatic relief. Two case reports have demonstrated successful treatment of EM with the use of botulinum toxin injections (15,16). Due to disease rarity, there is a lack of randomized controlled trials (RCTs) and standardized treatments comparing efficacy. Efficacy of interventional treatments in treating EM is limited to case reports and small-scale studies. The paucity of RCTs makes it difficult to judge which treatments would be most appropriate in those who fail conservative management. The mechanism by which interventional treatments provide relief in EM is likely multifactorial.

CONCLUSION

The stellate ganglion provides sympathetic innervation of the ipsilateral upper extremity and face. It may seem that the vasodilatory side effect of sympathetic blocks would be counterintuitive, but symptomatic relief may indicate a degree of peripheral sensitization involved in EM. The effect of local anesthetics that are

commonly used in such procedures and reported benefit of Botox injections support the channelopathy pathophysiology of EM, with the thought that sensitization is prevented via inhibition of pain-mediating neurotransmitters including substance-P, calcitonin-gene related peptide, and glutamate.

Each patient's treatment algorithm should be approached on a case-by-case basis, with trials of appropriate medical therapies and interventional procedures, as no uniform treatment guidelines exist. The employment of SGB for treatment of intractable upper extremity/facial EM may be worth considering, based on our experience. Further research is needed to assess the efficacy of SGBs in the treatment of EM.

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