

Krista Dicks, BA, RMT, CLT,¹ Philippe Rizek, BSc, MSc, MD²

¹Barrie, ON, Canada; ²University of Western Ontario, London, and Canadian Society for Life Science Research, Toronto, ON, Canada

Background: Erythromelalgia is characterized by temperature-dependent redness, pain, and warmth in one or more extremities. It may be a primary disease, or it may occur secondarily because of underlying illness. It is a chronic, debilitating condition often resistant to medical treatment.

Purpose: The present report evaluates massage as a complementary therapy to reduce pain and other symptoms associated with erythromelalgia.

Participant: A 31-year-old female with a longstanding history of erythromelalgia bilaterally in the lower extremities presented with complaints of acute pain exacerbation, anxiety, decreased quality of sleep, and difficulty with activities of daily living for prolonged periods of time. She had no previous experience with massage therapy or any other complementary therapies.

Intervention: Massage therapy was introduced over the course of 9 treatments, each 1 hour in duration, using various massage therapy techniques, remedial exercise, and recommended home care.

Results: In this patient with erythromelalgia, effleurage and petrissage as massage therapy techniques provided temporary pain relief in the lower extremities and long-term benefits that relieved anxiety, which improved restorative sleep and increased the patient's participation in activities of daily living.

Conclusions: For this treatment protocol, therapist observation and patient feedback suggest that massage therapy may lead to a state of increased relaxation, decreased stress, decreased muscle tension, and improved sleep. These positive effects may have an indirect role in the ability of the patient to cope with erythromelalgia day to day.

KEYWORDS: Erythromelalgia, massage therapy, pain management

INTRODUCTION

Erythromelalgia is characterized by temperaturedependent redness, pain, and warmth in one or more extremities. It may be a primary disease, or it may occur secondarily to an underlying illness⁽¹⁾. Primary erythromelalgia is caused by mutation of the voltagegated sodium channel α -subunit gene $SCN9A^{(2)}$. Primary erythromelalgia may be classified as either familial or sporadic, with the familial form inherited in an autosomal dominant manner. Secondary erythromelalgia can result from small-fiber peripheral neuropathy attributable to hypercholesterolemia, mushroom or mercury poisoning, or an autoimmune disorder⁽³⁾. Erythromelalgia interferes with daily living and ability to work, because diagnosed patients experience a high degree of pain. This interference can lead to increased isolation, causing depression and anxiety.

Because of the rarity of erythromelalgia, very little is known about how to effectively treat it⁽⁴⁾. Current treatment does not address the cause of the condition, but involves high-dose oral medications to control pain and symptoms related to pain⁽⁵⁾. Other options for partial relief from the symptoms of erythromelalgia include direct cooling and elevation of the affected limb or limbs⁽⁶⁾.

Chronic stress can exacerbate the pathological consequences of erythromelalgia, resulting in physiological or psychological dysfunction. Therapeutic techniques that reduce the consequences of stress—for example, massage therapy (MT)—are therefore beneficial tools for improving overall health⁽⁷⁾. Sleep disturbances are a major factor in many chronic pain syndromes, such as that with erythromelalgia. Therapeutic massage may support restorative sleep so that an optimal environment for healing and restoration may occur in the body. We believe this is the first case report about the use of MT as a treatment method for erythromelalgia.

Here, we present the case of a 31-year-old woman with erythromelalgia who was prescribed MT by her physician to manage her pain symptoms. MT treatments for this patient focused on reducing the severity of symptoms and side effects, on improving restorative sleep patterns, and on reducing stress and anxiety. It was hoped that, with MT, the patient would be able to resume activities of daily living. It was also hoped that, with the use of educational materials, she might be able to prevent and relieve subsequent symptomatic episodes.

METHODS

Client Profile

The patient in this case was diagnosed in 2005 with idiopathic-onset erythromelalgia. Her medical history included fibromyalgia, Raynaud phenomenon, and Henoch–Schönlein purpura. A family history showed symptoms associated with erythromelalgia, such as occasional redness and heat in the distal extremities. This patient had participated in a variety of indoor and outdoor physical activities before onset of her symptoms. She had held a full-time job, but at the time of treatment, was receiving disability benefits because the pain limited her ability to work. The patient had never previously experienced massage or any other complementary therapy.

Assessment

The initial MT assessment showed limited active and passive range of motion of the extremities because of hypertonicity in the muscles. The patient monitored her pain and discomfort using a pain scale of 0 - 10 (0 = no pain, $10 = \text{extreme pain})^{(\hat{8})}$. On initial assessment, the patient reported pain in the distal extremities as 8 bilaterally. She indicated experiencing more pain in the evening, peaking at night before bed, and she described experiencing painful symptoms of erythromelalgia approximately 3-4 days each week, with episodes lasting between 24 and 72 hours. She described the characteristics of the pain in the lower extremities as a cramping pressure, and she provided a photograph of the distal extremities showing redness and swelling as a result of erythromelalgia (Figure 1). Aggravating factors for the patient included extreme



FIGURE 1. Redness and swelling in the lower extremities of our patient with erythromelalgia.

heat, extreme cold, exercise, or standing for longer than 20 minutes, because gravity perpetuated swelling in the lower limbs. Elevation of the lower extremities would relieve swelling, but fail to alleviate pain. Signs and symptoms associated with the onset of erythromelalgia episodes included tension headaches, irregular periods, a decrease in restorative sleep because of pain, and irritable bowel syndrome, which the patient attributed to her medications. Daily medications prescribed by her primary care physician included oxycodone with acetaminophen as needed for management of the pain of fibromyalgia and erythromelalgia, and lorazepam and amitriptyline for depression and anxiety. The patient also took raberprazole, a proton-pump inhibitor, and senna glycosides, a mild laxative, to assist with bowel movements.

Contraindications to MT for a patient with erythromelalgia would include any technique or modality that would aggravate the peripheral arteries (and therefore exacerbate symptoms). These contraindications include local techniques during extreme symptoms of erythromelalgia, broad stroking techniques to the lower extremities at any time, and heat applications to the lower extremities. Additional contraindications to MT are fever or signs and symptoms of a kidney infection (secondary to the Henoch-Schönlein purpura).

Having given informed consent to MT treatment, the patient took the positive outlook that MT would be beneficial for her conditions of stress and anxiety and might improve the erythromelalgia symptoms.

Treatment Overview

The patient received 9 treatments 7 - 14 days apart, each treatment lasting for 1 hour. The long-term goals of MT were to reduce the severity of erythromelalgia symptoms in the lower extremities, reduce pain, increase relaxation, improve circulatory flow, improve restorative sleep, and enhance quality of life.

Treatments began with static contact and superficial effleurage. Static contact is the least mechanically stimulating of massage techniques; it is used to increase sedation and decrease anxiety⁽⁹⁾. Superficial effleurage is a light, flowing technique that can be used to facilitate the flow of interventions and to achieve circulatory effects, psychological effects, and a variety of physiological effects.

For subsequent treatments, broad-contact compression was introduced. Broad-contact compression is a non-gliding technique that is delivered with a broadcontact surface. It is a useful introductory stroke that can positively affect circulation⁽⁹⁾.

Passive range of motion and passive stretch, assisted by the massage therapist, were also introduced to all of the extremities. The patient was encouraged to continue passive stretching at home in an effort to improve flexibility and range of motion.

To reduce swelling and pain, superficial lymph drainage was introduced. The proposed mechanism

for superficial lymph drainage is slow, delicate, rhythmical stretching of the tissues, which stimulates contraction of the lymphatic vessels, propels lymph through the collapsible superficial lymphatic system, increases local blood flow, and reduces the time required for alternative pathways (that is, anastomoses) to form after lymphatic pathways have been interrupted by damage. When this technique is skillfully performed, it can result in sedation (parasympathetic activity), reduced pain (counterirritant analgesia), and improved general immune function⁽⁹⁾. The patient was also encouraged to keep her legs elevated when possible at home so as to reduce swelling.

Each technique was introduced on a separate treatment day for accurate feedback of its effect.

RESULTS

Over the course of 9 treatments, pain and discomfort were assessed using the 0 - 10 pain scale before and after treatment (Figure 2). For the purposes of this case report, activities of daily living, anxiety, and sleep (Table 1) were recorded as either improved or unchanged as a result of the MT treatments. Activities of daily living were measured by patient reports of her willingness to participate in certain tasks during her day, both inside and outside the home, and for how long she was able to tolerate the activity before debilitating symptoms occurred. Anxiety was measured by patient feedback about how she was coping with the daily change of symptoms. Sleep was measured by the number of hours the client slept each night, the quality of the sleep, and whether painful symptoms were present.

After the first week of massage treatment, the patient chose to reduce her prescribed pain medication.



FIGURE 2. Changes in pain level. This chart illustrates the pain scale (0 - 10 on the vertical axis): 0 = no physical pain; 10 = extreme physical pain. The patient was asked to use the pain scale to rate her pain and discomfort before and after treatment (1 - 9 on the horizontal axis).

TABLE 1. Pattern of Patient-Reported Improvements in Activities of Daily Living, Anxiety, and Sleep Across 9 Sessions of Massage Therapy^a

	1	2	3	4	5	6	7	8	9
Activities of daily living Anxiety Sleep	+ + +	+	+	+			+ +	+ + +	+ + +

^a Plus signs indicate improvement, and blank cells indicate no change; the patient reported no decrements.

The patient rated her physical pain and discomforts at 5 after the initial MT treatment, and averaged 6 on the pain scale over the course of 9 treatments (Figure 2). After the first treatment, the patient reported a decrease in her anxiety. The patient's anxiety was relieved after 6 of 9 treatments. The patient noticed an improvement in the quality of her sleep, and she slept for longer periods throughout the night after 5 of the 9 treatments. After 3 of the 9 treatments, the patient reported a willingness to participate in more activities of daily living (Table 1).

Overall, the results of the treatment plan show that the patient responded best to relaxation techniques (such as superficial effleurage) to the upper body and upper extremities after approximately 3 treatments. Superficial lymph drainage seemed to exacerbate the pain symptoms. The patient states that, since the start of the massage treatments, episodes of pain occur, but they have declined in frequency, duration, and intensity.

DISCUSSION

The goal in this case was to introduce MT techniques as a method of pain management for symptoms of erythromelalgia. For many patients with erythromelalgia, multiple pain medications are ineffective in achieving complete relief of pain symptoms⁽¹⁰⁾. Physicians commonly treat chronic pain syndromes with medications for symptomatic relief. Patents with chronic pain may seek alternative therapies to reduce pain and to increase the ability to carry out everyday activities. A few studies have assessed nonpharmacologic treatments that are used and preferred by patients with pain, concluding that massage and heat were the non-pharmacological treatments that resulted in the best pain alleviation⁽¹¹⁾. Compared with other healing modalities such as prescription drug programs, MT also boasts fewer, if any, negative side effects.

A review of the literature showed no other studies of MT treatment for erythromelalgia. However, in reports on chronic disease, rhythmical MT showed long-term reduction of chronic pain symptoms and an improvement in quality of life⁽¹²⁾. Our results reveal that relaxation massage techniques, such as superficial effleurage to the upper body and extremities, provided restorative sleep and helped to reduce anxiety for the patient. The patient was able to relax, to achieve more restorative sleep, and to enjoy a longer period of time between severe symptoms. She reported that massage helped to provide relief between severe episodes of erythromelalgia and that it improved her quality of life. Massage techniques applied directly to the lower extremities caused anxiety for the patient. Superficial lymphatic techniques seemed to provide temporary relief for the patient during treatment, but lasted for only a short time once she was standing.

Further studies about the use of MT in the treatment of erythromelalgia are required. Treatments provided more frequently may amplify the positive results observed in the present study. Patients must be individually treated because each patient experiences various degrees of discomfort caused by erythromelalgia. We feel that the introduction of superficial lymph techniques earlier in the treatment sequence should further reduce the painful symptoms of erythromelalgia. In patients that are willing to attend treatments on a more regular schedule, evidence might be obtained of a beneficial effect of MT on their condition during acute symptomatic episodes. Despite having additional health complications such as fibromyalgia, Raynaud phenomenon, and Henoch-Schönlein purpura, our patient noted a positive influence of MT on her pain management.

Because this report concerns just a single patient, it is important to recognize that the approach described is not a generalized treatment for all erythromelalgia cases. Special considerations in this particular treatment plan dealt with the effects that MT could have on symptoms of erythromelalgia, while taking account of how MT techniques might affect the other conditions present (Raynaud phenomenon, fibromyalgia). Raynaud phenomenon has a presentation similar to that of erythromelalgia: both deal with radical responses in the extremities and peripheral arteries to a change in temperature. The patient in this case report had been diagnosed with secondary Raynaud phenomenon, which occurs as a complication from an underlying problem. The clinical manifestation of Raynaud phenomenon may be the final expression of abnormal reactivity of the terminal arteries^(13,14). During local or whole-body cooling, blood flow to the skin is reduced to prevent heat loss. This response to cold is mediated by reflex activation of the sympathetic nervous system and by a direct effect of cold on cutaneous blood vessels⁽¹⁵⁾.

The patient expressed anxiety about techniques that might aggravate her symptoms. Often, this anxiety would prevent the therapist from working locally on the lower extremities. A generalized MT treatment plan for erythromelalgia is recommended to include relaxation techniques (static contact, gentle effleurage, and petrissage) applied to the whole body, especially the lower extremities. Lymphatic massage is also an important technique to use, with absolute knowledge of its execution so that it can be used at every visit depending on patient comfort and compliance.

We have several other recommendations for MT treatment plans designed to manage pain symptoms. It is important that the patient and the massage therapist have a trusting and professional relationship. The patient must understand that, based on this treatment protocol, he or she may experience side effects related to treatment techniques before experiencing benefits. Positive effects from each massage treatment may be temporary, but can have a lasting effect over several treatments. Therefore, patient compliance is important, and MT treatments must be regularly scheduled at the discretion of the therapist. For the patient to achieve long-term benefit, the patient and therapist must both be committed to regular treatments. A general recommendation is 2 treatment sessions per week, for approximately 3 weeks, with 48 hours between treatments. Patient feedback is encouraged before, during, and after treatment, so as to determine which techniques are helpful and which techniques seem to have no benefit for the patient. In addition, home care is strongly encouraged so that the patient can experience the benefits of treatment for a longer period of time. The introduction of one technique at a time into each treatment is important for recognizing the techniques that are helping the condition and those that may be making the condition worse.

Future studies should include a larger sample size. For more accurate assessments, future studies should use multiple pain assessments. Examples include selfreports, behavioral assessments, and physiological measures such as the McGill Pain Questionnaire, a visual analog scale (VAS), or the Multidimensional Pain Inventory. If depression or anxiety is present, the Beck Depression Inventory and the Spielberger State–Trait Anxiety Inventory may be used. Use of these assessments will more accurately represent the results of the treatment plan, helping the therapist and the patient to monitor progress.

Based on the treatment protocol presented here, MT may provide an increased state of relaxation, decreased anxiety, decreased muscle tension, and improved sleep in a patient with erythromelalgia. These indirect positive effects may have a role in the patient's ability to cope day to day with symptoms. This treatment plan and the associated guidelines can contribute to further studies of erythromelalgia patients seeking alternative therapies.

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CONFLICT OF INTEREST NOTIFICATION

The authors declare that there are no personal or financial conflicts of interest to report concerning this case study.

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Corresponding author: Krista Dicks, Barrie, ON Canada.

E-mail: krista.dicks@hotmail.com