

Graduate Medical Education Research Journal

Volume 5 | Issue 1

Article 7

June 2023

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Recommended Citation

Naber, W. J., Scribner, D. J., , Howard, G. Stellate Ganglion Block for refractory Raynaud's Phenomenon-A Case Report. Graduate Medical Education Research Journal. 2023 Jun 30; 5(1). https://digitalcommons.unmc.edu/gmerj/vol5/iss1/7

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Abstract

Introduction: Raynaud's phenomenon (RP) is predominately a sympathetically mediated ischemic vasospastic disease involving the distal limbs and is associated with sharp pains, numbness, and dermatological color changes. The symptoms are commonly induced by cold weather, stress, and trauma and it is known to affect 4.85% of the population. There are few well-defined treatments for patients with RP that are refractory to 1st-line therapies. We present a case of intractable RP with corresponding response to stellate ganglion block (SGB) which demonstrates the efficacy profile and safety of SNS blocks in treatment of resistant RP.

Methods: Per Bon Secours Mercy Health IRB policy, this case report does not qualify for Mercy Health North IRB, and a formal statement from them has been attainted.

Results: By week 1, our patient had a reduction in pain (71.4%), duration of symptoms (75%), and frequency (75%), with a simultaneous 66% improvement in severity/dexterity. Our patient showed therapeutic improvement from baseline in duration/frequency until week 6 and pain/severity until week 7.

Conclusion: With minimal significant prospective data on comparative efficacy of various RP intervention options, and a high disease burden in the general population, we highlight the importance of this paper's findings and the need for further research on this topic.

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Stellate Ganglion Block for Refractory Raynaud's Phenomenon- A Case Report

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https://doi.org/10.32873/unmc.dc.gmerj.5.1.007

Abstract

Introduction: Raynaud's phenomenon (RP) is predominately a sympathetically mediated ischemic vasospastic disease involving the distal limbs and is associated with sharp pains, numbness, and dermatological color changes. The symptoms are commonly induced by cold weather, stress, and trauma and it is known to affect 4.85% of the population. There are few well-defined treatments for patients with RP that are refractory to 1st-line therapies. We present a case of intractable RP with corresponding response to stellate ganglion block (SGB) which demonstrates the efficacy profile and safety of SNS blocks in treatment of resistant RP.

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Introduction

Raynaud's phenomenon (RP) is predominantly a sympathetically mediated ischemic vasospastic disorder involving the distal limbs and is associated with sharp pains, numbness, and dermatological color changes. The symptoms are commonly induced by cold weather, stress, and trauma. Commonly documented secondary generators include connective tissue diseases, vascular hematologic disorders, trauma, and certain medications.1 Affected areas change color as the vasculature is affected by spasms (pale/ white), tissue becomes cyanotic (blue/purple),

and spasms relent allowing for reperfusion (red/pink).¹ This cycle of vasoconstriction and vasorelaxation with corresponding ischemia and reperfusion is multifactorial, but the sympathetic nervous system (SNS) is largely implicated in RP related pain because of its role in the creation of the spasm and afferent ischemic pain transmission via the same SNS pathways.^{2,3} For this reason, SNS blocks in the corresponding ganglia have been used to minimize vasospasm generation and afferent nociceptive response in refractory cases of RP. Despite a relatively high prevalence in the general population (4.85%),⁴ there are few well-defined treatments for those with RP that are refractory to 1st-line therapies. To expand evidence on available interventions, we present a case of intractable RP with corresponding response to stellate ganglion block (SGB) which demonstrates the efficacy profile and safety of SNS blocks in treatment resistant RP.

Case

A 51-year-old female presented to the clinic with a several year history of bilateral paroxysmal hand/wrist pain, with the right worse than the left. Symptoms were most affected by extreme temperatures, greatest on the palmar/dorsal aspects of the right thumb and index fingers and caused color changes with associated pain radiating diffusely through the entire hand, particularly distal phalanges. She also reported associated bilateral numbness/tingling in all fingers. Previous electromyography and nerve conduction studies (EMG/NCS) studies were insignificant. Her past history was significant for fibromyalgia, Chiari malformation decompression/craniectomy and undifferentiated polyarthropathy being treated with Plaquenil. She had no history of tobacco/ smoking, diabetes, thyroid dysfunction, vascular disease, focal arm weakness, other paresthesias, balance/gait disturbances, and bowel/bladder incontinence. , Her physical exam was benign except for moderately reduced cervical range of motion (ROM) and diminished light touch sensation in her thumb and small fingers on the right. There were no skin lesions, rashes, or ulceration visible in her head, neck, or upper limbs.

Initial treatment modalities with low-dose steroids, non-steroidal anti-inflammatory drugs (NSAID), neuropathic agents, muscle relaxers, and occupational therapy desensitization techniques provided no relief by the 8th week of treatment. She continued to have right hand/wrist pain, numbness, and tingling, particularly in her thumb and index fingers in low temperature environments. As a result, an EMG/NCS and skin biopsy were performed to investigate small fiber neuropathy which vielded unremarkable findings. Although likely a multifactorial clinical presentation, the majority of her pain corresponded with Raynaud phenomenon symptomatology, and she denied interest in considering more oral agents. In response, a right stellate ganglion block (SGB) was performed to address the sympathetically mediated Raynaud reperfusion pain.

With informed consent obtained, an intravenous line was placed in the left hand. The patient was positioned supine on the fluoroscopic table. C6 vertebral body was identified under an oblique fluoroscopic view. A needle was inserted under fluoroscopic view until reaching Chassiaignac's tubercle. Injection of 1 cc of Omnipause showed an optimal spread of contrast along the longissimus colli. Next, 10cc of a mixture (Bupivicane, 1dexamethasone, Omnipaque) was injected in a graduated manner demonstrating good flow of contrast media to the T2 vertebrae (Figure 1A & 1B).

Within minutes of procedure completion, the patient demonstrated an effective SGB as shown by her right sided Horner's components (ptosis, facial anhidrosis, miosis) and warm erythematous right hand when compared to the left. The patient also reported significant pain relief in her right hand within minutes of SGB performance.

Discussion

Typical therapeutic modalities for RP include removal of underlying cause, avoidance of triggers (cold, stress, trauma, smoking), and warming of digits. When the aforementioned fail, oral agents, such as calcium channel blockers and phosphodiesterase inhibitors have shown minimal efficacy.⁵ Our patient denied interest in oral agents, and had minimal relief from other therapeutic modalities, so we elected to proceed with a SGB block targeting the T2 ganglion due to its sympathetic involvement on the upper extremity vasculature.⁶

In consideration of our patient's profession, which requires significant hand function and dexterity, we utilized the 4 data points in Table 1 to evaluate how much RP impacted her daily professional and non-professional life. Compared to Week 0's baseline, she had a reduction in pain (71.4%), duration of symptoms (75%), and frequency (75%) combined with a a 66% improvement in severity/dexterity by the end of Week 1 (Table 1). Although therapeutic benefit gradually trended back towards baseline after Week 1, our patient showed improvement in duration/ frequency until Week 6 and pain/severity until Week 7. During the initial post-injection follow up (Week 3), she reported increased post-injection numbness in the palmer/ thumb aspect of her right hand, which could be attributed to secondary/underlying pain generators, including her C5/6 radiculopathy. Additionally, this injection was performed during the spring, which had labile temperature fluctuations which historically cause her the most pain. Compared to prior spring symptoms, she reports a "baseline" improvement with the SGB beyond Week's 6/7, further demonstrating SGB efficacy despite confounding weather variables.

Further investigation with this case would be the examination of utilizing repeat injections to create summative improvement over time, as demonstrated in published literature.⁷ Ketamine, and other N-methyl-D-aspartate (NMDA) receptor antagonists, are believed to be able to effectively block the peripherally located NMDA receptors and better counteract the "wind-up' phenomena of chronic pain."8 As a result, SGB with adjuvant ketamine has also been shown to be a safe and effective technique in patients without destructive lesions, similar to other interventional modalities such as radiofrequency ablation of the ganglion.8 Although our patient did receive short therapeutic benefit from SGB, the current literature suggests that this is not predictive of the success of other interventions such as spinal cord stimulation, botulinum toxin injection, peripheral surgical sympathectomies or other complementary therapies.9 Importantly, the absence of predictive value from SGB should not discourage further interventional treatment as desired by the patient. Further interventional treatments should be thoroughly discussed and approached in a stepwise manner to maximize efficacy and limit adverse events. Botulinum toxin injections are considered very safe but will not have the same duration of effect as surgical sympathectomy which, in contrast, has a much higher incidence of undesired effects.9 Because RS related pain can be highly variable in responsiveness to

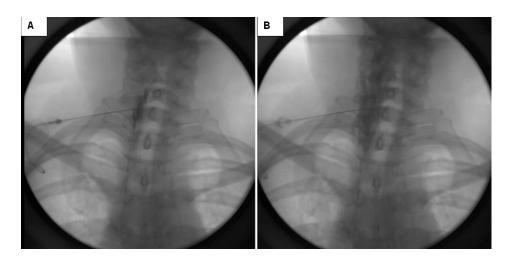


Figure 1: The flow of contrast media to the T2 vertebrae

 Table 1: Four data points to evaluate the impact of Raynaud's phenomenon in the patient's daily professional and non-professional life

Week Number	Pain (0-10)	Severity (0-4)	Duration (0-4)	Frequency
(Prior to SGB) Week 0	7	3	2	4
(Following SGB) Week 1	2	1	0.5	1
Week 2	3	1	1	2
Week 3	4	2	1	2
Week 4	5	2	1.5	3
Week 5	5	2	1.5	3
Week 6	6	2	2	4
Week 7-12	7	3	2	4

Ratings were based on the following.

Pain: Based off Visual Analogue Scale (VAS) for pain scaling

Severity: 0-4: based off hand dexterity, (0)-No reduction in hand function during Raynaud spasm(1)- mild (<25% reduction in hand function), (2)- moderate (>25% but <50% reduction in hand function), (3)- severe (>50% but <75% reduction function), (4)- incapacitating (>75% maximal intensity)

Duration: Total cumulative hours per day with Raynaud' symptoms on average

Frequency: Number of times per day that abortive hand warming was utilized

therapy, a personalized approach to treatment options is most likely to be successful. Lastly, with minimal significant prospective data on comparative efficacy of various intervention options,⁹ it highlights the importance of this paper's findings and the need for further research on this topic.

Conclusion

Chronic RP remains a challenging syndrome to treat and also a poorly studied disease state with ample room for continued research. Despite a relatively high disease burden in the general population, the available medical and interventional therapies are of limited efficacy for a significant group of refractory patients. Likewise, our case does show that SGB can provide effective relief for patients with refractory RP, albeit with unsatisfactory duration of relief. Our continued treatment plan will be focused on extending the duration of the relief that was provided by the stellate blockade to better improve her quality of life and daily functional abilities. Further research will be needed to elucidate more durable variations of current treatments or via creation of entirely novel therapies.

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