

The Efficacy of Spinal Ganglion Blockade in a Patient with Peripheral Circulation Dysfunction: A Case of Scleroderma

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Abstract: Scleroderma is a connective tissue disease characterized by the deposition of collagen in the skin, internal organs, and arteries and also by microvascular obstruction. Sympathetic ganglion blockade (cervicothoracic and lumbar sympathetic) is one treatment modality used in this population for circulation failure of the lower limbs, severe pain, gangrene, intermittent claudication, hyperhidrosis, sympathetic hyperactivity, and problematic peripheral circulation. In this case report, we aimed to discuss the efficiency of spinal ganglion blockade in a 30-year-old female patient with digital ulceration, pain, and circulation disorder caused by scleroderma.

Keywords: Ganglion blockade, Circulation dysfunction, Scleroderma

INTRODUCTION

Scleroderma is a connective tissue disease characterized by the deposition of collagen in the skin, internal organs, and arteries and also by microvascular obstruction. The etiology of scleroderma remains unclear [1]. There are two types of scleroderma: systemic and localized forms. Systemic scleroderma affects not only the skin but also the blood vessels or internal organs. The two main varieties of localized scleroderma are morphea and linear forms, which impact only the skin; rarely, tissues under the skin are also involved [2].

The prevalence of scleroderma is estimated to be 30 per 100,000. Scleroderma occurs four times more often in women than it does in men. Age varies depending on the type of scleroderma. The condition occurs more often between the third and fifth decades, although the linear form occurs under 18 years [3, 4]. Familial scleroderma is rarely reported [5].

Sympathetic ganglion blockade (cervicothoracic and lumbar sympathetic) is one treatment modality used in this population for both diagnostic and therapeutic purposes. This method is a component of treatment in herpes zoster, post-herpetic neuralgia, post-traumatic reflex sympathetic dystrophy, vasospastic diseases and circulation failure of the lower limbs, atherosclerotic disorder, severe pain, gangrene, intermittent claudication, hyperhidrosis, sympathetic hyperactivity, and problematic peripheral circulation [6].

In this case report, we aimed to discuss the efficiency of spinal ganglion blockade in a 30-year-old female patient with digital ulceration, pain, and circulation disorder caused by scleroderma.

CASE REPORT

A 30-year-old woman suffering from numbness in both of forearms, cyanosis from wrist to fingers for 17 years. She had a 12-year history of ulcerations on all fingers of both hands and all toes on both feet. Until 20 years old, she had received symptomatically treatment soon after she was diagnosed as scleroderma. She was treated with trental, acetyl salicylic acid, diltiazem, colchicine, and angiotensin converting enzyme inhibitors by the rheumatology and dermatology departments. The patient was admitted to our clinic due to worsening pain and cyanosis of the fingertips.

Physical examination upon admission to our pain clinic revealed hyperemia and infected wounds on the second and third fingers of the right hand, thickening of the ear and forehead skin, tapered nose cartilage, limitation of flexion in both forearms and legs, and thickened lips. The distal portion of the left and right fingers had become slimmer and been autoamputated. The distal portion of the left third finger contained a gangrenous wound, and the right second toe of the foot had eroded along with the heel. The patient suffered from hepatomegaly of 2 cm under the subcostal area. A complete blood count revealed hemoglobin: 10.7 g/dl, white blood cell count: 15.700/mm³, and platelet count: 380.000 mm³. Serum analysis showed ANA: 1/320 (discrete speckled), ENA: 110, ENA SCL: 70 (+), CRP: 1.01, C3: 275, C4: 44.5,

and RF: 22. Further analysis showed negative anti-centromere antibodies, anti-double strand DNA antibodies, anti-kardiolipin M antibodies, anti-kardiolipin G antibodies, anti-phospholipid M antibodies, and anti-phospholipid G antibodies.

We decided to recommend a spinal ganglion blockade and obtained informed consent from the patient. After the patient was transferred to the operating room, standard anesthesia monitoring was performed (electrocardiography, pulse oximetry, non-invasive blood pressure monitoring). Vascular access was established with an 18-G cannula in the antecubital vein. After placing the patient in the prone position, a pillow was placed under the abdominal region to correct lumbar lordosis. Following appropriate area cleaning, the patient was sedated, and right spinal ganglions were reached via a needle inserted at the L3 level. A lumbar sympathetic blockade was performed using 10 ml of 5% marcain. Following this diagnostic lumbar sympathetic blockade, the patient was taken to the recovery room, and the temperature in both feet was monitored. A three-degree increase was recorded in the right foot.

After this successful procedure, the patient underwent a second one. Following a physical examination, the patient was taken to the operating room, and lumbar sympathetic ganglions were reached using a 100-mm radiofrequency (RF) needle at the L3 level. A gradually progressive course of impulses was given, and the patient felt pain at 0.7 mV. After determining the placement of the sensorial ganglion, the impulse was increased gradually. Although a 2-mV level was reached, a pain response was not achieved. Therefore, a permanent sympathetic blockade was made with a conventional RF for 2 minutes at 80 degrees. The follow-up visit was done at 2 weeks postoperatively, and the temperatures in both feet were higher. Cyanosis and desquamation at the distal parts of the fingers had disappeared, and the ulcers in the toes as well as the patient's general pain had improved significantly. Six months postoperatively, the ulcers had resolved completely, and warming of the extremities continued.

DISCUSSION

Scleroderma is a connective tissue disorder characterized by the deposition of the extracellular matrix and fibrosis of the skin along with internal organ involvement. Systemic sclerosis is typically associated with Raynaud's Syndrome. Extreme sympathetic activity may lead to Raynaud's Syndrome. Hyperactivation of the sympathetic nervous system produces vasoconstriction, a decrease in the capillary surface area, an increase in vascular permeability, and impaired fluid mobilization. Treatment options include anti-depressants, calcium channel blockers, angiotension-converting enzyme (ACE) inhibitors, acetyl salicylic acid, corticosteroids, prostacyclin

analogues, N-acetylcysteine (NAC) infusion, and preventing exposure to cold temperatures. Sympatholysis or sympathectomy is a new treatment options used to resolve vasospasm and facilitate circulation in refractory cases [7-9].

Our patient had been diagnosed with scleroderma 17 years previously and was followed by the rheumatology and dermatology departments. The patient suffered from chronic pain and skin lesions related with ischemic wounds (autoamputations of the fingers and cyanosis and desquamation of the toes were detected) at admission. We recommended lumbar sympathetic ganglion blockade for her symptoms refractory to other treatment options. The positive outcomes of the procedure were a short duration of immobilization, increased temperature in both extremities, and improvement of her pain, desquamation, and wounds. In addition to these outcomes, psychiatric symptoms of the patient was ameliorated.

Improved circulation due to developed arterial collateral secondary to sympathectomy relieves skin lesions and symptoms related with Raynaud's syndrome. However, surgical sympathectomy seems a limited treatment option. In addition, patients with complex regional pain syndrome who have recurrent ulcers and delayed wound healing have limited surgical treatment options. Peripheral nerve blockade or sympathetic ganglion blockade is a less invasive treatment option that can be used to prevent sympathetic activation related with Raynaud's Syndrome [10, 11]. Another treatment modality includes continuous epidural injection of anesthetics for sympathetic ganglion blockade. In this method, patients receive continuous lumbar sympathetic ganglion blockade via a catheter inserted in the paravertebral region. Doppler ultrasonography is used both before and after the blockade to identify enlargement of the arteries and increases in blood circulation. Acute wound healing can be achieved using this procedure; however, the lifespan of catheters limit this method [10, 11]. Intravenous sympathetic regional blockade with guanethidine, bretylium, or reserpine is another treatment approach. In addition to the severe hypotensive effect of guanethidine, adverse systemic and gastrointestinal events may be seen with this method. Guanethidine is contraindicated in patients with suspected pheochromocytoma and patients with known monoamine oxidase inhibitors usage. Guanethidine must therefore be used with caution in patients with coronary artery diseases.

CONCLUSION

In conclusion, lumbar sympathetic ganglion blockade is an efficient and less invasive method with acceptable patient satisfaction that can be used in

patients with refractory Raynaud's disease in scleroderma.

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