

Introduction

This case report describes a series of stellate ganglion blocks in a patient with right upper extremity, right chest and right flank pain in the setting of Complex Regional Pain Syndrome Type I and autosomal dominant centronuclear myopathy.

Case Description

- A 22 year old woman with a medical history of autosomal dominant centronuclear myopathy, chronic respiratory failure requiring continuous nasal noninvasive ventilatory support, chronic lumbar pain secondary to scoliosis, wheelchair dependence and Complex Regional Pain Syndrome (CPRS) Type I presented for management of right upper extremity, right chest and right flank pain.
- The discomfort had begun 7 months prior as severe right lower rib and intercostal pain, radiating to the entire back, flank and right hip. There was no obvious precipitating injury.
- She had been treated inpatient with steroids, hydromorphone, ketamine, pregabalin, a transverse abdominal block and several intercostal blocks. The first inpatient TAP block completely relieved her lower front rib cage pain; subsequent blocks yielded no improvement.
- After discharge, her pain was controlled with buprenorphine, pregabalin, gabapentin, celecoxib, duloxetine and osteopathic manipulative treatment. Over the next few months, the patient’s right sided flank pain transformed into 7/10 intensity, sharp, shooting, electric-like constant discomfort and expanded in range to include the right upper extremity and right patella.
- Deep breathing and physical touch exacerbated the pain. In addition, the patient’s right hand 5th digit fingernail sloughed off without any overt trauma.
- Rib xrays, thoracic MRI and CT of abdomen, chest and pelvis were all negative for injury. The patient had undergone spinal fusion in 2015, but there was no hardware complication on imaging.

Case Description (Cont'd)

- EGD revealed only gastritis. Patient’s PEG tube functioned well with clean, dry insertion site.
- On physical exam, the patient exhibited exquisite tenderness in the right upper extremity and increased temperature in the right hand.
- The presentation was highly suspicious for atypical CPRS, and a diagnostic ultrasound guided **stellate ganglion block (SGB)** was attempted. However, the anatomical landmarks could not be visualized under ultrasound.
- The patient underwent a series of three SGBs under fluoroscopy.
- For the first block, a 25 gauge, 1.5 inch Quincke needle was used to inject 3 ml of 0.25% bupivacaine into the C7 pedicle target zone. The patient reported 80% pain reduction in her right flank, right rib cage and right upper extremity for the first six days after the first SGB. On the seventh day, the patient aspirated her beverage and had a severe coughing spell. Anterior chest wall pain returned with allodynia, but flank, leg and arm pain all remained improved.
- The patient underwent her second stellate ganglion block of 3 ml of 0.25% bupivacaine four weeks after her first. There were no further sympathetic symptoms, and the patient reported 90% pain reduction, again in all three regions.
- The third and final SGB of the series, with 5ml of 0.25% bupivacaine, was performed 2 weeks after the second SGB. Nine days after the procedure, the patient reported 95% pain relief as before in right flank, right rib cage and right upper extremity.

Discussion

- Budapest Criteria for CPRS diagnosis include: (a) disproportionate pain; (b) at least one symptom in three of four categories including sensory, vasomotor, sudomotor/edema and motor/tropic; (c) at least one symptom in at least two of the four categories when evaluated and (d) no other explanation for the symptoms (1). Our patient’s multifocal pain, allodynia, temperature differential and sloughed fingernail together support a CPRS diagnosis. The absence of nerve damage makes it Type I.

Discussion (Cont'd)

- SGBs are widely used to diagnose and treat upper extremity CRPS (2). However, SGBs for chest and abdominal pain are uncommon. In one case report, a patient with chronic aching, stabbing left anterior chest pain after aortic valve root replacement complicated by pericarditis reported 30% to 40% pain reduction 8 months after SGB with 5 mL of 0.25% bupivacaine at C6 level (3).
- Centronuclear myopathy is identified histologically by clustered, centralized nuclei in skeletal muscle cells, and associated with an array of genes including DNM2 and B1N1 that function in cell membrane remodeling (4). Common features also exhibited by our patient include muscle atrophy, masticatory muscle weakness and paravertebral muscle weakness leading to scoliosis and respiratory insufficiency (5).
- Wheelchair dependence, muscle weakness and spinal deformity all restricted her movement and made her more prone to muscle injury due to prolonged static positioning.

Conclusion

The fact that stellate ganglion block provided immediate relief chest and flank pain, in addition to upper extremity, that is, in the case of the flank pain, durable to mild trauma, offers a promising new application of stellate ganglion blocks to abdominal CPRS Type I in patients with centronuclear myopathy.

References

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