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Acute Vision Loss Sequela of Complex Regional Pain Syndrome Type 1 Successfully Treated with Stellate Ganglion Block: Case Report

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Background: Vision changes are rarely reported as a complication of complex regional pain syndrome (CRPS). Previous research has shown that photophobia and changes in pupil size associated with CRPS are the result of autonomic disturbances. Stellate ganglion blocks relieve pain by altering, at least temporarily, the dysautonomia associated with CRPS.

Case Report: A 30-year-old white woman with a history of cervical disc herniation and progressive radiculopathy was diagnosed with CRPS type 1 after C6-7 arthroplasty. Her case was complicated by ipsilateral acute vision loss in the left eye and left facial pain, which improved after stellate ganglion block.

Conclusion: This case broadened our knowledge about CRPS and suggested that associated dysautonomia can adversely affect visual acuity.

Key words: Case report, complex regional pain syndrome, dysautonomia, stellate ganglion block, visual loss

BACKGROUND

Complex regional pain syndrome (CRPS) classically presents with significant pain and sensory changes that are out of proportion to the inciting trauma. The syndrome may exhibit abnormalities in motor, sensory, vasomotor, sudomotor, and autonomic systems in the affected region, most often in the distribution of multiple peripheral nerves (1). CRPS is commonly seen after traumas and surgeries, including spine surgery (2). The pathophysiology of CRPS is a complex combination of neurological sensitization, dysautonomia, and inflammation, as well as multifactorial interactions of immunological, genetic, and psychological influences (3). Emerging evidence suggests that overactivation of the sympathetic system is one of the driving forces causing pain (4,5), which might be the underlying reason that peripheral sympathetic blockade relieves pain (3,6).

Visual changes are not commonly reported by patients with CRPS. There are only a few studies that have reported visual discomfort and changes in pupil size and pupillary reflexes associated with CRPS, which are postulated to be the result of dysregulation of autonomic pathways (7,8). We report on a rare but very interesting case of CRPS complicated by acute vision loss that improved after stellate ganglion block (SGB). We hypothesize that SGB improved visual loss by correcting dysautonomia, and we further discuss the neurophysiologic mechanisms underlying the visual changes.

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CASE REPORT

A 30-year-old woman with history of cervical disc herniation and progressive radiculopathy and myelopathy status post cervical spine surgery presented with left upper extremity allodynia, skin discoloration, mild edema, and sudomotor changes. Her initial injury symptoms (onset October 2020) were neck pain and bilateral arm pain and weakness. She subsequently developed left arm radicular pain and weakness of her left arm and leg, with resolution of right-sided symptoms. Conservative treatments failed and cervical spine magnetic resonance imaging (MRI) (December 2020) revealed a left C6-7 protrusion with canal stenosis and moderate cord deformity, but no foraminal stenosis or cord signal. Her neuropathic symptoms progressed despite conservative treatment, which prompted surgical decompression and C6-7-disc arthroplasty (April 2021). The surgery did not improve her symptoms and her pain syndrome continued to worsen. Postoperative cervical spine MRI (June 2021) revealed no stenosis or cord signal.

Upon presentation to our clinic (August 2021), her physical examination showed allodynia, global left upper extremity weakness, and distal pallor and edema without trophic changes (Fig. 1A). She was diagnosed with CRPS type 1, prescribed multimodal analgesics, and occupational therapy. She was subsequently scheduled for a SGB. While awaiting the procedure, she developed acute onset of left facial pain and vision loss in the left eye with pupil size changes (about 80% of the time, L>R, Fig. 1B1; the other time, L<R, Fig. 1B2), which was not her baseline. Urgent imaging included computed tomography angiography of the head and neck and MRI of the brain and spine, all of which ruled out stroke, multiple sclerosis, and orbital pathology. Ophthalmology examination confirmed vision loss OS 20/300, anisocoria (L>R). However, there was no ocular pathology to explain her vision loss. On our neurological exam, there was relative afferent pupil dilation of the left eye, fluctuating pupil sizes with continuous light stimuli, and no Horner syndrome. Visual evoked potential showed an intact visual pathway. She underwent an ultrasound-guided left SGB (October 2021, Fig. 1C). She tolerated the procedure well without any adverse effects. Her vision loss and facial and left arm pain significantly improved post procedure. Repeat ophthalmology exam one week after the block showed improved vision acuity OS 20/70, equal pupil sizes, and no more fluctuations of the left pupil size with light reflex (Fig. 1D).

DISCUSSION

Our patient developed CRPS type 1 after surgery of the cervical spine, which is a common potential complication after surgeries and traumas (2). The mechanisms of CRPS include dysregulation of inflammation, autoimmune systems, neurotransmitter systems, and autonomic systems (3). Sympathetic blockage, such as SGB (6,9), alleviates pain because sympathetically-maintained pain is a common feature of CRPS (4,5).

Visual changes with CRPS are not commonly reported; however, some evidence has shown that photophobia is associated with CRPS (8) and pupillary reflex is affected by CRPS due to autonomic disturbances (7). Our patient developed left-sided vision loss and fluctuating anisocoria, i.e., most of the time, her left pupil is larger (Fig. 1B1); other times, her left pupil is smaller (Fig. 1B2). Her exam consistently found anisocoria L>R, and her left pupil size abnormally fluctuated with continuous light stimuli. There is no obvious ocular pathology to interpret her vision loss and anisocoria. It was unclear why she suddenly developed visual changes.

To review the anatomy, the lens shape and pupil size are controlled by autonomic systems of both sympathetic and parasympathetic nerves (n.), which together determine visual acuity. Parasympathetic fibers derived from the optic n. synapse at the ciliary body and postganglionic axons innervate the ciliary muscles and pupillary sphincters. Optic n. activation leads to miosis when focusing on a near object. In contrast, sympathetic postganglionic axons from the superior cervical ganglion form the sympathetic fibers that pass through the ciliary ganglion and innervate pupillary dilators, causing mydriasis when looking at a distant object. Based on the anatomy, our hypothesis is that, in the setting of CRPS, an overactive sympathetic tone is the driving force behind the dilation of the left pupil for most of the time (Fig. 1B1 and Fig. 2). It is likely that parasympathetic input is trying to correct this abnormality with an overshoot, which results in the constriction of the left pupil for the other time (Fig. 1B2 and Fig. 2). As a result, dysregulation of the lens shape and pupil size due to imbalance between sympathetic and parasympathetic systems might be the reason for vision loss and fluctuating anisocoria. This is consistent with the improvement of visual acuity after SGB (Fig. 2) due to ipsilateral sympathectomy. SGB selectively reduces sympathetic outflow of the ipsilateral upper extremity and head by anesthetizing the ascending sympathetic fibers from the superior cervical ganglion (6).

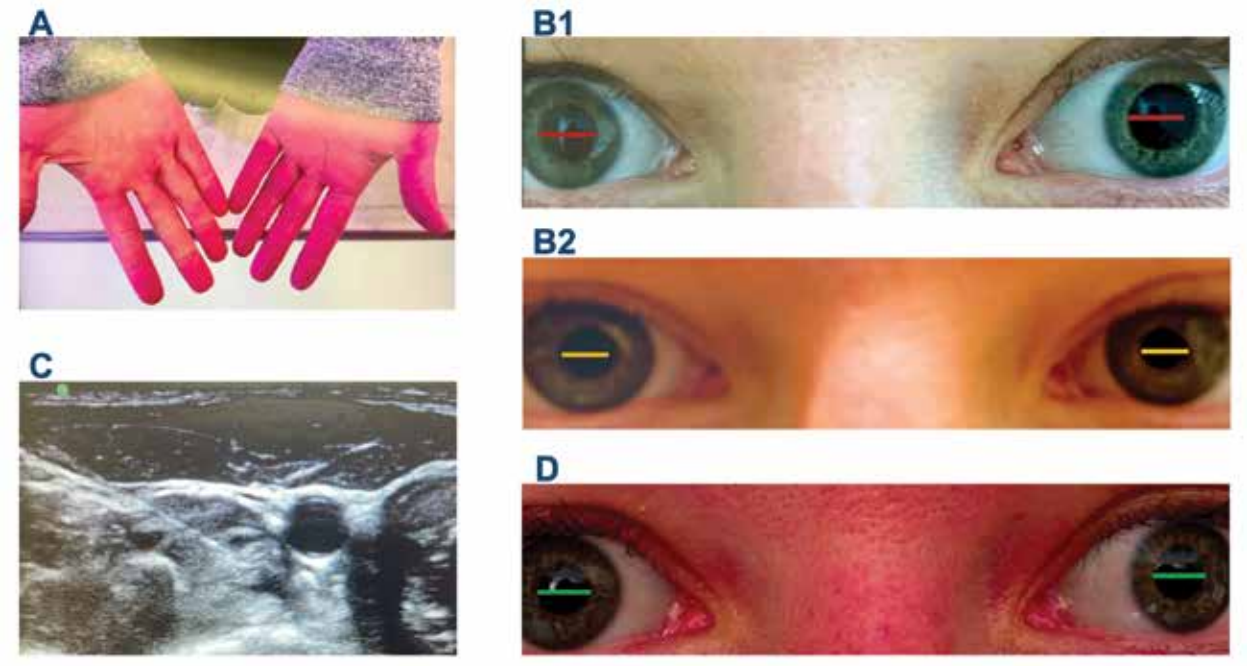

Fig. 1. A: Patient developed complex regional pain syndrome (CRPS) type 1 after surgical decompression and C6-7-disc arthroplasty. Note discoloration of left hand; B1/B2: Fluctuating anisocoria of left eye; C: Ultrasound-guided left stellate ganglion block (SGB); D: Vision and anisocoria significantly improved after SGB. Red, yellow, and green bars are the same length.

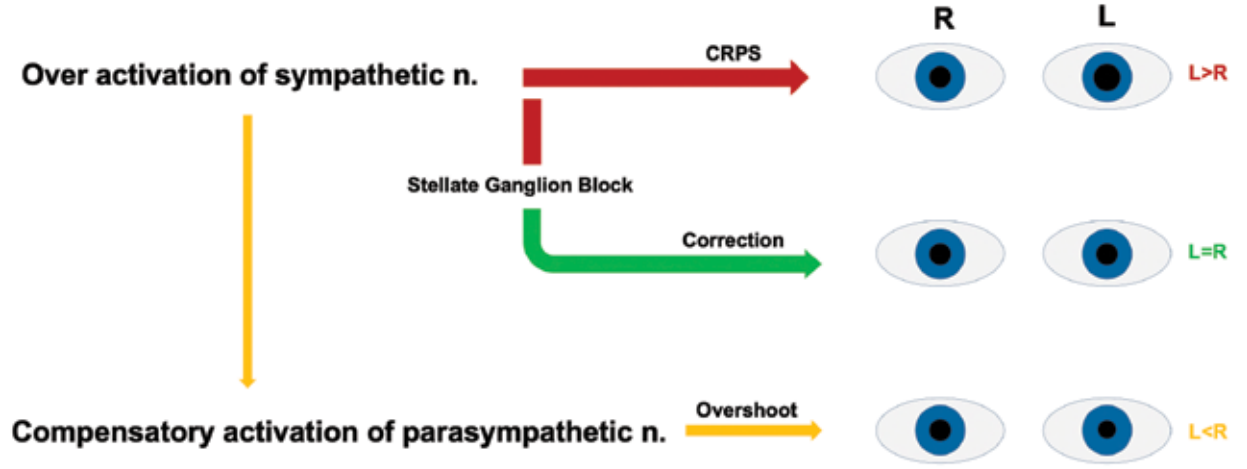

Fig. 2. Dysregulation of lens shape and pupil size due to imbalance between sympathetic and parasympathetic nerves in the setting of complex regional pain syndrome (CRPS) might be the reason for vision loss and fluctuating anisocoria. Stellate ganglion block (SGB) corrected the overactivation of the sympathetic system and improved vision loss and anisocoria.

CONCLUSIONS

Restated, given the vision recovery response after SGB, it is possible that the patient's vision loss and anisocoria are rare complications of CRPS secondary to dysautonomia. This phenomenon is further supported by clinical research on CRPS demonstrating that pupillary size and reflex are affected by dysregulated sympathetic pathways (7). This research found that ipsilateral to the side of CRPS, the pupil is smaller and pupillary dilation is delayed, which is slightly different from what we see

in our patient. The difference might be explained by the balance of the variable sympathetic and parasympathetic tone and fluctuating catecholamine levels at different phases of CRPS (3).

This case broadened our knowledge about CRPS and suggested that dysautonomia, especially overactivation of the sympathetic system, can cause vision changes that can be treated with SGB.

Disclosures and Conflicts of Interest

The patient has provided consent for this case to be published. We declare that there are no copyrighted figures. Both LL and MGH contributed to the writing of this manuscript, read, and approved the final manuscript. Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

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