Case Report

After 30 Physician Consults and Six Years: a Case Report Detailing a Successful Treatment of Posttraumatic Cervical Dystonia using Botox

William Ciurylo¹ and Terence K. Gray²

¹OMS II, University of New England College of Osteopathic Medicine, ME 04005, USA.
²Adjunct Professor of Clinical Research at University of New England College of Osteopathic Medicine and Medical Director of the Interventional Pain Center at Mercy Hospital, ME 04005, USA.

Background

Chronic pain is a challenging condition to diagnose and treat. It has been demonstrated that between 40 and 80% of chronic pain patients are misdiagnosed and, therefore, mistreated [1]. What this suggests is potential for mitigation of the emotional, physical, and financial tolls of chronic pain in many individuals should the right methodologies be employed. Handler posits that this prevalence of misdiagnosis is the result of inadequate history taking and the ordering of the wrong tests [1]. Other sources speak to a broader deficiency in the preparation of physicians to treat pain over the course of their education [2]. These deficiencies have led to low levels of physician satisfaction in their ability to treat those with chronic pain [3]. Overall, these findings indicate a need for case reports to educate physicians on how to appropriately recognize and manage the chronic pain conditions that they may encounter.

One such condition is chronic neck pain. It has been estimated that 13.7% of adults will have experienced some form of neck pain sometime within the past three months [3]. Additionally, it is believed that 15% of adults will experience neck pain that is considered chronic (lasting greater than three months) over the course of their lifetimes [4]. The prevalence of these conditions results in clinical burden, costs, and disability in the general population. The etiologies of neck pain and chronic neck pain vary but one of the most challenging to diagnose and treat is cervical dystonia.

Cervical dystonia is a chronic neurologic disorder that manifests as involuntary contractions of the cervical musculature resulting in painful postural changes of the head, neck, and/or shoulders [5,6]. As a condition, it has a heterogenous clinical presentation, variable clinical features, and, thus, various classifications that can be difficult to apply. In general, cervical dystonia is considered either primary or secondary distinguishing between idiopathic and exogenous origin respectively. Its prevalence varies widely by study methodology with one 2013 study finding it to range between 20 and 4,100 cases per million [7]. Part of the difficulty in determining its prevalence is the challenge of cervical dystonia being commonly misinterpreted or misdiagnosed [8-10]. Studies have suggested that one quarter to half of people with dystonia may be undiagnosed or misdiagnosed [11-14]. Furthermore, it has been documented that those with cervical dystonia often require multiple doctors’ visits and extended time before a diagnosis is reached [15,16].

Further compounding the difficulties incurred by cervical dystonia is the finding that roughly 12% of those afflicted had experienced a preceding trauma [17]. Investigators have claimed that post-traumatic cervical dystonia has clinical features that distinguish it from idiopathic cervical dystonia but the results of comparison studies have been inconclusive [6,17-19]. Furthermore, reluctance to make the diagnosis is reported by physicians. Post-traumatic cervical dystonia is difficult to accept because of the presence of recall bias, the fact that movement disorders usually occur in the absence of trauma, and that there is uncertainty over the length of time between injury and onset allowable for the diagnosis [20]. More so, there is a persisting perception that those who experienced an injury may have exacerbated symptoms due to pending litigation, creating the appearance of cervical dystonia rather than posttraumatic cervical dystonia being a distinct entity [17,20]. However, the classification of atypical posttraumatic cervical dystonia still endures to allow for the diagnosis of cervical dystonia of a believed traumatic etiology.

Texts that do report it as distinct point to unique clinical features brought on by the traumatic event. In a study of 95 patients with cervical dystonia, sixteen could identify a preceding injury. These sixteen patients were significantly more likely to report laterocolis, depression, and more pain than those who did not report a preceding trauma [20]. Other features of this type of dystonia include its impact on the side of the body opposite the side of the brain injured and paraspinal muscle hypertrophy [17,21]. Our aim is to present a case with these clinical features. It is one that we believe effectively illustrates an instance of posttraumatic cervical dystonia and the described diagnostic challenges emblematic of the condition. We also aim to show how atypical posttraumatic cervical dystonia can be alleviated to a high degree using botulinum toxin (Botox) as shown in previous studies and providing validation that it is a real condition [22-25]. We will also discuss recent adjustments to the classification scheme of posttraumatic cervical dystonia [26].

Presentation and Assessment

A 22-year-old Caucasian man arrived at the pain clinic for an evaluation of neck, upper back, and shoulder pain at the request of his primary care provider. Pain was described as his whole body feeling “inflamed” and “like a nerve burn”. Accompanying relevant history indicated that he had been struck by a sail boat boom, sustaining a concussion, in 2011: nearly six years prior to his current visit. During
that the patient complained of persistent axial neck, trapezius, shoulder, and lower back pain worsening with concomitant sore throat, redness, and swollen tonsils during the past six months. The original pain had been managed but not eliminated for several years using alternative medication, medical marijuana, acupuncture, and manipulation. The exacerbation of the pain with onset of six months prior to his visit had uncertain etiology but the patient noted that it coincided with studying in college. He believed that his studying may therefore have been contributory. Lifting, bending, lying, and sitting were described as provocative for his pain.

In addition, the patient stated that he had had constant involuntary spasms in his neck muscles since his 2011 injury. This symptom had worsened during the eight months prior to his visit. Because of this muscle activity, the patient complained of decreased range of motion in all neck planes. The patient also noted a frequent head tilt and rotation.

Social history was significant for major stressors both in addition to and due to his chronic pain since the 2011 injury. The patient stated that he had lost his mother to cancer within the year prior. He had also been having problems with his roommate and had had to close his business. While he stated that he was not currently suicidal, the patient noted that he had had some suicidal thoughts in the past for which he was receiving therapy. He stated that the suicidal thoughts originated at the thought of having his chronic persistent pain for the rest of his life. Compounding these problems was the reality that he had seen thirty physicians over the course of the prior six years: none of whom could provide clarity or a conclusion to his chronic pain condition.

Examination found a 22-year-old Caucasian male with pain and numbness described as pricking, aching, burning, throbbing, sharp, and stabbing. Pain score was assigned at 9/10. Numbness and tingling were found in the right forearm and tips of the fingers. Generalized weakness was found in the hands bilaterally. Cervical, thoracic, and lumbar paraspinal muscles were positive for spasm. Tenderness to palpation was found in bilateral trapezius, posterior neck, occiput, temporal, and supraorbital areas. Other positive findings included a left head tilt and rotation. Review of systems was significant for feelings of tiredness (fatigue), anxiety, depression, and sleep disturbances. Other systems were unremarkable.

Imaging studies included a MRI of the cervical spine without contrast ordered at the described visit and a recent CT scan. Findings were significant for minor irregularities of the vertebral endplates not typical of Schmorl’s nodes. These irregularities were most notable at the superior and inferior endplates of C6 and were presumed to be related to a form of degenerative disease or an old post-traumatic finding. Imaging of both modalities also described a small spur on the anterior superior corner of T1 and mildly prominent tonsils and adenoids. The lymphoidal findings might have been normal for a patient of his age. All other imaging findings were unremarkable with no acute nor active disease processes identified.

Given his presentation and imaging studies, it was determined that the patient might have had a form of atypical posttraumatic cervical dystonia, present since his injury and exacerbated by current life stressors.

**Treatment**

By the time of his visit, the patient had tried several treatment modalities for his chronic pain. Slight but temporary relief had been obtained using physical therapy, TENS (transcutaneous electrical nerve stimulation) therapy with ibuprofen, acupuncture, meditation, oxycodone, medical marijuana, ice, and heat. Additionally, a year prior to his visit, our patient had received a cortisone injection, possibly at the level of C2, from which he had good relief for a period of one to two weeks. Chiropractic adjustments, home exercise programs, and activity in general had all been unsuccessful in alleviating his chronic pain.

Current medications at the time of his visit included Intuniv 2 Mg oral tablet extended release 24 hour, medical marijuana, and OxyCODONE HCl 5 Mg Oral Tablet.

The proposed treatment plan following his visit included continuing these medications, starting Tizanidine 2 mg tablets 1-2 PO TID pm, and discontinuing other nonsteroidal anti-inflammatory medications in favor of Meloxicam 7.5 mg PO BID. Our patient was also advised to continue activity as tolerated. Physical therapy was prescribed.

In addition to these regular therapies, our patient was scheduled for a one-time nerve block and trigger point injections. Nerve blocks were administered bilaterally to the greater, lesser and third occipital, auriculotemporal, zygomaticotemporal, supraocular and supraorbital nerves. Trigger point injections were administered in the bilateral trapezius, rhomboids, and cervical paraspinal groups. Patient tolerated the procedures well with no complications. Pain level before the procedure was 5/10. Pain level after procedure was 0/10. He was given two weeks to monitor his response to the injections.

At his two-week follow-up for his nerve block and trigger point injections, patient reported that he had experienced a 100% improvement in his pain for approximately four days. This improvement was followed by a gradual reduction in his improvement but reports that he still is 20% improved since the procedure and 60% improved since his initial visit with us and the onset of the new treatment plan. Our patient reported a pain score of 4-5/10 at this follow-up visit. Review of systems was unremarkable given his known history. He received another set of nerve block and trigger point injections for another temporary relief of his pain from a score of 4/10 to 0/10.

The persistence of his pain prompted a discussion and education about the use of Botox injections as a treatment modality for posttraumatic cervical dystonia. He agreed to the procedure and received Botox in the procerus and bilateral corrugator, frontalis, temporalis, occipitalis, cervical paraspinal, and trapezius muscle groups roughly five weeks after his second treatment with nerve block and trigger point injections. The procedure comprised of the incremental injection of 165 units of Botox (5 units per location) using a 30 gauge 0.5-inch needle. The procedure was well tolerated with no complications resulting in a pain score reduction from a 2/10 before the procedure to 0/10 after. Our patient stated he had two months of 100% relief post Botox injections. This dropped to 95% after 2 months and at 5 months post injection he was still 90% improved. He had repeated Botox injections at this time with 100% relief immediately following the injections.
Outcome
Upon follow-up, our patient reported a marked reduction in pain as well as improvement in function. He stated that overall; he had an 80% improvement in his pain over his prior injections. He also stated that he was very happy with the results, had returned to his business and to school, and had become much more active; able to exercise and swim regularly. At six months, he reported that he remained 90% improved since his last injections and had two full months of 100% relief. He received additional Botox injections at that time with no complications and was discharged in good condition. Overall, this case represents a successful use of Botox in conjunction with other therapies to treat a posttraumatic cervical dystonia. It also highlights the need for physician education about this condition and treatment given that our success occurred in the context of some thirty prior unsuccessful doctors’ visits resulting in immense emotional burden for our patient.

Discussion
Chronic pain impacts the daily activity, productivity, and quality of life of the affected individual. These effects can have severe consequences both financially and emotionally. Those suffering from chronic pain have been shown to have a higher risk of developing major depression [27,28]. They also experience elevated suicidal ideation, with such thoughts being, on average, three times more common in people with chronic pain than in those without [29]. Some evidence exists to suggest that the likelihood of suicidal ideation correlates with duration of pain experienced [30]. What this highlights is a need for those with chronic pain to have a diagnosis and treatment plan in place as soon as possible. Achieving this has found to be difficult for both physicians and patients. Somewhere between 40 and 80% of all chronic pain patients are misdiagnosed [1]. Furthermore, physicians report low satisfaction with regards to their ability to treat chronic pain. One study of 111 primary healthcare providers resulted in an average satisfaction score of 1.9 (SD: 0.81) on a scale of 0 (not at all satisfied) to 4 (very satisfied) when asked to rate their satisfaction in their ability to treat chronic pain [14].

Patients afflicted with cervical dystonia are not immune to the pitfalls of chronic pain identification and diagnosis. A 2011 Canadian survey found that, on average, patients with a current cervical dystonia consulted approximately three physicians before receiving their diagnosis with average lag time between onset and diagnosis being 6.4 years [15]. Our patient experienced a similar lag time with a far greater number of provider consults at 30 physicians. This amounted to significant use of healthcare resources, personal obstacles, and an immense emotional toll including suicidal ideation. This was an extreme case illustrating both the challenge of diagnosing and treating cervical dystonia but also the consequences that such a missed diagnosis can have.

Why cervical dystonia evades diagnosis is multifactorial. Some point to physicians having little exposure to the condition in their training or practice due to its low prevalence [15]. By one estimate, there are between 20 and 4,100 cases per million [7]. Additionally, physicians have difficulty managing these types of conditions in general. In a 2016 review article, it was reported that physicians oftentimes do not take the careful and thorough history necessary for an accurate diagnosis of a pain condition. Furthermore, they often rely on anatomical testing, such as an upright cervical spine X-ray, rather than utilizing the more useful physiological tests, like flexion-extension X-rays, to guide their diagnostic determination [1]. Lastly, cervical dystonia presents with nonspecific and variable symptoms. Therefore, its diagnosis is highly based on specific clinical diagnostic skills. Meaning that it is a condition that can be evasive unless the patient encounters the appropriate specialist, like a neurologist or movement disorder specialist [15,31,32,]. In aggregate, these characteristics contribute to multiple doctors’ visits, prolonged lag time between onset and diagnosis, and misdiagnoses for those with cervical dystonia.

By presenting this case, our aim is to increase awareness of cervical dystonia and its treatment. Based on the consensus of a 2013 international committee of movement and disorder experts, dystonia is defined as a movement disorder characterized by 1) sustained or intermittent muscle contractions causing abnormal, often repetitive, movement postures or both, 2) typically patterned, twisting, and maybe tremulous movements, and 3) provocation by voluntary action and associated with overflow muscle activation [26]. The committee also devised a classification system based on age of onset, body distribution, temporal pattern, associated features, and etiology [26]. By their convention, our patient potentially had an acquired isolated focal dystonia with onset in adulthood. We use here the traditional convention of atypical secondary or posttraumatic cervical dystonia to avoid confusion considering the transition in terminology. Despite the diagnostic conventions championed by the committee, they still acknowledge the challenges of identifying and classifying dystonia. There is a large degree of phenotypic variability and frequent overlap among different syndromes [26]. Diagnosis and classification is based predominantly on recognition of clinical features with laboratory testing having minimal usefulness [12,33]. But once a diagnosis can be made, it is a treatable condition.

Botox injections have been found efficacious in improving the posture, pain, and disability of those afflicted with cervical dystonia [20,22,24,25]. These improvements are both subjective and objective and have been found to persist long term [25]. Such was the case for our patient who saw dramatic improvements in his pain and functionality; resulting in significant personal gains. This finding was remarkable in the context of the suffering that he endured prior to visiting our pain clinic. Our patient was seen by some thirty physicians over the course of six years. He had also expressed suicidal ideation. These features of his enduring condition are sadly not unique for those with chronic pain generally and dystonia specifically. Studies have shown a commonality about delayed diagnosis and treatment of these conditions. Others indicate that it is not simply the rarity of these conditions but a potential deficit in the education of physicians that results in such a high burden for the patient, the healthcare system, and our economy. By reporting this case, our aim is to increase awareness of cervical dystonia, and strengthen the perception that it is a valid and treatable condition, with the hope that those afflicted will receive the appropriate recognition, care, and treatment in a timelier manner [34-35].

Acknowledgement
‘We thank Dr. David J. Mokler, Ph.D of the Univeristy of New England for his generous support of this publication’.

References
2. Institute of Medicine (US) Committee on Advancing Pain


