Case Report

To spasm, or Not to Spasm, That is the Question

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Introduction

Typically, cervical dystonia (CD), or spasmodic torticollis, is described as a condition in which the cervical paraspinal muscle groups in the neck undergo uncontrollable and prolonged spasm. It has been well described in the literature that CD presents in most patients with an extreme head tilt or tremor due to spasm. This has become the all-encompassing definition of CD to many clinicians. However, in a medical review of over 260 patients with CD, the authors found that only 62% of patients present with the symptoms of spasmodic torticollis [1]. This means that 38% of CD cases do not manifest in a significant head tilt or tremor, which we will refer to as atypical cervical dystonia [1]. We describe a case of atypical cervical dystonia in which OnabotulinumtoxinA (ONA) was used to successfully treat the condition.

Case Study

A 77 year-old Caucasian female was referred to our clinic in 2013 with a three-year history of bilateral neck pain, which radiated into the posterior aspect of the cranium. She stated her pain had been steadily increasing over the past six weeks before her visit, but had been present for many years. The pain was described as an ache that the patient stated could range from a rating of a 5 to a 9 on the Visual Analog Scale. The patient also stated that the pain was more prominent on the left, but was present bilaterally. Holding objects or lying in bed exacerbated her pain. This made it difficult for her to perform daily tasks or to sleep. The patient claims to have tried physical therapy, occupational therapy, as well as treatment with NSAIDs and claimed that all treatment options had helped minimally. In addition to pain, the patient complained of bilateral arm and hand numbness and tingling, this was mainly in the ring and middle fingers. The patient had a history of low back surgery and did not complain of any bowel or bladder issues.

Upon physical exam, the patient’s deep tendon reflex was +2/4 in all of the upper and lower extremities. Hoffman’s reflex was negative bilaterally and Lhermette’s sign was also negative. Spurling’s test was positive bilaterally, which produced numbness in the hands. There was tenderness to palpation of the bilateral trapezius, upper neck, and occipital area, which produced 100% reproduction of pain symptoms. The patient had constant involuntary spasm of the cervical paraspinal muscles, constant moderate head tilt, and decreased range of motion of the head and neck in all planes due to spasm. Prior studies included an MRI in August of 2013. The results of which showed chronic spondylosis in the cervical spine from C4-C7 as well as chronic facet arthropathy. The patient had EMG nerve conduction studies, which determined that the tingling in her upper extremities was due to carpal tunnel syndrome and not a cervical radiculopathy.

The patient also described the pain as radiating from the neck and upper back to the occipital, temporal, and supraorbital areas and these areas accounted for 95-100% of pain complaints. Because of these complaints the patient was then treated with trigger point injections of the bilateral trapezius, rhomboid, and cervical paraspinal groups and nerve blocks targeted at the left occipital, ariculotemporal, and supraorbital nerves using local anesthetic (Bupivacine 0.25% and Lidocaine 1% in a 1:1 mixture with a total volume of cc’s) and steroid (Triamcinolone 40mg 40 mg/cc). She also received trigger point injections (5cc of each 0.25% Bupivacaine and 1% Lidocaine 1:1 mixed with 10mg Triamcinolone 40 mg/cc) into the bilateral trapezius, rhomboid and cervical paraspinal groups. She indicated these injections improved her pain and functionality by more than 80% at her 1 week follow up appointment. However, within 1 month the pain has returned close to baseline. These injections were repeated and at her 3 week follow up she noted improved sleeping pattern but only a 65% reduction in pain and improvement in functionality A third repeated series provided only a 1.5 week improvement with a blunted response that she noted “just took the edge off” before returning to close to baseline levels.

Because of the positive but diminishing response to the initial injections, the patient was given injections to the occipitalis, cervical paraspinal and trapezius muscle groups using ONA, brand name Botox. After the first round of ONA injections the patient reported a 75% improvement in both pain and functionality and stated she was continuing to improve week by week. After the second round of ONA injections the patient stated that her pain and functionality had been improved nearly 100%. Her range of motion had increased greatly in all planes after the first series of ONA.
injections and her range of motion improved even more after the second injection series. The patient also reported that her sleep had improved from 2-3 hours per night before ONA injections to 6-8 hours per night after the injections. Botox injections were started in September of 2014 and have continued to confer significant benefit to the patient with improvements ranging from 75-100% with each injection series.

**Discussion**

CD is the most common form of focal dystonia and has varying degrees of severity that are all grouped into the same category, classically [2]. The main symptom is uncontrollable spasm of muscles in the cervical paraspinal muscle groups causing decreased range of motion, head tilt, tremors, or all three. CD is classified as a neurological disorder, but torticollis can arise from other alterations in neck anatomy, like changes in vertebral structure and alignment or as a result of infection [3]. Forward, backward, rotational, and lateral head tilts are known as anterocollis, retrocollis, rotational torticollis, and laterocollis respectively [4]. The condition is more common in women than men at almost a 2:1 ratio [1]. There is also a genetic correlation in that individual’s with a family history of cervical dystonia are more likely to develop the condition [1].

Additionally, 20% of individuals with CD have some form of dystonia in another part of the body [4]. Our patient did not have a focal form of dystonia in any other part of the body. Reviewing the literature it appears the conventional classification is to label all cases of cervical dystonia as spasmodic torticollis, which entails a significant head tilt, twist or tremor. However, 38% of patients with CD do not exhibit spasmodic torticollis [1]. This means that up to 38% of patients with CD who could benefit from treatments that effectively treat classical CD, or spasmodic torticollis, are not considered for these treatments because they do not exhibit the classic or extreme symptoms. Therefore, we argue spasmodic torticollis is a form of CD, but not the only form of the condition. In terms of treatment options for CD, there is a wide range of treatment modalities. The treatment option that has come to the forefront is the use of ONA, brand name Botox, to treat CD. Oral medications, such as anticholinergic agents, can be used, but these treatments have shown little success [5]. ONA has been shown to reduce or prevent spasm in muscle groups [6]. The prevention of spasm can stop painful muscle contractures and help prevent the development of radiculopathies [7]. It is also an effective treatment for migraine headaches and some forms of neuralgia [8,9]. The effects of ONA can last for 3-4 months in many patients [10].

For those individuals who do not respond to ONA injections, there are other, more invasive, treatment options available. One is the employment of peripheral denervation to stop innervation of the muscle groups in the neck to reduce pain and spasm [7]. Another treatment option is the use of deep brain stimulation [11].

What we assert is that there are varying degrees of CD and a significant number of patients do not fall under the classical definition of Cervical Dystonia, or spasmodic torticollis. For this reason, we refer to the many of the remaining 38% of patients, who do not fall under the classical definition, as having atypical cervical dystonia. The treatment options that are effective for classical CD are also effective for individuals with atypical CD, as we demonstrate here through our clinical example. We believe more research into this poorly defined and specified group deserves attention.

**References**
