Posttraumatic Focal Dystonia of the Shoulder

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abstract

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Focal posttraumatic shoulder dystonia is a rare and not easily identifiable entity. Its true pathophysiologic nature, predisposing factors, and disease course remain debatable.

This article describes a rare case of a 40-year-old man with late symptoms of focal shoulder dystonia after peripheral trauma of his left shoulder girdle. The shoulder was indirectly injured from the impact of a fall off his motorbike 3 years earlier. He was referred to the authors’ institution because remarkable reduction of arm abduction, muscle spasms, and circumscribed hypertrophy of the trapezius muscle were noted while his head and neck were in neutral position and had a full range of motion. The left shoulder had a fixed elevated posture compared with the contralateral shoulder. A continuous burning pain was localized over the area of the hypertrophied trapezius muscle, radiating to the ipsilateral side of the head and neck. Dystonic movements of the trapezius, rhomboid, and supraspinatus muscles were observed. The abduction of the shoulder was significantly decreased, and any repetitive effort for arm abduction induced an exaggeration of his movement disorder, leading to a more pronounced shoulder elevation.

Plain radiographs and magnetic resonance imaging of the left shoulder revealed a suprascapular tendinitis with no other abnormalities. Repeated needle electromyography of the left trapezius muscle and neurography of the accessory nerve on both sides were normal. Injections of botulinum toxin A were effective in the resolution of muscle hypertrophy and abnormal posture.

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Diagnosis, adult-onset focal dystonias are common in movement disorders. They tend to involve the upper body and typically manifest in certain anatomic sites. Cervical dystonias (eg, spasmodic torticollis), cranial dystonias (eg, blepharospasm or oromandibular and lingual dystonias), and task-specific hand dystonias (eg, writer’s cramp or other occupational hand dystonias) are included in the setting of pathophysiologic features.

Isolated dystonic shoulder elevation in the absence of cervical dystonia is rare. It is a source of disability as a result of compromised shoulder motion, especially arm abduction, and is typically painful. Often the initial treating physician fails to recognize it as dystonia because of the unconventional presentation.

This article describes a rare case of posttraumatic shoulder dystonia that was recognized late and fully recovered after botulinum A toxin injections.

Case Report

A 40-year-old man presented to the authors’ outpatient clinic with chronic, unrelenting pain of his left upper limb and left shoulder girdle. Although his recent medical history was unremarkable, he reported a traumatic injury after a fall from his motorbike at the age of 37 years, touching with the left hand and the left arm held in extended position. The shoulder was indirectly injured from the impact of the left hand and the left arm held in extended position. The shoulder was indirectly injured from the impact of the left arm. The patient sought medical attention at that time; however, only rotator cuff tendinitis was diagnosed and all related laboratory and imaging studies were unremarkable. Initial management included nonsteroidal anti-inflammatory drugs and shoulder immobilization in a sling.

His symptoms did not improve, and the painful shoulder elevation was exaggerated in the following months. Shoulder elevation was reported to persist during sleep, with a reduction of pain during the first few morning hours. The patient returned to work 6 months after initial trauma, receiving analgesics and physical therapy for the diagnosed rotator cuff tendinopathy.

Three years later, he was referred to the authors’ institution for further evaluation because, during a routine follow-up visit at his physician’s office, a remarkable reduction of arm abduction (Video 1), muscle spasms, and circumscribed hypertrophy of the trapezius muscle were noted (Video 2) while his head and neck were in neutral position and had full range of motion. Along with muscle spasms of the trapezius muscle, the left shoulder had a fixed elevated posture compared with the contralateral shoulder. A continuous burning pain was localized over the area of the hypertrophied trapezius muscle, radiating to the ipsilateral side of the head and neck. Dystonic movements on the dorsal aspect of the left shoulder were observed. The movements concerned the trapezius, rhomboid, and supraspinatus muscles and led to a rotation of the left scapula. The abduction of the shoulder was significantly decreased, and any repetitive effort for arm abduction induced an exaggeration of his movement disorder, leading to a more pronounced shoulder elevation.

Repeated needle electromyography of the left trapezius muscle and neurography of the accessory nerve on both sides were normal. Plain radiographs and magnetic resonance imaging of the left shoulder revealed suprascapular tendinitis with no other abnormalities. Magnetic resonance imaging of the cervical spine was normal, with no signs of cervical myelopathy. The diagnosis was posttraumatic focal dystonia of the shoulder. A possible provocative factor for the development of this disease was his chronic heavy labor, including heavy lifting and the use of air drills.

Medications including analgesics, baclofen, anticholinergic drugs, antidepressants, and benzodiazepines were prescribed and improved his pain periodically; however, they did not improve his shoulder range of motion or arm abduction. Botulinum toxin A injections (40 IU to the supraspinatus, 25 IU to the anterior part and 25 IU to the posterior part of the deltoid, 50 IU to the teres major, and 50 IU to the subscapularis muscle) were administered and then repeated 3 months later. Four weeks after the last injection, shoulder range of motion and function significantly improved (Video 3). The patient’s Constant score improved from 35 to 87 of a maximum 100.

Two years after initial evaluation at the authors’ institution, the patient reported no remarkable pain, was satisfied with his shoulder function, and had successfully returned to his activities of daily living while receiving 1 mg of biperiden twice a day.

Discussion

Focal shoulder dystonia is a diagnosis that is usually missed or misinterpreted. Although it is rarely reported in the current literature, its true prevalence is unknown because the diagnostic criteria are not clearly defined. In most reports, the disease is characterized by isolated dystonic shoulder elevation following local trauma, which fits into the expanding category of dystonia provoked by peripheral (non-brain) injuries. A muscle spasm in response to a painful local injury to the shoulder girdle could be a common misdiagnosis, and focal dystonia should be considered as a potential cause of motion impairment. The persistence and severity of clinical symptomatology, the development of secondary muscular hypertrophy, and the resemblance of dystonic shoulder elevation to that associated with idiopathic cervical dystonia suggest that this disorder is not a simple spasm secondary to pain.

Should this pathologic entity be considered a new form of dystonia? In the largest series of 13 similar cases, Wright and Ahlskog reported that the term dystonia was appropriate in patients presenting with an abnormal, painless contraction of the trapezius muscle and the involvement of other muscles.

How can peripheral trauma cause focal or generalized dystonia? Damage to a pe-
Peripheral nerve or root may result in long-term changes in spinal or central synaptic mechanisms, a process in dystonia analogous to that in causalgia or phantom limb pain.\textsuperscript{3,12} Animal studies have shown that changes in afferent input into the spinal cord or brain stem can lead to reorganization in the central nervous system.\textsuperscript{15,16} Dystonia may result from synaptic reorganization of this kind, with a substrate of disordered intraneuronal function (possibly inherited).\textsuperscript{15,16} Moreover, it has been reported that a central motor reorganization, which plays a significant role in the development of the movement disorder, can be originated by a causative peripheral injury long before the development of focal dystonia.\textsuperscript{15,16}

Jankovic\textsuperscript{1} and Jankovic and Van der Linden\textsuperscript{17} reported that, in some cases of posttraumatic movement disorders, several features considered typical of dystonia, such as the presence of torsional movements, task specificity, or relief by antagonistic gestures, may be lacking. However, the features presenting in most of Wright and Ahlskog's\textsuperscript{5,7} and Cossu et al's\textsuperscript{8} cases did not exclude a nondystonic, peripheral nerve-induced spasm of the cervical muscles.

Several types of dystonia have been recognized, including the cervical type (eg, spasmodic torticollis),\textsuperscript{13} the cranial type (eg, blepharospasm or oromandibular and lingual dystonias),\textsuperscript{19} and task-specific hand dystonias (eg, writer's cramp or other occupational hand dystonias).\textsuperscript{20} They are usually related to disturbances of head and neck posture resulting from muscular or radiologically identifiable disorders (eg, posterior fossa and cervical spinal cord tumors, craniofacial junction or cervical spine abnormalities, focal myositis, or fibrosis).\textsuperscript{21} Focal posttraumatic dystonias are related to a recent traumatic event in an extremity that is not accompanied by central nervous (ie, brain or cervical) trauma.\textsuperscript{22} However, some authors consider this motor disorder a nondystonicic spasm or pseudodystonia, suggesting that it might have a different (peripheral) pathophysiology than true dystonia.\textsuperscript{23,24}

Contributory factors generally include positive family history of dystonia, early-life developmental delays, perinatal brain insults, use of stimulants or neuroleptics, and heavy labor.\textsuperscript{25,26} The current patient was a heavy laborer; however, no other contributory factor was present. Other authors have reported that trauma to peripheral body parts can trigger dystonia independent of brain injury.\textsuperscript{3,9} In such cases in which relatively minor trauma induces dystonia, it is presumed that some underlying inherent propensity likely exists for this to occur. Consistent with this presumption, Höllinger and Burgunder\textsuperscript{12} reported a patient involved in 2 motor vehicle accidents 2 years apart who developed a more widespread axial dystonia and dystonic foot inversion, similar to that of generalized idiopathic torsion dystonias. However, in a retrospective review of similar cases, predisposing risk factors were not always apparent.\textsuperscript{5}

Diagnosis is mainly based on the history of previous trauma and the clinical evaluation that reveals the presence of muscle hypertrophy and dystonic movement.\textsuperscript{5} Magnetic resonance imaging is essential to exclude potential pathology from the brain and cervical spine.\textsuperscript{24} The effectiveness of electrophysiological studies is debatable.\textsuperscript{24,27} Cases of isolated neurogenic muscle hypertrophy secondary to radicular or peripheral nerve injury have previously been described.\textsuperscript{10,11} In these cases, a peripheral (neurogenic) origin was indicated by electromyography with large and/or complex motor unit potentials and characteristic firing patterns (eg, complex repetitive discharges or myokymia).\textsuperscript{10,11} Wright and Ahlskog\textsuperscript{5} reported 2 of 13 patients with a typical clinical presentation whose electrophysiological studies showed inconsistency of the clinical and electrophysiological findings. However, the persistence of the condition during sleep in some cases suggested an organic cause for the patients' symptoms.\textsuperscript{5}

In the current case, no electromyographic abnormalities were revealed.

The prognosis and physical history of the disease is unknown.\textsuperscript{1,12} No study supports a spontaneous resolution of the disease. Several studies report dissociation between the degree of pain and the muscle hypercontraction, with the latter being a relatively constant state\textsuperscript{28,29}; reduced pain resulting from positioning, analgesics, or other factors did not result in alleviation of the muscle contraction state.\textsuperscript{28,29}

Treatment of these patients with oral medications is often unsuccessful, apart from analgesics for pain control. Administration of botulinum toxin A is reported to be beneficial, although not completely effective.\textsuperscript{30,32} The idea that dystonia is related to peripheral trauma has developed alongside the successful use of chemical trauma as a form of treatment. Botulinum toxin A injected into affected muscles will relieve cramps, improve neck twisting in torticollis, and restore vision in blepharospasm.\textsuperscript{19} However, sometimes little improvement occurs in patients with oromandibular or cervical dystonia, and the procedure must be repeated every 3 months.\textsuperscript{13,26,33} Alternatively, high-dose anticholinergic drugs may be effective, particularly in young people with generalized dystonia. Other forms of dystonia may respond to alcohol, levodopa, or carbamazepine.\textsuperscript{24} Future work includes the development of central muscarinic M2 receptor antagonists and drugs that act on central opioid receptors and may restore normal intraneuronal function.\textsuperscript{3} The more favorable outcomes from the use of botulinum toxin A injection in patients with focal posttraumatic dystonias compared with cervical dystonias may reflect the more limited involvement of muscles.\textsuperscript{2,13}

**Conclusion**

Focal posttraumatic shoulder dystonia is a rare and not easily identifiable entity. Its true pathophysiological nature, predisposing factors, and disease course remain debatable. However, it is generally accept-
ed that any patient with dystonic arm abduction and shoulder posture, combined with hypertrophy of the shoulder musculature, chronic unrelenting pain, and a history of trauma, could be a candidate for the disease. In the current case, botulinum toxin A injection was effective in resolving muscle hypertrophy and, consequently, abnormal posture and functional impairment. The significant clinical improvement of the patient may be explained by the ability of botulinum toxin A to reduce muscle hypertrophy and thus correct the abnormal posture and motion.

**REFERENCES**


