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Intermittent cerebral ischemia as a cause of dystonic storms in hypermobile Ehlers-Danlos syndrome with upper cervical instability, and Prolotherapy as successful treatment: 4 case series

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Abstract

Dystonic storms are a life-threatening condition that in many cases has no known etiology. We present, for the first time, 4 cases of patients with hypermobile Ehlers-Danlos syndrome whose dystonic storms were reproduced by vascular occlusion of either the vertebral or carotid arteries in the neck. This condition caused intermittent cerebral ischemia, which was documented by cervical motion during upright transcranial doppler examination. All 4 patients were found to have severe upper cervical instability at the atlantoaxial joint, diagnosed by cervical digital motion x-ray (motion videofluoroscopy). They responded favorably to Prolotherapy, resulting in either complete or near-complete resolution of their dystonic storms. Some forms of dystonic storms occur because of vertebral and carotid artery compression in the upper neck due to craniocervical instability, the latter of which can be successfully treated with Prolotherapy with resultant near-complete resolution of dystonic storms.

Keywords

Dystonia; dystonic storms; upper cervical instability; vertebrobasilar insufficiency; hypermobile Ehlers-Danlos syndrome; Prolotherapy; transcranial doppler.

Abbreviations

DMX: Digital motion x-ray; hEDS: Hypermobile Ehlers-Danlos syndrome; MRI: Magnetic resonance imaging; POTS: Postural orthostatic tachycardia syndrome; RIT: Regenerative injection therapy; TCD: Transcranial doppler.

Introduction/Background

Dystonic storm is a life-threatening form of generalized dystonia. Also known as status dystonicus or dystonic crisis, it manifests as increasingly frequent and severe episodes of generalized dystonia characterized by tachycardia, tachypnea, and autonomic instability. It requires emergent care when progression to bulbar dysfunction with dysarthria, dysphagia, and respiratory failure become evident. Dystonia itself is a hyperkinetic movement disorder characterized by sustained muscle contractions, leading to repetitive twisting movements and abnormal posture [1,2]. While dystonia, being a movement disorder, has commonly been associated with hypermobile Ehlers-Danlos syndrome (hEDS), dystonic storms are extremely rare even in a cohort of patients with hEDS and dystonia [3]. Although there are known triggers and brain structural lesions that can provoke dystonic storms, if initial history, blood tests, and MRI scans are unremarkable, the exact etiology in the majority of cases remains elusive [4].

We present the first 4 reported cases of patients with hEDS who each had full and partial dystonic storm episodes brought on by cerebral artery ischemia from neck motion (i.e., cervical motion-induced extracranial arterial occlusion), documented by extracranial and/or transcranial doppler ultrasound. All 4 patients were found to have extreme upper cervical instability seen on digital motion x-ray (dynamic videofluoroscopy) who experienced complete or near-complete resolution of their dystonic storms once their upper cervical instability was successfully treated with Prolotherapy, and they were counseled on the neck motions and positions that induced cerebral ischemia and their dystonic storms.

Case Series

Patient #1: A 44-year-old female with hEDS and quinolone-induced tendinopathy (taken for episodes of pneumonia and breathing difficulties), as well as a history of being in a severe car accident at the age of 17, came to Caring Medical complaining of having up to 20 dystonic storms a day and reported that they have been ongoing for the last 9 years. She visited many emergency rooms and underwent numerous hospitalizations. All diagnostic testing, including brain MRIs, were inconclusive. No medication was found very effective to reduce the dystonic storm activity. She was in total fear and distress, and unable to have gainful employment. When these episodes occurred, she was aware they were happening but was helpless to do anything about them because she had tremendous difficulty breathing and was unable to talk. During an attack, she said her body gyrated, rotated, twisted, and curled on its own while her diaphragm spasmed uncontrollably and her extremities became numb (Figure 1). Aside from the grueling dystonic storms this patient experienced daily, she also reported the following symptoms: dizziness, balance difficulty, severe neck pain, headaches, joint subluxations, intestinal bleeding, brain fog, light and sound sensitivity, dizziness, focal dystonia, colitis, ear fullness, dysautonomia, and postural orthostatic tachycardia syndrome (POTS).

Since the patient believed many of her symptoms could be due to cervical instability, she came to our office to have a videofluoroscopic evaluation called digital motion x-ray (DMX) taken and hopefully confirm this as a diagnosis. The DMX did in fact reveal significant upper cervical instability (Figure 2). As she had a myriad of symptoms, in addition to the dystonic storms, the patient then underwent a dynamic transcranial doppler (TCD) examination where the head and neck were put through a variety of movements

and held at the full end of the range of motion for 15-30 seconds. It revealed complete cessation of the left middle artery blood flow with neck flexion and right head rotation (Figure 3). This caused a mild dystonic storm. Transcranial and extracranial doppler also revealed cessation of vertebral artery flow (at V3 and V4 segments) with neck extension, followed by a full dystonic episode (Figure 4). These manifestations were significantly reduced immediately after her first Prolotherapy session and she was placed in a cervical collar. After it was shown that Prolotherapy treatment had stabilized her cervical spine, the patient no longer needed to wear the cervical collar and it was removed. It has been over a year since her last Prolotherapy visit, during which time she has been without a full dystonic storm episode and her focal dystonias are very rare. Her other symptoms were also markedly reduced.

Figure 1: Patient during a dystonic storm. She was found to have severe upper cervical instability, which caused compression of her carotid artery, leading to a myriad of dystonic storms. By stabilizing her upper cervical spine, Prolotherapy helped resolve her dystonic storms.

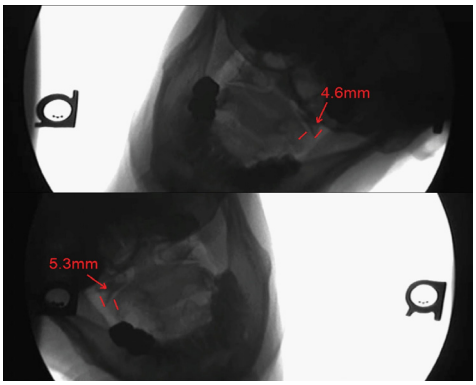

Figure 2: Digital motion x-ray demonstrating severe bilateral atlantoaxial (C1-C2) instability with open mouth view and lateral neck flexion.

Figure 3: Middle cerebral artery ischemia observed during motion transcranial doppler examination. Cessation of the middle cerebral artery wave from (finger point) when patient held a certain neck position. Middle cerebral artery circulation was restored when she put her neck back in the neutral position.

Figure 4: Patient having a dystonic storm after transcranial doppler examination showed decreased blood flow in her middle cerebral artery with specific neck motion.

Patient #2: A 21-year-old female presented to Caring Medical with a 5-year history of chronic illness and 1 year of "convulsions," where she would lose control of her body, causing it to twist and shake uncontrollably. These kinetic movements could last for seconds or minutes and would occur multiple times per day. She saw numerous physicians and had many lab and radiographic analyses, which were inconclusive. Over the years, she had numerous emergency room visits and hospitalizations. This patient had been previously diagnosed with hEDS, which was the apparent underlying cause for her myriad symptoms and multiple diagnoses (Figure 5).

Eventually, the patient had to drop out of college. At presentation, she was mostly bedridden but was using a wheelchair for ambulation. She came in with a central line for IV access because of hypotension and an inability to have necessary blood draws and was on 24 different medications. The patient had been told she was a candidate for cervical fusion but was leery of having this surgery and came to Caring Medical to see if Prolotherapy would be a better option for her.

After a physical examination and a DMX were taken, extreme upper cervical instability at the atlantoaxial joint was revealed. A TCD examination of the middle cerebral artery revealed complete cessation of the right middle cerebral artery blood flow within 10 seconds of neck flexion and left rotation, after which the patient had a full dystonic storm. To resolve the storm, her neck was turned to the right and placed in the neutral position. Once blood flow was restored in the middle cerebral artery, the dystonic storm gradually resolved. Further TCD examination after patient recovery revealed that extension of the neck also reproduced the dystonic storm after middle cerebral artery blood flow ceased.

The patient then underwent Prolotherapy to the areas in her neck that were unstable, especially the atlantoaxial joint, and was placed in a neck collar. While in the collar, she reported having no more dystonic storms. In total, she was consistently in the collar for approximately 2 months. She received a series of Prolotherapy injections to her neck over the course of 4 months. Follow-up DMXs showed improved stabilization of her cervical spine. Upon last correspondence, she reiterated not having any more full dystonic storms for over a year but does have rare "small" focal dystonias of hands/arms. Many of her other symptoms also dissipated, her medication list was down to 2, and she was able to have her central line removed.

Diagnoses	Symptoms	
• Dysautonomia • Dystonic storm • Endometriosis • Gastroparesis • hEDS • Joint dystonia • Migraines • Neurogenic bladder • Tethered spinal cord • Vertebrobasilar occlusion • Vertigo	• Cerebral spinal fluid leaks • Deep vein thrombosis/ pulmonary embolism, postural orthostatic tachycardia syndrome, cervical instability, neck pain • Dizziness • Drop attacks • Ear fullness • Eye tearing • Facial pain and numbness • Frequent joint subluxations/ dislocations, body pain	• Heart palpitations, mast cell activation syndrome • Hoarseness • Incontinence • Intermittent eosinophilia • Memory decline • Nausea, weight loss • Poor balance • Temperature dysregulation • Vision changes, sensitivity to sound and light

Figure 5: The additional diagnoses and symptoms of patient #2.

Patient #3: A 26-year-old female with a history of hEDS came to Caring Medical, saying she had been having episodes where she lost control of her body ever since she was in a motor vehicle accident 4 years ago. She described these episodes as being whole body seizure-like activities where she would almost lose consciousness. They initially occurred many times a day and lasted seconds to minutes. All diagnostic tests for the cause of the whole body's twisting and posturing were inconclusive. Because the episodes were triggered by neck motions, she had been careful moving her neck, which would decrease their frequency.

This patient had also been diagnosed with joint dystonia, restless leg syndrome, POTS, mast cell activation syndrome, chronic fatigue syndrome, vertigo, and dysautonomia. Her many symptoms could keep her bedridden for weeks at a time and included the following: joint dislocations, loss of ambulation, balance problems, dizziness, neck pain, drop attacks, headaches, fevers, temperature dysregulation, tremors, difficulty swallowing, brain fog, nausea, poor digestion, strange smells, ear fullness, and sensitivity to light and sound. She had numerous hospitalizations because of these symptoms, was taking numerous medications, and had a central line implanted because she also needed frequent doses of intravenous medications and fluids due to her nausea, severe pain, and hypotensive dizziness.

During her visit to our office, a cervical DMX was taken and revealed severe upper cervical instability. The neck motions of flexion and right rotation produced a significant drop of the left middle cerebral artery blood flow on TCD, shortly followed by the beginnings of a dystonic storm. Neck flexion and left lateral flexion caused a significant drop in the right vertebral artery blood flow that also induced dystonic storm activity. Moving her neck out of the induced postures restored blood flow to both cerebral arteries and her dystonic storms were resolved. She was given Prolotherapy to treat her upper cervical instability and placed in a cervical collar. Almost immediately, the frequency and severity of her dystonic storm symptoms dropped considerably. Prolotherapy was able to stabilize her cervical spine to the point where she was dystonic storm-free for 6 months. At that point, she needed to have 2 abdominal surgeries and some of her dystonic activity returned, but only at 10% of the level it was before. A repeat DMX did reveal some upper cervical instability had returned so she is currently under Prolotherapy treatment. Her myriad symptoms continues to improve, but as she has had some other joint instability issues, she is still under periodic care.

Patient #4: When this 22-year-old female who had been diagnosed with hEDS came to Caring Medical, she stated her dystonic storm symptoms all started one day after she caught a cold and was coughing a lot. Dystonic storms were the chief problem she wanted to resolve. During a storm, she lost control of her body, which would twitch and writhe around, and said she could not stop her body movements. The storms became almost nonstop, so she was immediately taken to the emergency room and hospitalized. Despite numerous tests, including brain scans and a myriad of consultants, no answers for her dystonic storms were found. She continued to have almost nonstop twitching in her arms and neck, which was affecting her cognition and breathing. Eventually, she was prescribed clonazepam, which decreased her constant body movements. The patient reported having other symptoms, including severe debilitating fatigue, insomnia, pounding headaches, dizziness, light and sound sensitivity, lower back and neck pain, and joint subluxations, all of which adversely affected her ambulation and ability to work. Initial evaluation by DMX confirmed severe atlantoaxial instability. TCD examination showed no significant drop in MCA velocities with neck motions, but several motions, including extension and left rotation, decreased vertebral artery flow, causing severe dizziness and the start of a dystonic episode. She was immediately started on a course of Prolotherapy and neck immobilization. Treatment resolved her joint instability, causing her dystonic storms to cease and alleviating many of her other symptoms. She has been dystonic storm-free for 6 months.

Discussion

The diagnosis of dystonic storms carries with it significant morbidity and mortality. Once one dystonic storm occurs, the most common scenario is an ever-increasing progression of symptoms and severity. Even with treatment, the mortality remains 10% [5,6]. Recovery from one storm can take weeks. The most serious immediate threat to the life of dystonic patients is respiratory failure triggered by excessive contraction of the muscles of the upper respiratory tract, chest, and abdomen [7]. The severity of this condition is evident by its other names, including status dystonicus, desperate dystonia, dystonic crisis, and life-threatening dystonia. [5,6,8-10]. While known etiologies include structural brain masses and lesions, febrile infections, dehydration, toxicity, dyselectrolytemia, various medications, and neurodegenerative diseases, the etiology of dystonic storms is unknown in approximately half of patients [5,11,12].

Dystonic storms usually occur in patients with known dystonia [5] that is severe or poorly controlled [5,13]. In one-third of events, there is no preceding known provoking trigger [6]. While dystonia is uncommon in the general population, noting a prevalence of 0.7% [14], one clinic that diagnosed 2,212 patients with Ehlers-Danlos syndrome over a 10-year course noted that 75% of the patients had some form of dystonia, including dystonic storms. [3] Many of the patients had symptoms similar to the case studies presented [3] (Figure 6).

hEDS, previously known as Ehlers-Danlos syndrome, hypermobility type 1, is diagnosed strictly on clinical grounds [15]. It is one of a group of connective tissue disorders that are characterized by joint hypermobility, a term defined as “the capability of a joint or group of joints to move, passively and/or actively, beyond normal limits along physiological axes” [16]. Craniocervical instability (upper cervical instability) is known to be associated with Ehlers-Danlos syndrome with a potential host of symptoms including headache, visual disturbances, tinnitus, lightheadedness, dystonia, and dizziness [17-19] While vertebrobasilar and carotid artery insufficiency are well-known causes of many of these symptoms [18,20], dystonic storm is not one of them. These are the first case reports of dystonic storms occurring immediately after compromise of cerebral blood flow with neck motions as documented by TCD.

Neurosonology with transcranial and extracranial doppler provides real-time determination of velocity and spectral waveform of blood flow in the basal intracranial and extracranial (neck) vessels. It has the added advantage that dynamic motions can be done to see their effects on cerebral blood flow [21,22]. It has long been established that head and neck position does influence cerebral circulation, as the internal carotid and vertebral arteries run through the neck [23-25]. The internal carotid arteries furnish blood to the anterior 75-80% of the brain via the middle cerebral artery and the anterior cerebral artery. The posterior part of the brain is supplied via 2 vertebral arteries which join to form the basilar artery, which then communicates with the posterior cerebral arteries. The communication between the anterior and posterior brain circulatory networks comes via the circle of Willis. To assess the brain’s anterior circulation, the middle cerebral artery is typically chosen during TCD because of its superficial location (compared to other brain blood vessels) and it receives the majority (60%) of the blood from the internal carotid artery. The brain’s posterior circulation can be assessed via the posterior cerebral arteries and the

basilar or cranial portion of the vertebral arteries.

The close association between the spinal bony anatomy and the vertebral artery is well appreciated, as it traverses up the cervical transverse foramina from C6 to C2, then loops and turns lateral to ascend into the transverse foramen of C1. It then traverses the posterior border of the lateral mass of C1, making its way intracranially through the foramen magnum (Figure 7). The common carotid artery runs up the neck in the carotid sheath, which is only a few millimeters from the anterior cervical borders as it branches into the external and internal carotid artery at about the level of C3-C4 [26]. From here the internal carotid artery, still in the carotid sheath, transverses just anterior to the upper cervical vertebrae, making a bend around the border of the anterior lateral mass of C1 to enter the cranium through the carotid canal (Figure 8). The brain's blood supply is especially vulnerable at the atlantoaxial joint because of its close association and being the most mobile joint in the whole spine, this joint is thus prone to instability [27,28]. Because of its "native looseness," the C1-C2 articulation is prone to instability; the few millimeters of extra motion then make the internal carotid and vertebral arteries a risk for compression or kinking with certain neck motions.

In all 4 cases described, transcranial and extracranial doppler documented blood flow compromises with specific neck motions, and shortly thereafter (within 10 seconds) a full dystonic storm (patients 1 and 2) or partial storm (patients 3 and 4) followed. Once blood flow normalized, the dystonic storm resolved. All 4 patients were found to have severe atlantoaxial (C1-C2) instability by digital motion x-ray (Figure 9). Motion videofluoroscopy has been shown to have a high degree of diagnostic accuracy for the identification of vertebral instability in patients with symptomatic cervical injuries [29].

Using DMX and TCD technology, our office was able to diagnose each of these patients with cervical instability and carotid or vertebral artery occlusion, respectively. Prolotherapy injections given at the fibro-osseous junction of soft tissue structures such as ligaments stimulate the body to thicken and tighten ligaments [30-33]. Several series of Prolotherapy injections were given under ultrasound or x-ray guidance to the incompetent posterior cervical ligaments at the craniocervical junction (Figure 10). Prolotherapy, a form of regenerative injection therapy (RIT), was successful in correcting the cervical joint instability found in each of our 4 patients, whose medical history included a diagnosis of hEDS and a succession of dystonic storm episodes, as well as a myriad of symptomology spanning multiple diagnoses. The cervical instability, and vertebrobasilar and carotid occlusion, were responsible for causing the dystonic storms in these patients and the majority of symptomology these patients had endured (Figure 11).

Symptoms	Patients Affected (%)
Diplopia	74
Dysautonomia	70
Dystonia	75
Fatigue	93
Gastroesophageal reflux	72
Hemorrhages	93
Hyperacousia	75
Memory decline	72
Multiple Pains	93
Proprioceptive problems	92
Shortness of breath	76

Figure 6: Percentages of various symptoms in 153 patients with hypermobile Ehlers-Danlos syndrome (hEDS). These patients were all examined by the same physician.

Hamonet C, Ducret L, Marie-Tanay C, Brock I. Dystonia in the joint hypermobility syndrome (a.k.a. Ehlers-Danlos syndrome, hypermobility type). SOJ Neural. 2016;3(1): 1-3. doi: 10.15226/2374-6858/3/1/00123

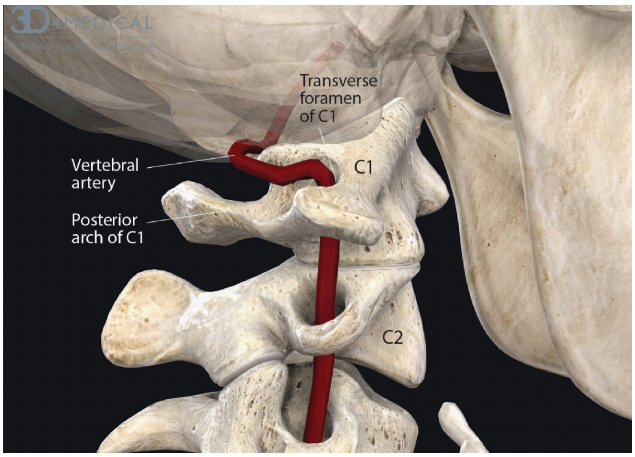

Figure 7: Relationship of the upper cervical vertebrae and vertebral artery. The vertebral artery passes through the transverse foramen of C2-C6 and across the posterior arch of C1.

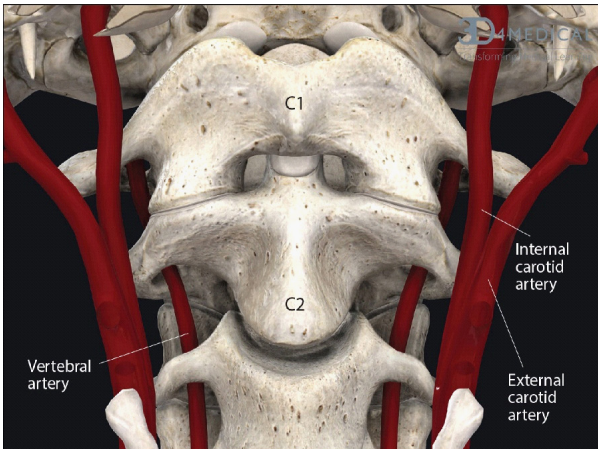

Figure 8: Relationship of internal carotid artery to the upper cervical vertebrae. The internal carotid artery sits just anterior to the transverse processes of the atlas (C1) and axis (C2) Its blood flow can be constricted or blocked by atlantoaxial (upper) cervical instability.

Patient #	Initial Overhang of Atlas (mm)	
	Right	Left
1	5.3	4.6
2	4.0	4.0
3	5.0	4.7
4	3.6	4.6

Figure 9: Lateral displacement of atlas (C1) on axis (C2) on open-mouth digital motion x-ray with lateral flexion.

Figure 10: Prolotherapy cervical injections under mini c-arm x-ray guidance.

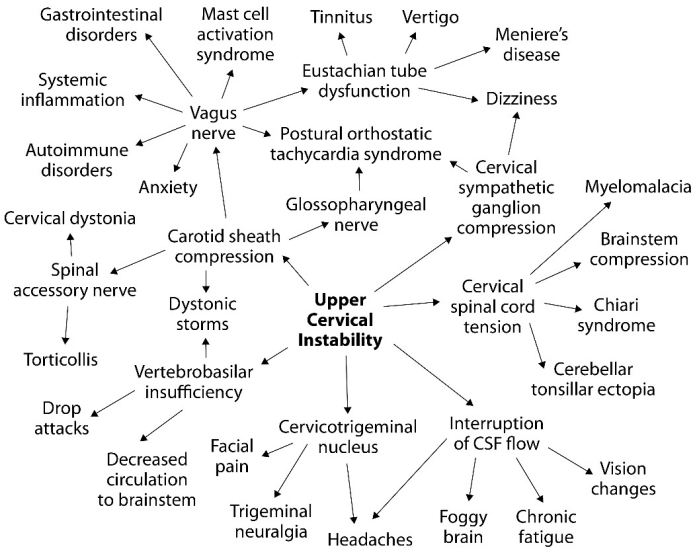

Figure 11: The many syndromes caused by upper cervical instability.

Conclusion

Carotid or vertebral artery occlusion is a treatable cause of dystonic storm and can be identified by extracranial and transcranial doppler ultrasound. Upper cervical instability can be the root cause of both conditions in patients with hEDS. Along with advising hEDS patients on proper neck motion and position awareness, Prolotherapy can be used to successfully stabilize the upper cervical joints, which allows normal blood flow in the vertebral and carotid arteries, causing the dystonic storms to resolve.

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