

Case Report

A Case of Cervical Dystonia - Non Responsive to Usual Noninvasive Treatment

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Abstract

Cervical dystonia is a focal dystonia characterized by neck muscles contracting involuntarily-causing abnormal movement & awkward posture of the head & neck. It is a rare disorder that can occur at any age even infancy. Symptoms generally begin gradually & then reach a point where they don't get substantially worse. Asymmetry of the Sternocleidomastoid is often present in untreated patient. There is no cure for cervical dystonia. Injecting botulinum toxin into the affected muscles often reduce the signs & symptoms of cervical dystonia. Here, it has been described a rare case of a forty seven year old male patient with a history of cervical dystonia presented with struggle with head position, resisting but never overcoming the tendency of his head to assume an unnatural position. The management including drugs like oral analgesic, muscle relaxant, anti-parkinson drug & even antipsychotic drug were used. Psychiatric approach like different modalities of heat such as Short Wave Diathermy, Microwave diathermy & Ultra sound therapy were used. Brace like cervical collar and different types of exercise was also prescribed, even botulinum toxin was injected into suspected muscles but all treatments discussed above failed in this case.

Key word: Cervical dystonia, sternocleidomastoid (SCM), botulinum toxin.

Introduction

Cervical dystonia also called spasmodic torticollis¹ is a painful condition in which neck muscle contract involuntarily, causing head to twist or turn to one side, can also cause head to uncontrollably tilt forward or backward.² Movement is a key finding in cervical dystonia. Another prominent finding is evidence of active muscle contraction in the form of muscle thickening and hypertrophy. The movement may be sustained (tonic), jerky (clonic) or a combination.³ Spasms in the muscle or pinched nerve in the neck can result in considerable pain and discomfort and expanded neck size can occur. Asymmetry of Sternocleidomastoid muscle is often

present in untreated patient. The condition appears sporadically in the absence of a documented family history of the disease. In about 10-15% of cases more than one family member maybe affected. Several families have been attributed with autosomal dominant,⁴ adult onset, primary dystonia. That is focal in distribution affecting the neck region. This form of condition has been called familial torticollis.

Case Report

A 48 years old male patient attended the out patient department of Physical Medicine & Rehabilitation at NICVD, with the complaints of slight pain in neck on one side with deviation of chin to the right shoulder for 1 year. Initially it was occasional & mild but later the symptoms increased with severity. Patient could not hold his neck properly & it makes his neck turns to the right shoulder. Sometime he struggles with head position, resisting but never overcoming the tendency of his head to assume an unnatural posture, which prevents his normal daily life activities & makes his life miserable. Previously he was treated as a case of cervical spondylosis by doing cervical spine X-ray

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where mild osteophytic lipping, were seen in C_{4,5} & C_{6,7} level. On physical examination, the patient's chin turned toward right shoulder & his head turned towards left posteriorly. His sternocleidomastoid muscle (SCM) of left side was prominent & hard. There was also difficulty to keep face forward & look to the left without holding it. But right SCM muscle was normal. Sensation was normal. Neurological examination revealed, no abnormality except mild weakness of right thumb. Routine biochemical investigations were done in addition to copper analysis in urine which were found normal. MRI of brain & neck was done, which were normal except mild cortical atrophy & mild degenerative change of cervical spine. EMG of neck muscle shows increased activity of left sternocleidomastoid & right splenius capitis muscle. There was also increased activity of right semispinalis capitis. Based on history, physical examination, different imaging modalities & EMG (Electromyogram), a diagnosis of cervical dystonia was confidently made. The patient was trying to be managed accordingly by the drugs, physical therapy & orthosis. To relieve the pain, oral analgesic was used. Different muscle relaxants like beclofen, tizanidine etc., different antipsychotic drug like haloperidol, clonazepam etc, antiparkinson drug like carbamazepine, were tried to relieve muscle spasm & dystonia. Sometimes it decreased the intensity of dystonia & pain subsided. Different exercises such as strengthening exercise, range of motion exercise, & stretching exercise were prescribed to reduce the dystonia. The patient was advised to wear a cervical collar during journey & work. Along with changing daily activities & training in stress management technique was also advised. The judicious use of botulinum toxin was tried twice directly into the affected neck muscle. But all these procedures were failed in this case with little recovery of his cervical dystonia.

Discussion

Cervical dystonia is a rare disorder that can occur at any age even infancy. Cervical dystonia most often occurs in middle aged people women affected more than men. In most cases, the exact cause of cervical dystonia (C/D) is usually unknown⁶. The

condition appears sporadically in the absence of a documented family history of the disease. In about 10-15% of cases more than one family member may be affected. Several families have been attributed with autosomal dominant. Initial symptoms of C/D are usually mild. Over time involuntary spasm of the neck muscles will increase the frequency & strength until it reaches a plateau⁷. Symptoms can worsen during stress. Both agonist & antagonist⁵ muscles contract simultaneously during dystonic movement. Usually muscles involved are contralateral sternocleidomastoid & ipsilateral splenius capitis⁵. Other symptoms include muscle hypertrophy, neck pain, dysarthria & tremor. Physical therapy⁸ helps with some of the movement of the head and neck. By stretching out the muscles spasm decreases the length and severity of the attacks. Strengthening exercises are also great for easing pain.^{9,11} The most common treatment for cervical dystonia is the use of botulinum toxin type A¹². This paralyzing agent can be injected directly into the neck muscles affected by cervical dystonia. Most people with cervical dystonia seen an immediate improvement with this treatment but may be repeated every 3-4 months^{1,2}. In our case even botulinum toxin failed.

Conclusion

There is no cure for cervical dystonia. In some people signs and symptoms may disappear without treatment. But recurrence is common. Even surgery like deep brain stimulation may be required to repair the affected muscles. Psychiatric intervention plays an important role by exercises that improve neck strength & flexibility to aid the patient in keeping their head in proper alignment with their body. Judicious use of a neck brace and training in stress management technique also helps a lot. The disability & pain caused by cervical dystonia may result in depression. A high index of clinical suspicion is required for the early diagnosis of cervical dystonia and avoid missing the potentially life miserable condition.

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