

A Case of Cervical Dystonia with 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 and awkward posture of the head and neck. A rare disorder can occur at any age even at infancy. Symptoms generally begin gradually and then reach a point where they don't get substantially worse. Asymmetry of the sternocleidomastoid (SCM) is often present in untreated patient. There is no cure for cervical dystonia. Injecting botulinum toxin into the affected muscles often reduces the signs and symptoms of cervical dystonia. In this case, 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. Physiatric approach like different modalities of heat such as Short wave diathermy (SWD), Microwave diathermy (MWD) & Ultra sound therapy (UST) were used. Brace like cervical collar and different type of exercise was also prescribed, even botulinum toxin injected into suspected muscles but all discussed above failed in this case. [J Shaheed Suhrawardy Med Coll, 2014;6(1):44-46]

Key Words: cervical dystonia, Sternocleidomastoid (SCM), botulinum toxin

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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 backward². 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 maybe sustained (tonic), jerky (clonic) or a combination³. Spasms in the muscles 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 dominant⁴, adult onset, primary dystonia. That is focal in distribution affecting the neck region. This form of condition has been called familial torticollis. Cervical dystonia may be primary (meaning that it is the only apparent neurological disorder with or without a family history) or be brought about by secondary^{3,4} causes such as physical trauma. Cases of inherited cervical dystonia may

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occur in conjunction with early onset generalized dystonia which is associated with the DYT1 gene⁴.

Case Report

A 48 years old male patient attended the outpatient department of physical Medicine & Rehabilitation at NICVD, with the complaints of slight pain in neck one side with deviation of chin to the right shoulder for 1 year. Initially it was occasional & mild but now the symptoms increased in 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 the normal daily life activities and makes the life miserable. Previously he was treated as a case of cervical spondylosis by doing cervical spine X-ray where mild osteophytic lipping, were seen in C4,5 and C6,7 level. On physical examination, the patients chin turns towards right shoulder & his head turns 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 and 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 and 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 manage accordingly by the drugs, physical therapy and orthosis. To relieve the pain, oral analgesic was used. Different muscle relaxant like baclofen, tizanidine etc. different antipsychotic drug like haloperidol, clonazepam etc. anti-Parkinson drug like carbamazepine, were tried to relieve muscle spasm & dystonia. Sometimes it decreases the intensity of dystonia & pain subsided. Different exercise 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. 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 focal dystonia characterized by neck muscles contraction involuntarily-causing abnormal movement and awkward posture of the head and neck. It is a rare disorder that can occur at any age even infancy.

Cervical dystonia most often occurs in middle aged people woman more than men. In most case, 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 affected⁴. 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 and strength until it reaches a plateau⁷. Symptoms can worsen during stress. Both agonist and antagonist⁵ muscle contract simultaneously during dystonic movement. Usually muscle involved contralateral sternocleidomastoid and ipsilateral splenius capitis muscle⁵. Other symptoms include muscle hypertrophy, neck pain, dysarthria and tremor Physical therapy⁸ helps with some of the movement of the head and neck. By stretching out the muscles that spasm it decreases the length and severity of the attacks. Strengthening exercise is also great for easing pain⁹⁻¹¹.

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 months¹⁻². In this case even botulinum toxin was failed. 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.

Conclusion

Physiatric intervention plays an important role by exercises that improve neck strength and 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 help a lot. The disability and 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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