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Case Report

Cervical Dystonia Caused by Chronic Nonunion C2 Fracture: A Case Report

Shangming Zhang, MD [a], Jamie Sibel, MD [a], Majid Dadgar-Kiani, DO [a], Paul Overdorf, DO [a], Donald J. Flemming, MD [b], David R. Gater, MD, PhD [c]

[a] *Department of Physical Medicine and Rehabilitation, Penn State Hershey Medical Center, Hershey, PA*
[b] *Department of Radiology, Penn State Hershey Medical Center, Hershey, PA*
[c] *Department of Physical Medicine and Rehabilitation, University of Miami, Leonard M. Miller School of Medicine, Miami, FL*

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Rehabilitation;
Spinal stenosis;
Torticollis

Abstract
Chronic nonunion cervical fracture leading to cervical dystonia (CD) is very rare. This study reports a 72-year-old man who presented with 9-month history of progressively worsening neck tilting, neck tightness, neck pain, headache, and difficulty with swallowing. The patient was referred to speech therapy and confirmed to have dysphagia on modified barium swallow study. A cervical spine radiograph showed a chronic C2 nonunion fracture. Subsequent cervical spine magnetic resonance imaging confirmed chronic C2 nonunion fracture with kyphotic deformity of the cervical canal with associated cord compression at C1-C2 and severe central canal stenosis. Needle electromyography revealed dystonic or spasmodic neck muscles, consistent with diagnosis of CD. Botulinum toxin injection resulted in marked clinical improvement. The patient finally underwent occipital to C4 posterior segmental fusion. No recurrence of CD had occurred 12 months after botulinum toxin injection and surgery, which supports the conclusion that chronic C2 nonunion fracture is most likely responsible for CD in this case. The authors suggest that all patients with CD receive dysphagia evaluation and more importantly cervical spine imaging to rule out chronic C2 nonunion fracture.

List of abbreviations: CD, cervical dystonia; CT, computed tomography; MRI, magnetic resonance imaging.
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Cervical dystonia (CD), also called spasmodic torticollis, is a neurologic syndrome characterized by sustained or intermittent involuntary muscle contractions and movements of head, neck, and shoulders associated with neck pain.[1] CD is commonly a primary idiopathic condition but can be inherited or acquired. Acquired causes include brain injury, infection, drug, toxin, vascular lesion, cancer, and psychogenic cause.[2,3] Odontoid osteomyelitis-, atlantoaxial rotatory subluxation-, and odontoid fracture-caused torticollis have been reported.[4-6] In this article, we describe the diagnosis, treatment, and satisfactory recovery of a chronic nonunion C2 fracture-caused CD and dysphagia. We hope that this article will raise awareness that chronic nonunion C2 fracture and dysphagia should be assessed in patients with CD among clinicians involved in the assessment and management of patients with CD.

Case description

A written informed consent was obtained from the patient for publication of this case. A 72-year-old white man with medical history significant for herpes encephalitis diagnosed in 2011 presented with a 9-month history of progressively worsening neck tilting, neck tightness, neck pain, and headache. The patient also reported a 1- to 2-month history of difficulty swallowing with unintended weight loss, which was characterized by coughing with meals and regurgitating through his nose. The patient reported decreased balance without weakness, sensory deficits, bowels or bladder dysfunction. The patient initially did not recall history of fall or trauma. However, on the follow-up visit, his daughter reported the patient did have a fall approximately 10 months ago. On examination, the patient's right ear was tilting to his right shoulder (fig 1A). Neck active and passive range of motion were very limited in all planes. Passive neck range of motion was painful. The patient revealed no focal weakness of the bilateral upper or lower extremities. The patient was diagnosed with torticollis and dysphagia. Because of extremely restricted range of motion, the patient was instructed to stop driving. The patient was referred to speech therapy for dysphagia. Modified barium swallow study confirmed oropharyngeal dysphagia. Given his history of herpes encephalitis, it was initially assumed torticollis was secondary to herpes encephalitis or idiopathic origin. Botulinum toxin injection was recommended. To assure cervical spine stability and tolerance to physical therapy post-botulinum toxin injection, a cervical spine radiograph was obtained and revealed cervical spine malalignment (fig 1B) and a chronic C2 nonunion fracture, which is better demonstrated on cervical spine computed tomography (CT) scan (fig 1C). A subsequent cervical spine magnetic

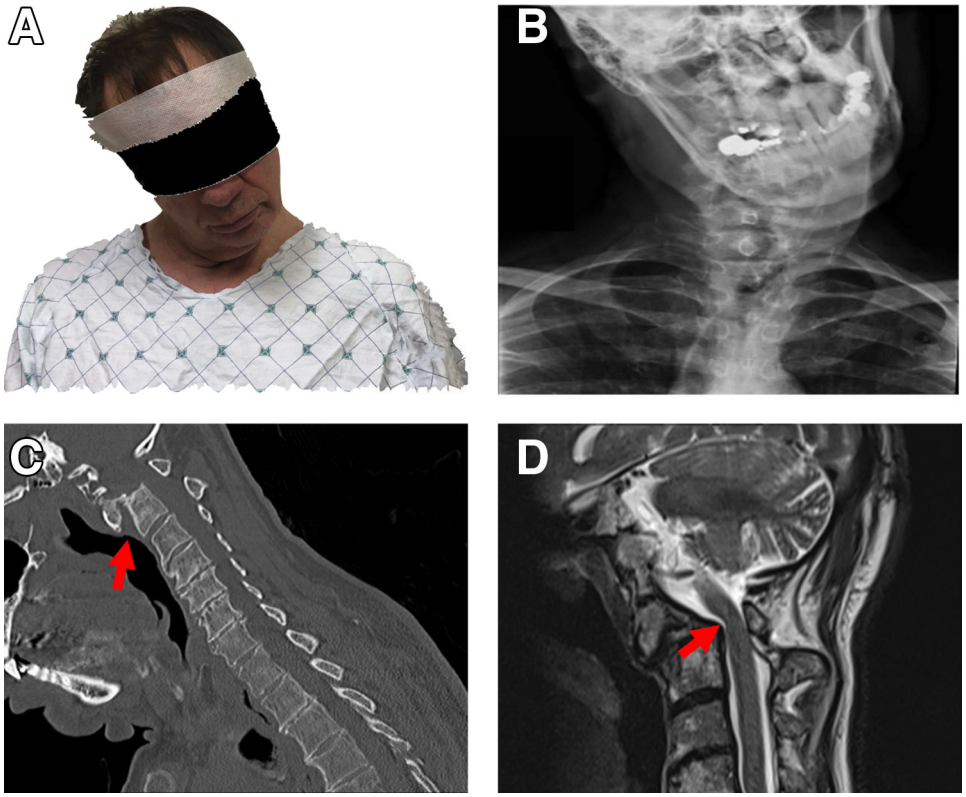

Fig 1 (A) Frontal photograph shows patient's right ear was tilting to his right shoulder with nose and face rotating to the left side, consistent with CD. (B) AP radiograph of the cervical spine shows head tilting to his right and rotating to his left. A chronic nonunion C2 fracture is not shown on this view. Abbreviation: AP, anterior posterior. (C) Midline sagittal reconstruction of cervical spine CT shows C2 fracture with anterior displacement of the dens (red arrow). C1 is anteriorly subluxed relative to C2 at the spinolaminar line with marked narrowing of the cervical canal. (D) Sagittal T2-weighted MRI image of the cervical spine MRI shows cord kinking and narrowing of the cervical canal (red arrow).

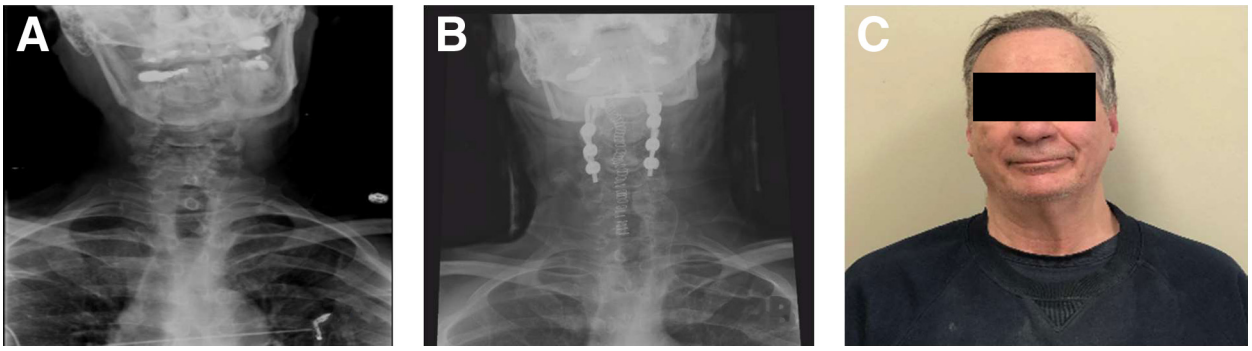

Fig 2 (A) AP radiograph of the cervical spine shows improvement in patient's head and neck rotation, 19 days post-onabotulinumtoxin A injection. (B) Postoperative AP radiograph of the cervical spine shows posterior occiput to C4 fusion with reduction of cervical and occipital malalignment. (C) Frontal photograph of the patient showed no recurrence of CD more than 12 months after onabotulinumtoxin A injection and operation. Abbreviation: AP, anterior posterior.

resonance imaging (MRI) confirmed chronic C2 nonunion fracture with kyphotic deformity of the cervical canal with associated cord compression at C1-C2 and severe central canal stenosis (fig 1D). MRI also revealed partially visualized right lateral ventricular temporal horn hydrocephalus and temporal lobe cystic encephalomalacia (imaging not shown here). The possible etiologies for CD in this case included history of herpes encephalitis with abnormal MRI, idiopathic origin and chronic C2 nonunion fracture with severe central canal stenosis, and cord compression. After consultation with the neurosurgeon, to whom we referred the patient, botulinum toxin injection was performed as planned. A total of 200 units (U) onabotulinumtoxin A were injected into the right sternocleidomastoid (75 U), splenius capitis (50 U), splenius cervicis (25 U), levator scapulae (25 U) and longissimus (25 U) muscles, using anatomic landmarks and needle electromyography or stimulation guidance. Needle electromyography confirmed dystonic or spasmodic neck muscles consistent with diagnosis of CD. The patient responded very well to the injection. His neck tilting to the right and rotation to the left were greatly improved, as seen on cervical spine radiography 19 days postinjection (fig 2A). Neck pain and dysphagia were markedly improved. The patient subsequently underwent surgical intervention for the displaced C2 fracture and C1/C2 dislocation with occipital to C4 posterior segmental fusion (fig 2B). The patient recovered well from surgery with further improved posturing of the cervical spine. Dysphagia, neck pain, and headache all resolved. The patient regained 20 lb of weight lost 3 months after surgery and subsequent resolution of dysphagia. No recurrence of cervical dystonia was appreciated 1 year after surgery (fig 2C).

Discussion

CD is the most common form of focal dystonia and characterized by sustained or intermittent involuntary muscle contractions of the neck, head, and shoulders leading to various disabling abnormal postures and movements.[7] CD has been reported to be underdiagnosed and undertreated.[7] CD patients often present with shoulder elevation, restricted range of motion, neck pain and headaches secondary to sustained abnormal postures and twisting movements. As seen in this case, dysphagia is very common in up to 36%-51.2% patients with CD.[8,9] Early diagnosis and management of dysphagia is essential for prevention of aspiration pneumonia. On the other hand, dysphagia is the most common adverse effect of botulinum toxin therapy, which is commonly used to treat CD patients. In a large double-blind, randomized, placebo-controlled clinical trial of onabotulinum toxin A, dysphagia was reported in 19% of patients post-botulinum toxin injections.[10] It is believed to occur because of regional spread of the toxin to adjacent pharyngeal muscles, particularly after injection of the sternocleidomastoid muscles.[11] Therefore, the assessment of preexisting dysphagia prior to the injection is important.

The treatment choices for patients with CD includes oral medications, botulinum toxin injection, deep brain stimulation, and surgery.[2,3,12] No oral medication has been found effective. However, botulinum toxin injection is considered a revolutionary symptomatic treatment for dystonia.[13-15] For torticollis, tilting to the right shoulder with nose and face rotating to the left side as seen in this case, right sternocleidomastoid and right splenius capitis, splenius cervicis, levator scapulae, and longissimus muscle are often injected.[16,17]

As part of a routine workup of CD, in addition to head CT scan or brain MRI, the authors suggest cervical spine imaging study to rule out cervical vertebral fracture and central canal stenosis. In addition, imaging evidence of stable cervical spine is warranted prior to physical therapy after botulinum toxin injection.

Conclusion

Chronic C2 nonunion fracture is a rare cause of CD. The authors suggest that all patients with CD receive dysphagia evaluation and, more importantly, cervical spine imaging to rule out chronic C2 nonunion fracture.

Corresponding author

Shangming Zhang, MD, Department of Physical Medicine and Rehabilitation, Penn State Hershey Medical Center, Hershey, PA 17033. *E-mail address:* szhang5@pennstatehealth.psu.edu.

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