Accepted Manuscript

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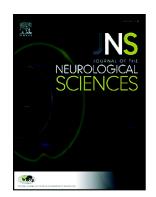
PII: S0022-510X(16)30830-9

DOI: doi: 10.1016/j.jns.2016.12.037

Reference: JNS 15029

To appear in: Journal of the Neurological Sciences

Received date: 25 August 2016 Revised date: 14 December 2016 Accepted date: 20 December 2016



Please cite this article as: Aparna Wagle Shukla, Sol De Jesus, Fan-Gang Meng, Wei Hu, Focal cervical dystonia presents in the setting of acute cerebellar hemorrhage. The address for the corresponding author was captured as affiliation for all authors. Please check if appropriate. Jns(2016), doi: 10.1016/j.jns.2016.12.037

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Focal cervical dystonia presents in the setting of acute cerebellar hemorrhage

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Running Title: Cerebellar hemorrhage and cervical dystonia

Keywords: Cerebellum, dystonia, stroke, intracerebral hemorrhage

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Highlights:

Cervical dystonia can present following an acute cerebellar hemorrhage.

Dystonia symptoms respond to botulinum toxin therapy however are resistant to oral pharmacotherapy

Cerebellar stroke is important for pathophysiology of focal dystonia.

Number of words in the manuscript body: 996

Number of Figures: 1

Number of references: 10

Dear Editor of the Journal of the Neurological Sciences:

Cervical dystonia is the most common type of focal dystonia. [1] Traditionally, pathophysiological models of cervical dystonia have focused on the basal ganglia; however other nodes in the motor network such as the cerebellum have also been found to contribute to dystonia. Abnormalities in the cerebellum have been shown in animal studies, in functional imaging and in neurophysiological studies, all collectively lending support to the role of cerebellum in dystonia. [1] Furthermore, the autopsy studies in dystonia, although rare, have shown that there are subtle changes in the cerebellum. [2] Whether these cerebellar abnormalities are causal or compensatory, is not known. It is unclear whether a combined dysfunction of the basal ganglia and the cerebellum will play a dominant role in the pathophysiology of dystonia. Alternatively it is unknown whether the cerebellum alone can trigger the symptoms of dystonia. We report a single case of cervical dystonia with mild blepharospasm presenting in the setting of an isolated acute cerebellar hemorrhage.

CASE REPORT: A 39-year-old left handed male was evaluated in the movement disorders clinic for an acute onset of neck pain and involuntary movements involving the muscles of the neck and eyes. His symptoms began soon after an acute intracerebral hemorrhage (ICH) in the right cerebellum. At the time of ICH, the patient presented to the emergency department with severe headache, drowsiness, disorientation, and a blood pressure of 240/120. The physical examination revealed a score of 13/15 on the Glasgow coma scale and there was only a slight dysmetria in the right upper extremity. The remainder of the physical examination was unremarkable. A CT scan of the head demonstrated a 3.8 x 3.0 x 2.8 cm intracerebral hemorrhage with cerebral edema, a midline shift, cerebellar herniation, and a brainstem compression. (Figure 1) Subsequently a CT angiography did not reveal any vascular anomalies or aneurysm. There was no history of anticoagulant use or cocaine abuse. The patient reported noncompliance with treatment for hypertension. Using the history and physical examination findings, he was diagnosed with an acute hypertensive ICH. Within 8 hours of onset, he underwent an emergency posterior fossa craniotomy, evacuation of the blood clot and placement of an external ventricular drain for treatment of hydrocephalus. In the two days following surgery, he noticed discomfort over the scar site that responded well to pain medications. Subsequently, within a week of onset of ICH, he began to experience a new onset of neck stiffness and pulling of muscles with involuntary turning of head to the right. His head also began to exhibit an intermittent "no-no" tremor. Additionally, he felt there was involuntary closure of the eyes and there was a tightening sensation on the right side of the face. He

endorsed improvement of the neck which pulled into specific gestures (e.g. placing a hand over the chin). (See Video) He denied abnormal movements involving the other body parts. Prior to presenting to the movement disorders clinic, he tried using a soft collar for neck stiffness without clinically significant relief of his symptoms. A follow-up CT scan was performed at six weeks and it revealed resolved hydrocephalus and hemorrhage reduction. During the physical examination in our movement disorders clinic, the involuntary turning of the head was noted to be more pronounced during walking. He was provided a diagnosis of cervical dystonia which developed in the setting of an acute cerebellar hemorrhage. He underwent several treatment trials for neck and facial symptoms. He was first prescribed a trial of baclofen at a dose of 15mg three times/day and clonazepam at a dose of 1mg/day. There were no noticeable clinical improvements with these medications. He then underwent a trial with onabotulinum toxin injections at escalating doses. The third attempt at injections revealed that at 250 units, he began to endorse clinical improvements.

DISCUSSION: Although cerebellum is considered an important contributor to the pathophysiology of dystonia, there are a few case reports and retrospective case studies that have specifically implicated cerebellar strokes/hemorrhages on imaging as related to dystonia. [3-7] In the available reports, dystonia was observed to be focal and involving the craniofacial region (cervical dystonia or oromandibular dystonia). The dystonia symptoms in these reports presented nearly 15 months to 40 years after the cerebellar stroke, and this timing could lead to doubt about the etiological role of cerebellum in dystonia pathogenesis. [6,8] Furthermore although 3/188 cases of cervical dystonia had cerebellar infarcts, the study was retrospective and the causal relationship between cerebellar lesions and dystonia could not be determined. [9] The remaining cases reporting cerebellar abnormalities in dystonia included slow growing tumors. [7,10] The tumors frequently involved other brain structures (e.g. brainstem), therefore it would be difficult to definitely determine whether cerebellum was a primary contributor to the movement disorder.

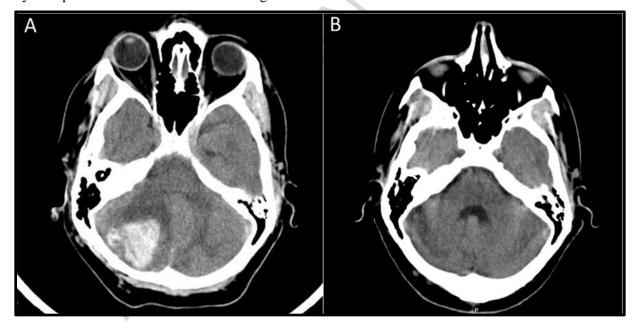
Our patient, who developed symptoms of cervical dystonia within a week of onset of a cerebellar hemorrhage, raises two important findings. First, a clear temporal relationship between the onset of ICH and the dystonia strongly suggests that cerebellum alone could trigger the dystonia. This finding suggests that cerebellum had a causal, and not a compensatory role. Second, cerebellar hemorrhage induced focal dystonia in our case. The pathophysiology of focal dystonia is usually distinct from generalized and segmental dystonias and therefore this finding is interesting. Further, our case shared similar features with previously reported focal dystonia cases and in particular and consistent was the direction of head turning was ipsilateral to the side of the hemorrhage.[3] Additionally, and similar to a previous report, our patient

did not respond to oral dystonia medications but did show improvements once the dosage for botulinum toxin was optimized.[5]

In summary, dystonia involving the craniofacial region should be included in the clinical profile of cerebellar stroke or hemorrhage. Although our case suggested a causal relationship based on the temporal course, longitudinal follow-up studies will be required in a larger stroke and hemorrhage population. Future studies should focus on the cerebellar region which may help us to understand the pathophysiology of focal dystonia and may also be a potential target for future therapies.

Legend

Figure 1. (a) A head CT revealed intracerebral hemorrhage (3.8 x 3.0 x 2.8 cm) with cerebral edema in the right cerebellum, cerebellar herniation and a brainstem compression. (b) A follow up head CT scan (40 days later) revealed postsurgical changes from suboccipital craniectomy and hematoma evacuation and there was mild encephalomalacia involving the right cerebellar hemisphere. In addition, the hydrocephalus resolved and the hemorrhage volume was reduced.



Legend of video

This video highlights the cervical dystonia with head turning to the right and tilting to left. There was intermittent irregular "no-no" head tremor, and the dystonia partially improved when touching the chin

(sensory trick). He has mild involuntary closure of eyes. The cervical dystonia was increased during walking.



Still photograph of the patient

The patient signed informed consent to be filmed for publication on line.

Conflict of Interest Statement:

The authors declare there is no conflict of interest.

Acknowledgement: We would like to thank NIH K23 NS092957-01A1 (AWS) and the American Brain Foundation (WH) as well as Tyler's Hope Foundation for Dystonia Cure. Other authors have nothing to disclose regarding conflicts of interest or commercial relationships including grants, honoraria, speaker's lists, significant ownership, or financial support from pharmaceutical or other companies

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