

## Bilateral Cerebellar Stroke Presenting with Acute Dysphonia and Late Palatal Tremor

Richard A. Walsh,<sup>1</sup> Patricia Gillivan-Murphy,<sup>2</sup> Ciara Murphy,<sup>2</sup> Michael Colreavy,<sup>3</sup> Killian O'Rourke,<sup>1</sup> and Timothy Lynch<sup>1\*</sup>

<sup>1</sup>Dublin Neurological Institute, Mater Misericordiae University Hospital, Dublin, Ireland

<sup>2</sup>Speech and Language Therapy Department, Mater Misericordiae University Hospital, Dublin, Ireland

<sup>3</sup>Department of Otolaryngology, Mater Misericordiae University Hospital, Dublin, Ireland



A 74-year-old woman presented with acute dysphonia and ataxia. Her husband reported that she had been asymptomatic with no speech abnormality immediately prior to the event. She had no significant past medical history nor prior exposure to neuroleptic medications. There was no family history of dystonia. She had been a lifelong and accomplished choir singer, practicing daily for at least an hour.

Fast atrial fibrillation was noted during her initial assessment, requiring diuresis for pulmonary congestion and metoprolol for rate control. Speech was mostly incomprehensible with a strained-strangled quality, slow rate, occasional voice arrests, and inappropriate pauses. Of note, phonation in the context of laughter was relatively preserved. The clinical impression was of a spasmodic dysphonia (SD) with additional features of an ataxic dysarthria. Examination of the tongue and palate was initially unremarkable. Tone, power, and reflexes were normal throughout. There was mild appendicular intention tremor with dysmetria but severe truncal ataxia causing retropulsion, even when seated.

### Investigations

Bilateral cerebellar hypodensities consistent with acute infarction were seen on computed tomography (CT) performed on the day of admission. One week after presentation, magnetic resonance imaging (MRI) of brain identified bilateral hyperintensities involving the superior cerebellar arterial territories, including the dentate

nuclei outflow tracts (Fig. 1). Restricted diffusion consistent with subacute infarction was evident on diffusion-weighted imaging (DWI). No acute brainstem or basal ganglia lesions were seen, although a small focus of hyperintensity was noted in the ventral medulla on sagittal T2 but not DWI sequences. Bilateral scattered deep white-matter hyperintensities were seen without restricted diffusion, consistent with established infarcts. Dopamine transporter imaging looking for evidence of secondary nigrostriatal dysfunction was normal and <sup>18</sup>F-DG positron emission tomography (PET) imaging did not demonstrate any evidence of altered metabolism affecting the basal ganglia, thalamus, or cortical structures as a consequence of the cerebellar infarcts.

Voice assessment found that on a sustained "ah" vowel task, her voice was forced and strained to the extent that it was almost aphonic with intermittent bursts of breakthrough sound (see Video and Supporting Fig. 1). Fiber optic laryngoscopic evaluation of the vocal cords at presentation was normal at rest and no tremor was evident. Marked supraglottic constriction on voicing was noted, characterized by adduction of the ventricular (false) cords to the midline, almost completely obliterating the view of the vocal cords. These features were still present in addition to a new palatal tremor on repeat examination five months later. She received intensive speech therapy on the acute stroke unit. Following a 6-week program there was no functional improvement in her effortful, strained vocal quality. She was later treated with botulinum toxin. One thyroarytenoid muscle was injected with definite improvement in her ability to maintain vowel sounds and evidence of less effortful speech.

### Discussion

We postulate that this patient had acute SD due to bilateral lesions of the dentato-rubro-thalamic pathway and delayed palatal tremor due to involvement of the dentato-rubro-olivary tract (Guillain-Mollaret triangle). The cerebellum and basal ganglia are involved in motor control via functional loops with common cortical and subcortical targets in the frontal cortex and thalamus.

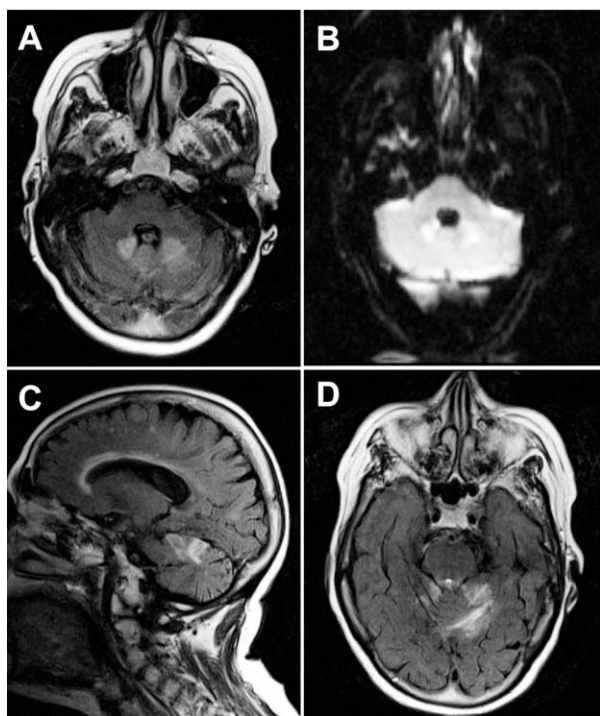
Additional Supporting Information may be found in the online version of this article.

\*Correspondence to: Dr. Timothy Lynch, The Dublin Neurological Institute, Mater Misericordiae University Hospital, Eccles Street, Dublin 7, Ireland; tlynch@mater.ie

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**FIG. 1.** MRI imaging performed 1 week after presentation. **A:** Hyperintense signal in the superior cerebellar artery territory bilaterally on FLAIR. **B:** Diffusion weighted imaging in the same region (ADC map not shown). **C:** FLAIR hyperintensity in superior left cerebellar hemisphere. **D:** Corresponding axial FLAIR image. ADC, apparent diffusion coefficient; FLAIR, fluid-attenuated inversion recovery; MRI, magnetic resonance imaging.

Both structures may have separate or interacting roles in the pathogenesis of dystonia.<sup>1</sup> Evidence from animal models points to reciprocal polysynaptic connections between these structures that may interact in the genesis of dystonia.<sup>2</sup> There is a growing literature to support a role for the cerebellum in the etiology of primary dystonia, including spasmodic dysphonia.<sup>3</sup> Animal models of primary dystonia can improve with cerebellectomy, which suggests that dysfunctional cerebellar outputs may be causative.<sup>1</sup> Conversely, secondary dystonia occurs acutely following cerebellar stroke, presumably via disruption of normal cerebellar interaction with the basal ganglia and cortical structures.<sup>4</sup> Palatal tremor is associated with lesions of the Guillain-Mollaret triangle, affecting projections from the dentate nucleus to the contralateral red nucleus or descending fibers from the red nucleus to the inferior olive that can then become hypertrophied. Follow-up MRI imaging at 6 months did not, however, reveal olivary hypertrophy in our patient.

The main differential diagnosis to consider is muscle tension dysphonia (MTD), a term used to describe a functional speech disorder associated with a spectrum of vocal symptoms related to excessive tension of the (para)laryngeal muscles. It can be difficult to discriminate between SD and MTD. The coexistence of the 2 disorders can arise as MTD can be seen as a secondary compensatory phenomenon in response to any vocal cord pathology. MTD can also be caused by psychological factors or maladaptive changes with overuse.

The acute onset of maximal and persistent dysphonia at the time of stroke onset in this patient makes a purely compensatory etiology unlikely. We had no suspicion of a psychological component. As a high-level performer it could be said that she had a lifetime of vocal “overuse” and how this background might have interacted with her acute cerebellar injury is unknown. Therefore, we believe that MTD does not explain the acute nature of her laryngeal hyperkinesia (or indeed tremor) but certainly could represent a later compensatory phenomenon for laryngeal dystonia.

Adductor spasm of the vocal cords could not be confirmed on laryngeal examination, although this is not always of discriminating value when considering SD or MTD. Some additional clinical features can sometimes assist.<sup>5</sup> SD is more likely to be task-specific with relative preservation of emotive phonation (eg, laughing or crying) and typically will not respond to speech therapy, both features observed in this patient. SD tends to be restricted to the vocal cords but supraglottic variants exist and the clinical spectrum of secondary laryngeal dystonia is unknown. Acoustic analysis has been proposed as a useful discriminating factor, with the presence of frequent acoustic breaks being more suggestive of a dystonic etiology than MTD.<sup>6</sup> Botulinum toxin can improve both SD and MTD. The therapeutic response in this patient was to a unilateral thyroarytenoid muscle injection and not the supraglottic larynx, thus favoring a diagnosis of laryngeal dystonia as the principal pathology.

Dystonia typically develops weeks to years after stroke onset and is most commonly associated with putaminal infarction. The acute onset of vocal disturbance in this patient is also atypical for dystonia; however, acute cervical dystonia has been reported 1 and 3 days after cerebellar stroke.<sup>4,7</sup> Our understanding of the pathophysiology of “cerebellar dystonia” and its temporal relationship to acute lesions is currently limited.

In conclusion, we propose the primary phenomenology in this patient to be dystonic. We cannot rule out an element of compensatory MTD in response to the acute laryngeal dysfunction and there are additional features of ataxic dysarthria. While the precise underlying neural substrate in primary focal dystonia remains unknown, clinicoradiological correlation in secondary cases such as this will remain important in attempting to reveal the importance of structures outside the basal ganglia.

## Legends to the Video

### Speech Assessment and Fiber Optic Laryngoscopy

**Video Segment 1.** Speech is strained and strangulated with telescoping of syllables, distorted and prolonged vowels and articulatory errors. Verbal dysdiadochokinesia on performing ooo-eee; p-p-p-p; p-t-k.

**Video Segment 2.** At laryngoscopy, phonation when laughing and impersonating the local accent is considerably better. During normal speech extreme hyperadduction of the supraglottic larynx is then seen.

**Video Segment 3.** Palatal tremor was noted 5 months later.

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## Commentary on “Bilateral Cerebellar Stroke Presenting with Acute Dysphonia and Late Palatal Tremor”

Kristina Simonyan, MD, PhD<sup>1\*</sup> and Andrew Blitzer, MD, DDS<sup>2</sup>

<sup>1</sup>Departments of Neurology and Otolaryngology, Mount Sinai School of Medicine, New York, New York, USA

<sup>2</sup>Department of Otolaryngology, Columbia University College of Physicians and Surgeons and New York Center for Voice and Swallowing Disorders, New York, New York, USA

The case report by Walsh et al described the development of acute spasmodic dysphonia (SD) and late palatal tremor secondary to cerebellar stroke. The patient was presented with hyperfunctional, hyperadductive voice disorder affecting the supraglottis, including the ventricular (false) cords. Speech was incomprehensible with a strained-strangled quality and occasional voice arrests. The diagnosis of SD was made based on these speech symptoms, hyperfunctional supraglottis visualized with fiberoptic laryngoscopy, the lack of benefits from voice and speech therapy, and some improvement of voice problems following unilateral botulinum toxin injection into the thyroarytenoid muscle.

As such, this is an atypical SD case for several reasons. Laryngeal spasms in SD primarily affect the (true) vocal cords rather than the ventricular cords, although supraglottic hyperadduction involving ventricular cords may also be present. Although SD patients have problems with coordinating voice onset and offset during connected speech because of SD-specific voice breaks, they usually are able to sustain sound, and their speech remains comprehensible. The lack of response to speech therapy and the benefits from botulinum toxin injections into the laryngeal muscles are typical but not specific to

SD only. On the other hand, speech therapy is beneficial for patients with supraglottic hyperadduction, whereas persistent supraglottic squeeze may be managed more successfully with botulinum toxin injections into the supraglottic musculature.<sup>1</sup>

This case further suggests that cerebellar lesion without functional, structural, or dopaminergic abnormalities in any other brain regions may cause SD. Conversely, previous studies have shown relief of dystonia symptoms following cerebellar lesions and cerebellectomy,<sup>2,3</sup> and SD has been characterized as a brain network disorder involving abnormalities in the basal ganglia, thalamus, sensorimotor cortex, and cerebellum.<sup>4,5</sup>

In conclusion, the diagnosis of secondary SD in this patient was complicated by an unusual clinical presentation, atypical diagnostic criteria, and the presence of cerebellar pathology without abnormalities in other dystonia-related brain regions. The differential diagnosis is likely to include other cerebellar disorders such as spinocerebellar atrophy, other movement disorders such as chorea, and muscle tension dysphonia, as a compensatory voice disorder secondary to cerebellar pathology. ■

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\*Correspondence to: Dr. Kristina Simonyan, Department of Neurology, One Gustave L. Levy Place, Box 1137, New York, NY 10029, USA; kristina.simonyan@mssm.edu

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