Movement Disorders
CLINICAL PRACTICE

CASE REPORT

Lingual and Facial Myoclonus Associated with Early Corticobasal Syndrome

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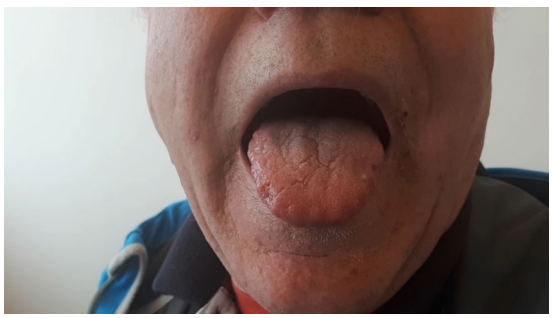
The early clinical features of corticobasal syndrome (CBS) may include limb myoclonus, occurring in approximately 15%.[1] Isolated lingual dystonia was recently reported as a new clinical feature heralding early CBS,[2] and lingual dyskinesia is also a recognized phenomenon in CBS.[3] Here, we add a case with isolated lingual and facial myoclonus that we hypothesize as a potential sole initial manifestation of CBS for more than 3 years before other features emerged.

Case Report

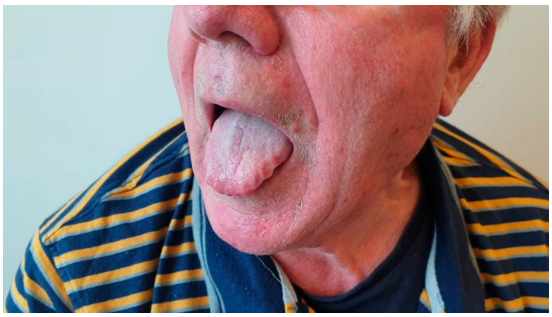
A 66-year-old right-handed man presented with a 15-month history of abnormal tongue and facial movements and dysarthric speech. He felt no compulsion to perform the movements and no sensory trick, and they could not be suppressed. He had no history of epilepsy and no other neurological symptoms. Medical history was significant for duplex kidneys, dyslipidemia, hypertension, and obstructive lung disease. He was taking rosuvastatin and valsartan and was using a bronchodilator. He had never taken neuroleptics. Family history was unremarkable.

On examination, there were brief, spontaneous, nonrhythmic, left-sided lingual and facial movements. These heightened in frequency with speech and volitional facial activation (Video 1). In addition, although not well captured on video, we observed that the intermittent lingual flickers often synchronized with flickers on the face. Tongue power and muscle bulk were normal. The remaining cognitive and neurological examinations were normal.

Initial investigations were unilluminating, including normal/negative hematological–biochemical profiles, iron, ferritin, copper, ceruloplasmin, and autoantibodies (anti-nuclear antibody, extractable nuclear antigen antibodies, anti-neutrophil cytoplasmic antibody, anti-thyroid peroxidase antibody, classic onconeuronal



Video 1. Patient at 15 months from symptom onset, demonstrating brief, spontaneous, nonrhythmic, left-sided predominant lingual and facial movements that appeared to be stimulus sensitive, along with a slight dysarthria.
Video content can be viewed at https://onlinelibrary.wiley.com/doi/10.1002/mdc3.13861



Video 2. Patient at 3½ years from symptom onset with worsening lingual and facial movements and dysarthria. Furthermore, he has developed left upper limb small amplitude myoclonic jerks along with rigidity, bradykinesia, and abnormal ideomotor limb apraxia on the same side.
Video content can be viewed at https://onlinelibrary.wiley.com/doi/10.1002/mdc3.13861

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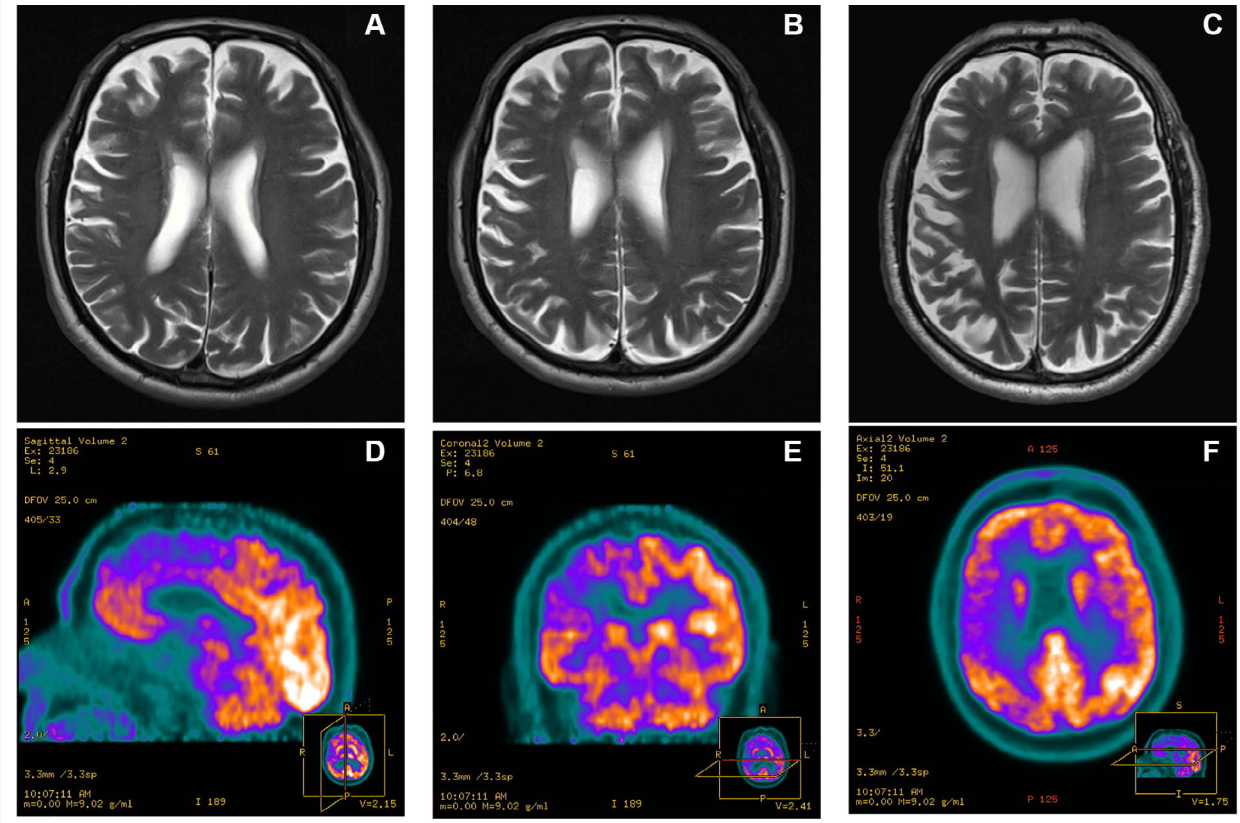

Fig. 1. Representative T_2-weighted magnetic resonance imaging brain axial cuts at initial outpatient presentation (**A**), 18-month follow-up (**B**), and 36-month follow-up (**C**) show subtle progressive asymmetric brain atrophy, predominantly affecting the right frontal and parietal lobes, and positron emission tomography–computed tomography of the brain after the 36-month follow-up with representative sagittal (**D**), coronal (**E**), and axial (**F**) views shows asymmetrical cortical metabolism with reduced uptake in the right frontal and parietal cortices and in the basal ganglia in keeping with corticobasal syndrome.

antibody panel, and antibodies to glutamic acid decarboxylase-65, glycine receptor, leucine-rich glioma-inactivated 1, and contactin-associated protein-like 2). Genetic testing for Kennedy's spinal and bulbar muscular atrophy was negative. Initial brain magnetic resonance imaging (MRI) showed scattered nonspecific white matter changes felt unremarkable for age. Electromyography showed very short duration spontaneous bursts of motor units in the mentalis, orbicularis oris, and tongue. These were myoclonic (unlike myokymia) and involved the tongue (unlike hemifacial spasm). Routine electroencephalogram (EEG) did not identify any focal or epileptiform abnormalities, and there was no correlate for the abnormal movements.

During a further 18 months of close clinical observation, the lingual and facial myoclonus remained an isolated issue although accompanied by progressively worsening dysarthria. The movements were unresponsive to successive trials of carbamazepine, baclofen, and levetiracetam.

By 24 months from initial presentation, however, he reported significant deterioration of the abnormal movements and dysarthria, along with difficulties with repetitive and coordinated tasks using his left upper limb. On examination, the lingual and facial myoclonus had worsened, and his speech had become profoundly dysarthric. Whereas initially the dysarthria represented speech interruption by myoclonus, now additional apraxia of speech was observed. In addition, he now had small-amplitude, stimulus-sensitive myoclonus of the left upper limb; rigidity, bradykinesia, and pathologically brisk reflexes on the left; and abnormal bedside testing for praxis (ideomotor limb apraxia and ideational apraxia)[4] and agraphesthesia on the left (Video 2).

Follow-up brain MRI showed progressive asymmetric atrophy predominantly affecting the right frontal and parietal lobes, and positron emission tomography–computed tomography of the brain showed asymmetric cortical and basal ganglia hypometabolism (Fig. 1).

At this point, he was diagnosed with probable CBS (and possible clinical corticobasal degeneration) as per the Armstrong et al criteria.[1]

Discussion

There have been no previous reports of lingual myoclonus as an early sole manifestation of neurodegenerative conditions or CBS.

CBS has been associated with orobulbar and speech issues at presentation in up to 40% of cases, but these tend to involve apraxia of speech,[1] often described as slow, effortful, and/or groping. Other movement disorders of the tongue have been previously described in CBS, specifically lingual dyskinesia[3] and recently lingual dystonia.[2]

In our case, we hypothesize that the dysarthria and abnormal tongue movements seen represent a cortical myoclonus[5–7] given the nonrhythmic, jerky, stimulus sensitivity observed (worse with movement and with speech). Owing to cortical generation, there is also synchronous involvement of the nearby face observed clinically. Back-averaging on neurophysiology may have allowed a cortical correlate to be identified, however, in practice this can be technically difficult and was unavailable.[5,6] We cannot definitively exclude the possibility that the isolated lingual and facial movements were coincidental with the later separate emergence of CBS.

Lingual myoclonus is itself a very rare phenomenon, to date described sporadically as either a primary essential phenomenon, often rhythmic,[8,9] or as a secondary phenomenon, including various cortical and subcortical causes.[10–12] The lack of response to antiseizure medications in our case is noteworthy, as previously reported cases of lingual myoclonus of other etiologies have responded at least in part to, for example, clonazepam,[8,12] valproate,[9] lacosamide, levetiracetam, phenytoin,[10] and topiramate.[11]

Lingual myoclonus could be mistaken for tongue fasciculations, palatal tremor, or epilepsia partialis continua. The asymmetry, the simultaneous facial myoclonus, and the stimulus-sensitive dysarthria may be helpful for differentiation. Individuals with lingual myoclonus warrant longitudinal follow-up for the emergence of CBS.

Author Roles

(1) Research Project: A. Conception, B. Organization, C. Execution; (2) Manuscript Preparation: A. Writing of the First Draft, B. Review and Critique.

M.M.O.: 1A, B, C, 2A

S.C.: 1A, B, C, 2B

Disclosures

Ethical Compliance Statement: The authors confirm that the approval of an institutional review board was not required by our University College Cork for this work given its nature. Informed verbal and written consent was obtained for this article from the patient as per the Journal's guidelines. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

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