Movement Disorders
CLINICAL PRACTICE
CASE SERIES WITH LITERATURE REVIEW

Chorea and Levodopa-Induced Dyskinesia in Corticobasal Syndrome: Two Case Reports with Pathological Insights and Literature Review

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Background: Corticobasal syndrome (CBS) is a rare, clinically heterogeneous form of atypical Parkinsonism. Hyperkinetic movements, aside from myoclonus and dystonia, have rarely been reported in CBS.
Cases: We present two patients with CBS, one with pathologically confirmed corticobasal degeneration (CBD) and generalized chorea, and another with probable CBS and Levodopa-induced dyskinesia (LID). Case 1 exhibited late-onset generalized chorea, which was preceded by several years of dystonia, rigidity and apraxia affecting the right upper limb. Case 2 presented with dystonia, cortical sensory loss, and apraxia in the left upper limb, while LID affected the face and the right side of the body.
Literature Review: A systematic review of published cases of chorea or levodopa-induced dyskinesia (LID) in CBS was performed. The literature search was executed in PubMed from its inception for cases of chorea or LID associated with CBS. Twelve patients were identified across eight studies. Only five cases of pathologically confirmed CBD with chorea were found; chorea developed after 4 years of disease progression.
Conclusions: Chorea and LID are rare but may represent late manifestations of CBS. The exact mechanisms are unclear; they may relate to variability in anatomical involvement, particularly relative sparing of the GPi. Greater understanding of topographical disease progression may improve diagnostic precision and phenotypic classification in CBS and related tauopathies.

Corticobasal Syndrome (CBS) is characterized by asymmetric levodopa-resistant parkinsonism, apraxia, cortical sensory loss, alien limb phenomenon, cortical myoclonus, and focal dystonia.[1] Common overlapping phenotypes include frontal behavioral-spatial syndrome, nonfluent/agrammatic variant of primary progressive aphasia, and progressive supranuclear palsy syndrome.[1–4] The 4-R tau pathology of corticobasal degeneration (CBD) accounts for between 25 and 50% of CBS cases; other pathologies, particularly progressive supranuclear palsy (PSP) and Alzheimer's disease, account for the remainder.[5] Apart from CBS, the clinical presentations of 4R-tauopathies show considerable overlap, suggesting that the phenotypic spectrum reflects variability in anatomical distribution.[6] For instance, PSP-Richardson syndrome has been observed with CBD pathology.[7]

Hyperkinetic movements other than myoclonus or dystonia, such as oral stereotypies, have been occasionally reported in CBS.[8] Levodopa-induced dyskinesias (LID) have been rarely described in FTD-related atypical parkinsonian syndromes, mainly in PSP.[9,10] In the first description of CBD, Rebeiz et al reported three cases with the main commonality of a similar age group and asymmetric limb "slowness, awkwardness, and clumsiness."[11] However, the authors stated that two developed

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involuntary, uncontrollable movements. In the discussion, J. Clifford Richardson (Toronto, Canada) suggested that the movements were "choreic" in one of them.[11]

Here, we present two cases of CBS with unusual hyperkinetic movements: one pathologically confirmed CBD patient with generalized chorea, and a CBS patient with LID. Additionally, we conducted a systematic literature review of chorea and LID in CBS to identify similar cases.

Case Series

The two patients were evaluated at the Moncton Interdisciplinary Neurodegenerative Diseases Clinic, New Brunswick, Canada. Informed consent for this report was obtained from each. Probable CBS was defined according to the established diagnostic criteria.[1] The case of pathologically confirmed CBD was determined using neuropathological criteria[12] by a neuropathologist (third author, MR) at Centre de Recherche de Québec, Université Laval.

Case 1

An otherwise healthy 65-year-old French Canadian female, with no relevant family history, displayed a tightness and pressure sensation around her jaw, accompanied by slurred speech. One year later, she developed difficulty opening her mouth, almost simultaneously with abnormal posturing of her right arm, hand, and neck, and reported that her right hand felt useless. Physical examination demonstrated right limb apraxia, rigidity, and segmental dystonia affecting the jaw, neck, and right arm. Subsequently, cognitive impairment became apparent to the family members. A neuropsychological assessment identified difficulties with attentional control, working memory, and naming, with intact functioning in word knowledge and verbal abstract reasoning (MoCA 19/30). After approximately 6 years, large, generalized amplitude movements emerged, accompanied by postural instability and falls. The patient was never exposed to dopamine receptor blocking agents or other dopaminergic drugs. On examination, generalized chorea was observed, combined with segmental dystonia, bilateral hand apraxia, cortical sensory loss, jerky ocular horizontal pursuit, and severe vertical ophthalmoparesis (Video 1). The brain MRI revealed nonspecific symmetric generalized atrophy and white matter hyperintensities (Fig. 1). Genetic testing for Huntington's disease (16 and 17 trinucleotide repeats), spinocerebellar ataxia type 17 (ATX-*TBP*), dentatorubral-pallidoluysian atrophy (ATX-*ATN1*), Huntington's disease-like-2 (CHOR-*JPH3*), and a frontotemporal dementia (FTD) panel were negative. Antiphospholipid antibodies were negative, and there was no polycythemia or thrombocytosis. Metabolic studies, vitamin B12, and thyroid-stimulating hormone levels were normal. Multiple blood smears did not show acanthocytes, and serum IgLON 5 antibodies were negative. One year later, the patient was wheelchair-bound, had increasing swallowing difficulties, and passed away from aspiration pneumonia.

Video 1. The first case during two separate visits. In the first segment, generalized chorea, segmental dystonia of the jaw, cervical region, and right upper limb, along with bilateral hand apraxia, are observed. In the final portion of the first segment, severe reduction of vertical and horizontal eye movements is observed. In the second video, generalized chorea is again observed, along with severe akinesia and dystonia of the right upper limb. She was able to ambulate with a mild, wide-based gait and reduced cadence.
Video content can be viewed at https://onlinelibrary.wiley.com/doi/10.1002/mdc3.70420

Neuropathological findings were consistent with CBD (Fig. 2). There was mild gyral flattening across all lobes, mild loss of pigmentation in the substantia nigra with no Lewy bodies detected on routine sections, significant gliosis, and neuronal loss observed in the parietal cortex, along with ballooned neurons in the cingulate gyrus. Mild vacuolization was seen in the white matter, with some collagenization of blood vessels in the basal ganglia and thalamus. The p62 and tau (AT8) immunostains revealed dense cortical neuronal (pre-tangles) inclusions along with thread-like glial pathology in both the cortex and white matter of the frontal, parahippocampal, and insular regions, and astrocytic plaques were identified. Very dense tau thread-like

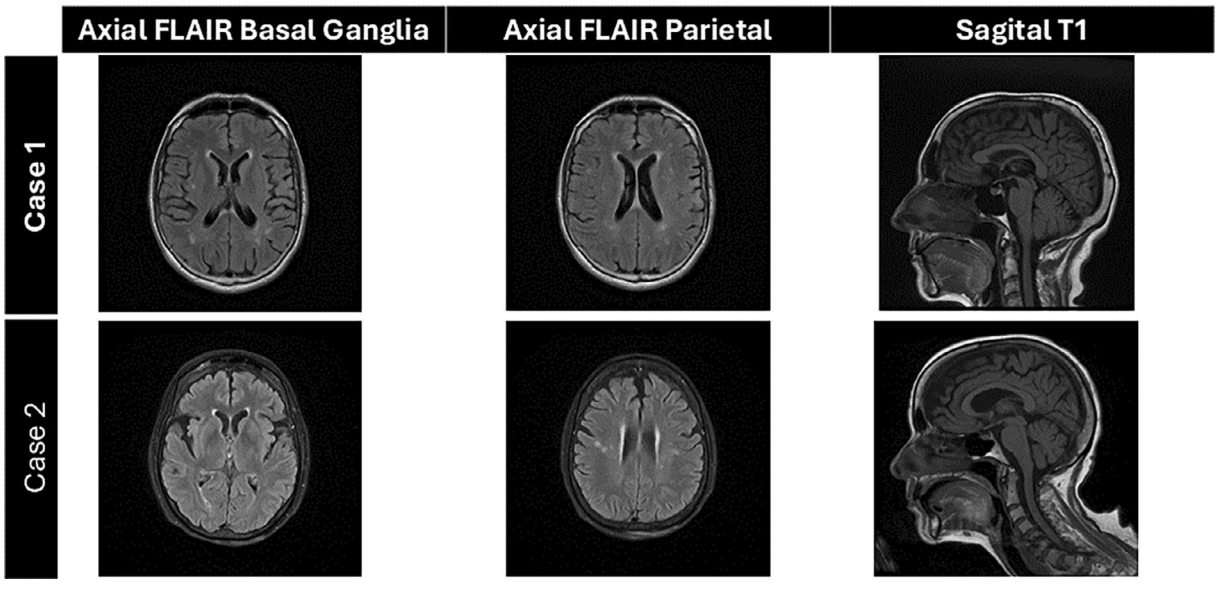

Figure 1. 1.5-T Magnetic Resonance Imaging (MRI) axial fluid-attenuated inversion recovery and sagittal T1 displaying in both cases nonspecific symmetric generalized atrophy and white matter hyperintensities.

pathology was detected in the putamen, subthalamic nucleus, and amygdala, with significant involvement of the globus pallidus and thalamus also seen. Beta-amyloid plaques were confined to the middle and superior temporal gyri. TDP-43 and alpha-synuclein stains were negative.

Case 2

A 67-year-old English Canadian female, with no relevant family history, complained of rigidity and reduced mobility of the left body, leg first, and insidiously over less than a year, her arm became affected. After approximately 6 months, her left hand adopted a fixed posture (wrist and finger flexion), and she complained that the hand was unusable. The left foot dystonia limited her mobility. Levodopa-carbidopa 800 mg/day had provided no clear clinical improvement. On physical examination, the patient had reduced comprehension, attention, and bradyphrenia. She also displayed some difficulties with saccade initiation, both horizontal and vertical, yet with normal velocity and complete horizontal and vertical range. She had apraxia and astereognosis in the left hand. A neuropsychological assessment identified multi-domain difficulties, including visuospatial, attention, short-term memory, and orientation (MoCA 20/30). The brain MRI (Fig. 1) showed nonspecific generalized atrophy and scattered white matter hyperintensities. Negative or normal studies included genetic testing for ATX-*ATN1* and an FTD panel, which included expansion for the C9orf72 gene and MAPT, paraneoplastic, and autoimmune encephalitis panels, including IgLON5, vitamin B12, p-tau217, thyroid-stimulating hormone, and other metabolic workups.

After approximately 18 months of starting levodopa-carbidopa treatment, she developed choreic movements of the right hemibody, more prominent in the lower extremity, also with forceful contractions of the eyes and jaw (Video 2). The abnormal movements lowered when levodopa-carbidopa was progressively reduced to 300 mg/d and did not return after levodopa was discontinued (Video 2). At the time of this reporting, approximately 7 years after symptom onset, the patient is still alive in a nursing home.

Literature Review

The literature search strategy was conducted in PubMed and was created with the assistance of an experienced librarian. There were no time or language restrictions, and it was executed on June 4, 2025. Controlled vocabulary and terms searching for chorea, "OR" dyskinesia, "OR" hyperkinetic were combined with corticobasal (syndrome or degeneration) plus additional tauopathies-related vocabulary, including corticodentatonigral degeneration with neuronal achromasia and cortical basal ganglionic degeneration. The complete search strategy can be found in Supplementary Material S1.

The citations were uploaded to the Covidence platform for title/abstract screening and full-text review stages were done independently by two authors (first and second authors, AME and second author MR). Abstracts were included if they reported chorea, dyskinesia, or hyperkinetic movements as a manifestation of any tauopathy. In the full-text review stage, studies were

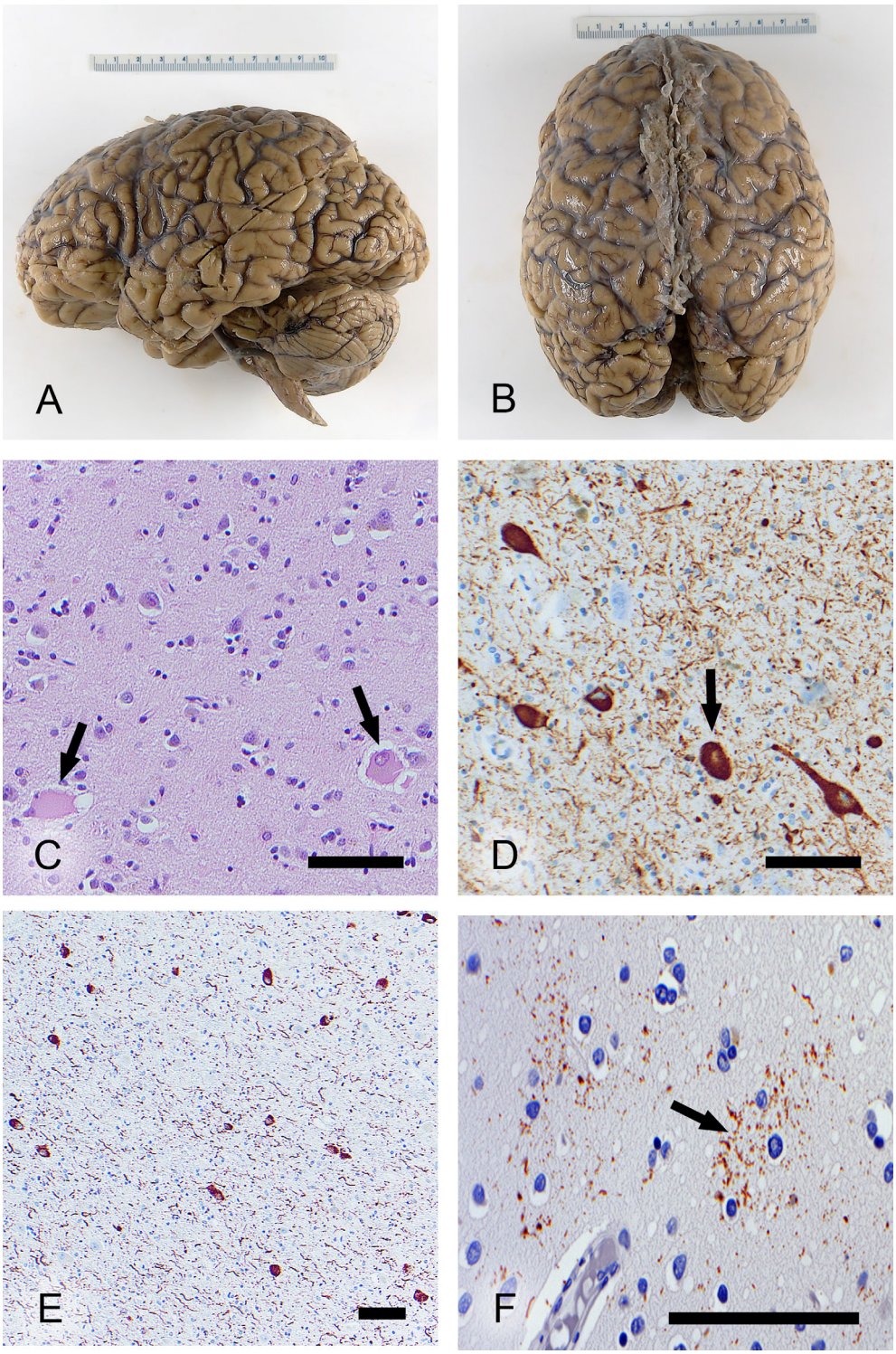

Figure 2. Neuropathological findings of Case 1: (A) Lateral view of the left cereberal hemisphere. (B) Superior view of the brain showing mild gyral flattening and sulcal narrowing across all cerebral lobes without any asymmetry. (C) Cingulate gyrus showing ballooned neurons (arrow), gliosis and neuronal loss. (D) Substantia nigra with extensive thread-like pathology, numerous pretangles (arrow), and sparse oligodendroglial coiled bodies. (E) Dense tau thread-like pathology in the subthalamic nucleus. (F) Dense tau pathology in the cerebellum, including pretangles and astrocytic plaques (arrow), along with numerous pretangles in the granule cell layer of the dentate gyrus. The magnification bar in image C-F represents 100 μm.

Video 2. The second case during four different assessments. In the first segment, the patient is on 300 mg of levodopa and exhibits dyskinesia affecting the right side of her body, as well as the upper and lower parts of her face. The second segment captures a separate admission, showing a similar pattern of dyskinesia but with slightly more prominence in the upper and lower face while on 600 mg of levodopa. The third segment was recorded approximately 4 years after the onset of her symptoms, showing abnormal posturing on the left lower extremity. In the final segment, the patient is off levodopa and manifests apraxia and dystonia in her left hand, alongside slow initiation of horizontal and vertical pursuit movements, but with a normal range.
Video content can be viewed at https://onlinelibrary.wiley.com/doi/10.1002/mdc3.70420

selected if CBS or CBD were described with chorea, dyskinesia, or hyperkinetic movements.

Systematic Review Results

The flow chart of the literature systematic review is depicted in Figure S1. A total of eight studies published between 1989 and 2009 contained data on hyperkinetic movements in 12 patients with CBS or CBD,[13–20] of which seven patients presented with chorea, three with unspecified hyperkinetic movements, and two had LID. In total, only five out of 12 had a pathological confirmation of CBD. There was limited clinical information; however, two of the pathologically confirmed cases displayed chorea after 4.5 years. In addition, two cases were reported to have co-pathologies: Huntington's Disease (HD), with 40 repeats in one of the alleles[14], and Alzheimer's Disease Pathology, identified as Alzheimer's-type neurofibrillary tangles.[16] The clinical findings are summarized in Table 1.

Discussion

We report two cases with hyperkinetic movements unusual for CBS, including a pathologically confirmed CBD with generalized chorea and a patient with LID. In both cases, the hyperkinetic movements were an important source of disability. Comparatively, the chorea manifested late in the clinical course, suggesting that these features may relate to advanced pathological stages, whereas the LID emerged after approximately 3 years. Both clinical phenotypes were representative of probable CBS.[1] These cases illustrate the heterogeneity of the motor spectrum of CBS in advanced stages and do not necessarily represent distinct phenotypes.[21]

We infer that these findings are unusual, and the mechanisms behind chorea remain unclear. A possibility is that the CBD patient had less severe involvement of the Globus Pallidus internus (GPi). Neuropathological studies already suggest that the variability in clinical presentation may be explained by the distribution of neurodegeneration.[22] Different densities of tau pathology involvement in specific neuroanatomical structures have been linked with clinical presentations.[6,22,23] From a clinical standpoint, through our systematic review, we identified late-onset patterns of chorea in CBS.[15,19] The authors theorize that a possible mechanistic explanation is a relatively intact striatum. However, there is no specific pathological description of the different basal ganglia structures mentioned in the literature. Taken together, this supports that the emergence of chorea and LID may rely on a variable topographical extension. Future research could aim to identify if there are specific neuroanatomical substrates in different stages of CBS. Clinical variability has also been reported by Ouchi et al,[24] who found that eight out of 10 patients did not meet CBS criteria in the early stages, indicating that the progressive emergence of core clinical features was only observed in more advanced stages. The main challenge is that proposing staging systems for CBS-related conditions may require a multicenter collaboration and the validation of biomarkers. Likewise, a critical aspect would also be determining the factors driving pathological progression to specific brain areas.

While there is a plethora of studies reporting the overlapping features of 4R-tauopathies,[25] especially between CBD and PSP, there are inherent oculomotor abnormalities in CBD, which are clinically challenging to separate from those of PSP.[24,26] This is

TABLE 1 *Clinical characteristics of CBS or CBD patients with chorea, unspecified hyperkinetic movements, or levodopa-induced dyskinesias*[a]

Author year	Patient ID	Sex	Age	Disease duration until hyperkinetic movement development	CBD neuropathological confirmation	Co-pathologies	Type of hyperkinetic movements	Levodopa-induced dyskinesias
Anhaus, 2000[13]	1	M	59	8 months	NA	NA	Orobuccofacial and unilateral limb dyskinesias	No
Caparros-Lefebvre, 2009[14]	1	F	70	0 months	Yes	Yes (HD)	Jerky arrhythmic involuntary limb movements	No
Lang, 2005[17]	1	F	NA	NA	NA	-	Large amplitude bizarre movements of her limbs	Yes
Vanek, 2001[20]	NA	NA	NA	NA	NA	-	Chorea	No
Vanek, 2001[20]	NA	NA	NA	NA	NA	-	Chorea	No
Vanek, 2001[20]	NA	NA	NA	NA	NA	-	Chorea	No
Vanek, 2001[20]	NA	NA	NA	NA	NA	-	Chorea	No
Frucht, 2000[15]	1	F	74	4.5 years	Yes	No	Dystonic blinking, neck and torso wiggling, mild choreic movements of the right arm and both legs	Yes
Rinne, 1994[19]	5	F	72	4 years	Yes	No	Choreiform movements of the fingers of the right hand	No
Riley, 1990[18]	1	NA	NA	NA	No	-	Orolingual dyskinesia	
Gibb, 1989[16]	2	F	62	NA	Yes	No	Speech-induced limb chorea, orofacial dyskinesia	No
Gibb, 1989[16]	3	F	66	NA	Yes	Yes (AD)	Choreiform movements of the limbs	No

Abbreviations: NA: not available. CBD: Corticobasal Degeneration. HD: Huntington's Disease. AD: Alzheimer's Disease.
[a]Drug-induced dyskinesia, chorea, or hyperkinetic movements are not included, except for those induced by levodopa.

exemplified in our first case, the patient exhibited prominent vertical gaze involvement, indistinguishable from that of PSP. It appears that this is generally a late feature of CBD; a careful examination and linear documentation are necessary. A central drawback is defining clinically late-onset vs early-onset oculomotor dysfunction, as in PSP, this feature may not be apparent early in the course.[27] Comparably, studies on PSP and CBS have indicated that classifying patients into specific phenotypes is challenging, as patients may meet criteria for multiple phenotypes over the course of their illness or be unclassifiable.[27] Even grouping PSP and CBD into probable 4R-tauopathies has a low sensitivity within the first 3 years of disease progression, particularly for CBD.[25]

There is increasing evidence of copathology contributing to atypical manifestations in atypical forms of parkinsonism.[28] Although the neuropathological details were not available in all the cases identified in our systematic review, two out of five cases with neuropathological descriptions were reported as having mixed pathologies.[14,16] Notably, one of these cases involved genetically confirmed HD,[14] suggesting that HD could explain the involuntary movements.[14] Likewise, there is growing interest

in the role of intermediate alleles in HD[29,30]; however, our case had normal CAG repeats in both alleles, which supports primary CBD pathology as the main factor leading to chorea. In clinical practice, investigating the pathology or co-pathologies in CBS at present is challenging, even with the advent of novel biomarkers.[28] For instance, in the systematic review one of the pathologically confirmed cases had CBD and AD pathology.[16] Indeed, amyloid-β was present in 41% of CBD cases in one large series,[31] which may suggest that a positive biomarker for AD pathology is not diagnostic of CBS-AD but could simply indicate the presence of AD copathology. In our second case, p-tau 217 was negative, and, therefore, the utility of fluid biomarkers for AD may be related to their negative predictive value. However, the interpretations of AD biomarkers in the context of CBS warrant further investigations with neuropathological confirmation.

There are multiple limitations that we need to outline. First, we could not perform 4RT staining, but the CBD diagnosis was made based on validated pathological criteria.[12] While we identified tau pathology in different basal ganglia structures, we were unable to compare the relative neuronal loss, which could further support our explanation of chorea emerging due to a minor involvement of the GPi. Without additional functional imaging studies and particularly neuropathology, we can only speculate as to why our second case developed such severe LID in contrast to the usual experience of patients with CBS treated with levodopa.

We can conclude that chorea can occur in CBS as supported by our cases and the systematic review. Chorea and LID are likely associated with the progression of the disease. Both our cases and those identified in the systematic review presented after an existing CBS phenotype, which suggests that they may represent late-onset manifestations. Future research should aim to quantify the extent of involvement of various neuroanatomical structures to better understand the pathological mechanisms that lead to chorea and LID. There is an urgent need for biomarkers that can define the specific underlying pathology, stage the disease, and indicate the level of neuroanatomical involvement.

Author Roles

(1) Research project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript: A. Writing of the First Draft, B. Review and Critique.

A.M.E.: 1A,1B, 1C, 3A.

M.R.: 1B, 1C, 3A, 3B.

M.R: 1C, 3B.

S.G.: 1B, 3B.

A.L.: 1A, 1B, 3B.

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Disclosures

Ethical Compliance Statement: 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. The authors confirm that institutional review board approval or patient consent was not necessary for this work, as it was solely based on a review of their routine clinical practice. Informed consent was obtained from the patients or their power of attorney.

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Data Availability Statement

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions. ■

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Supporting Information

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

Supplementary Material S1. Search strategy.

Figure S1. Preferred Reporting Items for Systematic reviews and Meta-Analyses (PRISMA) flow diagram.