A case of corticobasal degeneration presenting with alien limb syndrome

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Abstract
Alien limb syndrome (ALS) is a very rare condition where the affected persons are not able to recognise the affected limb as their own, and regard it as being foreign or alien to them. We present a patient with ALS secondary to corticobasal degeneration, which is a rare neurodegenerative parkinsonian disorder. We discuss the clinical features, neuropathology and management of corticobasal degeneration.

Keywords: alien limb, CBD, corticobasal degeneration, taupathy

Case report
An 80-year-old man was referred to the outpatient clinic in April 2004 with uncontrolled hypertension and deteriorating mobility. The patient had a past medical history of hypertension and was taking bendrofluazide, atenolol, valsartan and clopidogrel.

His main complaint was difficulty using his left arm and hand, which had started 2 years before and gradually progressed. He said that it pained him when he tried to use it, and it had become useless. He lost control over it and referred to it with phrases like ‘it has a life of its own’, ‘it doesn’t do what I want it to do’, and ‘it can wander off by itself’.

His mobility deteriorated markedly from being able to swim and play golf a year ago, to having a very slow shuffling and unsteady gait. Further findings on examination revealed a masked facial expression and marked apraxia of the left upper limb. He scored 29/30 on the Mini Mental State examination. Brain CT scan showed cortical atrophy with no clear asymmetry.

The clinical diagnosis was that of corticobasal degeneration (CBD) presenting with alien limb phenomenon. This was based on the gradual onset and progressive nature of a parkinsonian disorder associated with cortical features, and the highly asymmetrical features. Parkinsonism was evidenced by the slow shuffling gait, and the cortical features by severe apraxia with alien limb phenomenon and dystonia. Parkinsonism did not respond to levodopa, but painful dystonia was helped by levetiracetam. His condition gradually progressed and he died at home some 2 years after initial presentation.

Discussion
CBD, also known as corticobasal ganglionic degeneration, is a rare progressive neurodegenerative disorder that was first reported by Rebeiz in 1967 [1].

Neuropathologically CBD is characterised by asymmetrical degeneration of the cerebral cortex and the basal ganglia (hence the name corticobasal). The perioladic posterior frontal and parietal cortex are primarily affected while the temporal and occipital cortex are relatively spared. Other structures involved include the substantia nigra, and to a lesser extent the putamen, pallidum, thalamus and hypothalamus [2].

Microscopically there is neuronal loss, gliosis and tau-positive swollen achromatic neurons (ballooned neurons) especially in superior frontal and parietal gyri [3].

Similar to fronto-temporal dementia and progressive supranuclear palsy (PSP), CBD is a taupathy (characterised by abnormal deposition of the microtubule-associated protein tau). There is evidence of genetic and phenotypic overlap between these disorders, especially between CBD and PSP [4].

CBD usually begins in the sixth decade with equal distribution between sexes. Due to its rarity, the exact prevalence is not known, but is thought to be at least two per million [5].
Clinical features can be explained by involvement of the cortical and sub-cortical structures. Cortical-related features may include apraxia, alien limb phenomenon, focal dystonia and myoclonus. Cognitive and behavioural problems can be a feature and may include non-fluent dysphasia, frontal executive impairment, acalculia and visuospatial impairment. Eye movements are usually preserved, although there may be a delay in initiation of saccades [6]. Involvement of sub-cortical structures usually results in highly asymmetrical parkinsonism, where tremor is uncommon [7].

Bogen defined alien limb syndrome (ALS) in 1979 as ‘a circumstance in which one of the patient’s hands behaves in a way which the patient finds foreign, alien or at least uncooperative’. Levitation and mitgehen (continuous tactile pursuit) have been observed, but spontaneous limb levitation on its own is not sufficient to diagnose ALS. ALS has also been reported with callosal tumours, callosotomy and, occasionally, frontal strokes [8].

Neuroradiological imaging in CBD may show asymmetrical atrophy, but anatomical imaging can be normal especially in early disease. Functional imaging with PET or SPECT scan can be helpful and may show focal reduction in cerebral blood flow and glucose metabolism [6].

Prognosis of CBD is poor with a mean survival of only 7 years [5]. Parkinsonism generally responds poorly to dopaminergic medication. The main emphasis of management is supportive with a multidisciplinary team approach.

Key points
- Alien limb syndrome is a rare presentation of CBD, callosal tumours and frontal strokes.
- CBD is a taupathy; its diagnosis is mainly clinical. Neuroradiological imaging may be inconclusive as was in our case.

Conflicts of interest
None

References

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