Crossed cerebellar diaschisis in corticobasal syndrome

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INTRODUCTION

Corticobasal degeneration (CBD) is a progressive neurodegenerative disorder characterized by the asymmetric presentation of limb rigidity and/or akinesia, dystonia, myoclonus, cortical sensory deficit, and alien limb phenomenon, in addition to specific neuropathological hallmarks [1]. The term corticobasal syndrome (CBS) is used to define the clinical features originally described by Rebeiz, Kolodny and Richardson in 1968 [2], in absence of the histopathological confirmation. In fact CBS could also represent the clinical presentation of other neurodegenerative diseases, including progressive supranuclear palsy, Alzheimer's disease, frontotemporal dementia with TDP-43 inclusions [3,4].

We describe a patient affected by CBS, who showed a crossed cerebellar diaschisis at 18FDG-PET imaging.

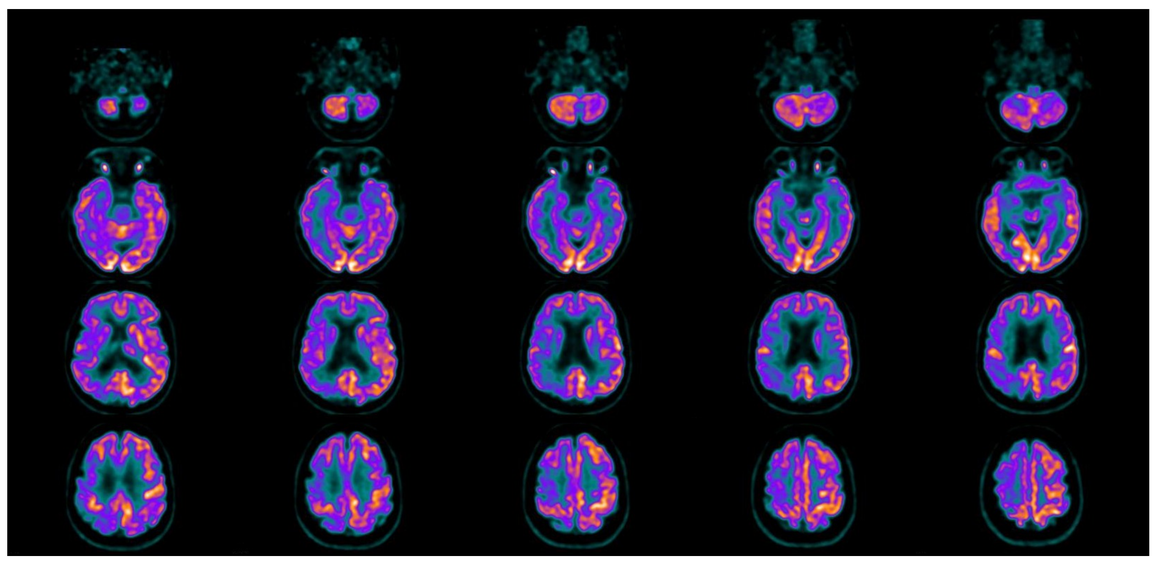
CLINICAL HISTORY

At the age of 63 years, a right-handed woman came to our attention because of progressive difficulty in walking, caused by right leg numbness. She was no longer able to drive after two not traumatic car accidents, due to difficulty to readily press the foot brake pedal. At the same time she noted a right hand clumsiness with difficulty in the use of domestic instruments. Handwriting became illegible. Language and memory were preserved. Past medical history was unremarkable.
On first neurological examination, an increased muscular tone with rigidity was pointed out in her right upper and lower extremities. Right side bradykinesia and dysdiadochokinesia were noted, the latter due to mild dystonic posture of the forearm. A dystonic posture was also noted in right foot only during walking. No limb apraxia or alien limb phenomena were present at that time. Sensory examination was normal. The extraocular movements appeared normal except for saccadic intrusion during smooth pursuit. Tendon reflexes were brisk on the right and normal on the left side, plantar responses were flexor.
Neuropsychological evaluation revealed a mild executive deficit, with sparing of short term memory. Line bisection task revealed a left visual field inattention. Language production and comprehension were preserved.
The subsequent year we observed a worsening of the right side dystonia. She was unable to walk without support. Her right arm raised during conversation against her will. An abnormal two-point discrimination task was performed on her right side. Her right foot was intra-rotated in a fixed dystonic posture. Mild dysarthria and dysphagia were showed.
Based on the clinical findings and the disease course, CBS was assumed.
The following year she was confined in a wheelchair. Dystonia was marked to both upper and lower right extremities, with muscular pain, requiring local botulinum toxin treatment. Emotional incontinence was present. Dysarthria and dysphagia worsened. Short term memory was preserved.

Blood and CSF analysis (included dosage of β-Amyloid 1-42, tau protein and phosphorylated tau protein) were unremarkable. Brain MRI and cerebral FDG-PET were performed.

DISCUSSION

Crossed cerebellar diaschisis (CCD) is defined as metabolic depression in the cerebellar hemisphere contralateral to supratentorial lesion. It is caused by loss of excitatory afferent inputs to the cerebellar hemisphere by the contralateral cortical area from which originate the cortico-ponto-cerebellar (CPC) tract. It is typically described in patients affected by acute cortical stroke.
At our knowledge, only two single-case reports described a similar PET finding in a patient affected by CBS [5] and in an autopsy-proven CBD [6]. Finally, Akdemir et al. [7] reported a 63-year-old patient with CBD who had complaints of left-hand apraxia, in addition to her findings of parkinsonism. The authors described an hypometabolism in right basal ganglion, thalamus, and frontal, parietal and cortical regions. Further an hypometabolism in the left cerebellar hemisphere is evident in 18FDG-PET slices.
18FDG-PET metabolism in CBS patients was first studied by Eidelberg et al. [8], who found a decreased metabolism in thalamus and inferior parietal cortex contralateral to the most affected side. Further investigations reported an asymmetrical decrease of glucose metabolism in primary motor and somatosensory cortices, thalamus, caudate nucleus, temporo-parietal and parieto-occipital regions [9].
Human DTI studies found that the CPC tract is divided into four portion (fronto-, parieto-, occipito-, temporo-ponto-cerebellar tracts); the temporal and frontal contributions to the CPC tract are greater than the parietal and occipital portions [10].
Our finding indirectly confirms the asymmetrical involvement of the fronto-temporo-parietal cortices in CBS, with secondary progressive axonal loss of the ipsilateral corticopontine tract, causing hypometabolism of the contralateral cerebellar hemisphere.



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