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Short communication

Subacute corticobasal syndrome following internal carotid endarterectomy

Video Animation

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ABSTRACT

The present report is of two patients who, immediately after internal carotid endarterectomy, presented with unexplained hemiplegia, despite normal findings on repeated MRI scans, which secondarily evolved into homolateral subacute corticobasal syndrome (CBS), with asymmetrical hemispheric hypometabolism and evidence of dopaminergic denervation. This prompted us to propose an hypothesis of transient cerebral hypoxia arising during the surgical clamping period that might have provoked a prolonged or permanent functional lesion of the left hemisphere and basal ganglia, with no visible infarction on MRI but only synaptic rearrangement of the neural networks, thereby revealing or exacerbating a potentially preexisting silent impairment.

1. Introduction

Corticobasal degeneration (CBD) is clinically characterized by a progressive asymmetrical presentation of limb rigidity or akinesia and apraxia, with other findings suggestive of additional basal ganglia and cortical dysfunction (such as dystonia, tremor, myoclonus, alien limb phenomena, cortical sensory deficit and/or impaired cognition) [1]. When first described, CBD was considered a distinct clinicopathological entity, but since then, it has been shown that, despite its many clinical diagnostic criteria, an ante-mortem diagnosis of CBD was pathologically confirmed in only 25–56% of cases, with other cases turning out to be progressive supranuclear

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palsy, frontotemporal dementia, Alzheimer's disease or Parkinson's disease [1,2]. These clinicopathological inconsistencies have led to the preferential use of the term 'corticobasal syndrome' (CBS) for the clinical presentation, while reserving the term CBD to describe the histopathology [1]. A few cases of CBS secondary to internal carotid artery (ICA) stenosis have recently been reported [3–5]. These vascular CBS cases presented with an insidious onset and gradual progression. Subacute CBS has previously so far only been described in cases of Creutzfeldt–Jakob disease [6].

Our present report is of two patients who, immediately after ICA endarterectomy, presented with unexplained hemiplegia, despite normal findings on repeated magnetic resonance imaging (MRI) scans, which secondarily evolved into homolateral subacute CBS. These cases were seen 1 year apart, and followed procedures undertaken by two different surgeons at two different institutions.

2. Observation

Both these patients had a history of diabetes mellitus, hypertension and dyslipidemia. Patient 1 (P1; a 67-year-old left-handed male Caucasian) presented with an 80% right ICA stenosis and two episodes of transient amaurosis of the right eye, whereas Patient 2 (P2; a 71-year-old right-handed male Caucasian) was asymptomatic, but had a history of a stent for ischemic heart disease and a left middle cerebral artery stroke 7 years previously, with left ICA thrombosis (and persistent partial dysphasia), which led to the decision to perform a right carotid endarterectomy. Neither patient had a family history of movement disorders nor any reported bradykinesia prior to the surgery.

Both procedures unfolded with no problems (each had a clamping period of 30 min and 38 min, respectively), yet both patients awoke from their operations with left-sided proportional hemiplegia. Angiography plus computed tomography (angio-CT) of both patients and CT perfusion scanning (P2 only, Fig. 1) performed immediately failed to reveal any abnormality in the right carotid field. There was also neither restenosis of the right ICA nor any stenosis of the other carotid axis, and the circle of Willis was complete with no hypoplasia. Yet, after 24 h and 72 h, both patients were still presenting with proportional left hemiplegia while angio-CT remained normal.

When axial fluid-attenuated inversion recovery (FLAIR) and diffusion-weighted (DWI) MRI (1.5T, 5-mm slices) sequences were performed on day 6 (P1) and day 15 (P2), the scans revealed only mild atrophy and no infarcts in the right hemisphere. Left frontal hypersignalling compatible with vascular sequelae was identified in P2, whereas extremely sparse white-matter hyperintensities were noted in both patients (P1: Fazekas scale = 0; P2: Fazekas scale = 1; Fig. 2) [7]. When P1 underwent another MRI (3T) on day 20, it showed only scant hypersignalling in the periventricular white matter with no evidence of a recent stroke. While gradually recovering from the hemiplegia, P1 developed myoclonus on day 40 (video 1) whereas P2, on day 20, developed a tremor of the left superior limb (LSL) associated with rigidity and bradykinesia. The clinical evolution and investigations performed in both patients are summarized in Table 1. Neither patient presented with any clinical manifestations other that the ones described here.

Neuropsychological assessment performed in P1 at 3 months revealed moderate left-sided apraxia (symbolic gestures), with preserved global cognitive functioning except for a mild episodic memory impairment, which was apparently present, according to the patient's wife, before the endarterectomy. At 6 months after the surgery, an improvement in myoclonus and LSL dexterity was observed, although an akinetic-rigid syndrome persisted (video 1). After 9 months, a rapid cognitive decline of mainly dysexecutive symptoms was evident. Surprisingly, however, at the last assessment performed 20 months after endarterectomy, there was clear regression of the cognitive impairment in line with a global improvement reported by the patient.

For P2, neuropsychological assessment at 3 months revealed anterograde verbal and visual memory impairment associated with a dysexecutive syndrome, and visuoconstructive apraxia and left-sided hemineglect. Another assessment conducted at 9 months found mostly praxis disorders involving the LSL (ideomotor and melokinetic apraxia; video 2), with no dysexecutive impairment, but an overall slowing of cognitive processing instead.

Fluorodeoxyglucose (^{18}F-FDG) positron emission tomography (PET) identified, in both patients, severely decreased metabolism in the right frontoparietal area and right striatum (Fig. 2) which, in P2, was associated with milder decreased metabolism in the left frontal area, corresponding to a sequela of a previous stroke. Use of ^{123}I-ioflupane (^{123}I-FP-CIT, DaTscan, GE Healthcare, Chicago, IL, USA) showed, in both patients, reduced radioligand uptake in the right striatum, compatible with mild dopaminergic nigrostriatal presynaptic denervation in the right putamen (Fig. 2).

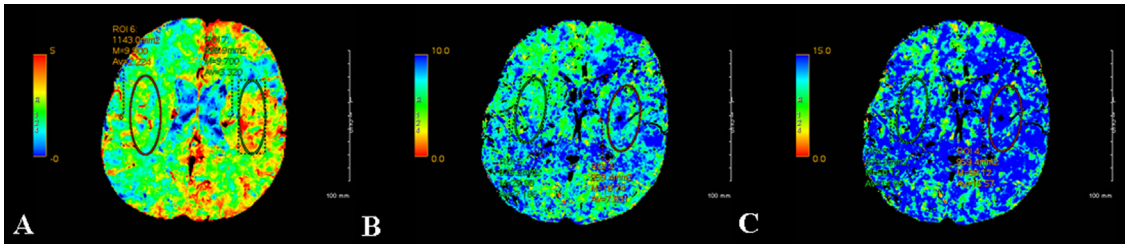

Fig. 1 – Perfusion parameters on computed tomography (CT) of Patient 2: (A) mean cerebral blood volume (CBV; right: 2.54 mL/s/100 g; left: 3.93 mL/s/100 g); (B) mean maximum time (T_{max}; right: 7.16 s; left: 7.93 s); and (C) mean transit time (MTT; right: 14.06 s; left: 15.57 s).

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