

Hemic chorea–hemiballism syndrome following a thrombo-embolic striatal infarction

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Received: 11 November 2011 / Accepted: 12 April 2012 / Published online: 25 April 2012
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Case report

A 70-year-old man was admitted to our emergency department for sudden onset of abnormal involuntary movements involving his upper and lower left limbs. Hemic choreal movements were restricted to his left limbs, associated with rare episodes of hemiballism (video). There were no cranial nerve alterations, no sensory loss or limb weakness. The patient reported mild hypertension under treatment. There was no history of diabetes mellitus or epilepsy, no prior exposure to neuroleptics or recent change in therapy. He denied any similar previous episode or family history of movement disorders.

Chest radiography, electrocardiography, and laboratory tests including glucose, glycated hemoglobin, coagulation parameters, and immunoglobulin electrophoresis were normal. Brain non-contrast CT scan on admission showed no signs of acute ischemia (Fig. 1a). A supra-aortic-trunk Doppler sonography revealed an irregular surface plaque, partly ulcerated, on the origin of the right internal carotid artery with stenosis of about 90 %, confirmed at CT angiography (Fig. 1b).

Brain MRI examination, performed 3 days after onset of symptoms, showed a linear altered signal area in the right basal ganglia, mostly in globus pallidus, on diffusion weighted imaging (DWI) sequences (Fig. 1c). The area was also visualized on the apparent diffusion coefficient (ADC) map (Fig. 1d), and on fluid attenuated inversion recovery (FLAIR) sequences (Fig. 1e). T1-weighted sequences of the basal ganglia were unremarkable (Fig. 1f). MRI angiography did not reveal abnormalities in the intracranial circulation.

Our patient was thus diagnosed having an acute left hemic chorea–hemiballism (HCHB) syndrome, caused by a carotid-originating embolic stroke, which affected the right lenticular nucleus.

Medical treatments included immediate unfractionated heparin infusion, followed by warfarin and atorvastatin, while waiting for surgical treatment of the right carotid artery stenosis. The movement disorder was symptomatically controlled by oral administrations of clonazepam. 2 months later, he underwent surgical revascularization of the internal right carotid stenosis by thromboendarterectomy. 3-month follow-up revealed considerable symptom reduction with minimally disabling residual movements.

Electronic supplementary material The online version of this article (doi:10.1007/s10072-012-1098-6) contains supplementary material, which is available to authorized users.

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Discussion

Hemic chorea–hemiballism is a hyperkinetic movement disorder consisting in involuntary, aimless, large amplitude limb excursions, which mainly involve distal body parts.

Choreal movements are an uncommon finding in emergency context. Such a disorder can be associated with different conditions, including infective, autoimmune or central nervous diseases, oestrogen or levodopa replacement therapies, polycythaemia rubra and chorea gravidarum

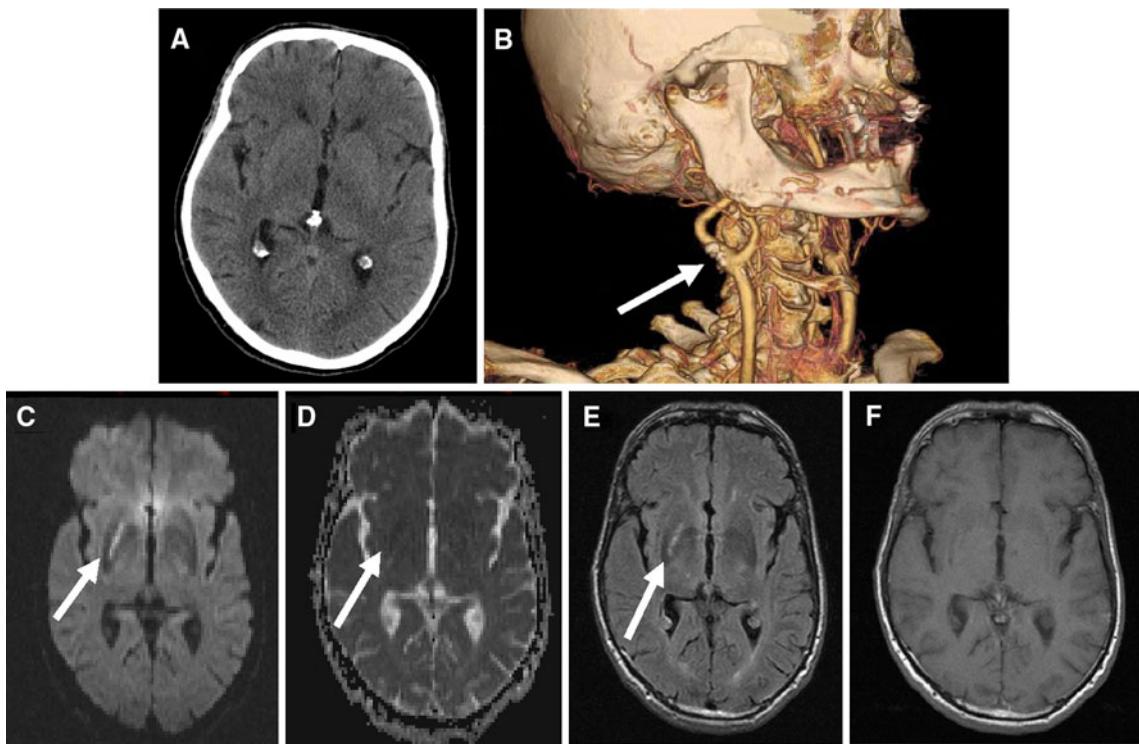


Fig. 1 Brain non-contrast CT scan (a) and brain T1-weighted MRI (f) do not show any lesion. **b** Tridimensional reformatted CT-angiography shows the ulcerated plaque (arrow) at the origin of right internal carotid artery likely responsible for the acute ischemic event.

DWI sequences (c) and ADC maps (d) show a linear ischemic area in the right basal ganglia. **e** T2-FLAIR confirms the acute ischemic lesion in the basal ganglia and in the periventricular areas (arrows)

[1, 2]. Chorea associated with these diseases have consistently a slow onset. Whenever onset is acute and apparently isolated, vascular events and metabolic disorders are most likely cause of the disorder.

Basal ganglia blood supply is mostly provided by lenticulo-striatal arteries. These are small diameter, penetrating vessels branching at right-angle from M1 segment of the middle cerebral artery [3]. Since their distribution territory has no collateral blood supply, they are functionally regarded as “terminal” thus making basal ganglia particularly vulnerable to hypoxia or ischemia.

In the present case, the neuroimaging evidence of acute ischemia and congruent carotid atherosclerotic lesion supported the clinical diagnosis of thrombo-embolic striatal infarction.

Emboli coming from the internal carotid probably compromised the terminal lenticulo-striatal circulation causing a circumscribed pallidal infarction. Our syndrome was associated with a typical lesion within the basal ganglia. Others have reported cortical lesions to cause similar clinical expressions [4].

Although hemichorea usually develops within few hours from stroke onset (as in our patient), time intervals as long as 5 days have been reported. Such a delay has raised the hypothesis of restorative neuroplasticity-mediated

pathogenesis [4]. Our report seems inconsistent with such a hypothesis and stresses the relevance of the ischemic lesion itself as a source of acute basal ganglia motor circuit dysregulation for HCHB determination. Ischemia, thus, would induce an increased dopamine striatal release with consequent interruption of the typical transmission pattern among basal ganglia circuitry. The acute circuitry dysfunction would result in loss of thalamus movement control and in excessive motor facilitation [1, 3, 4].

Among differential diagnosis for acute onset hyperkinetic disorders we should consider transient ischaemic attack limb-shaking syndrome (LSS).

Limb-shaking syndrome is a rare manifestation of transient ischaemic attacks due to carotid-occlusive disease. The LSS may assume either choreic or tremor-like appearance involving an arm or leg, and it manifests as single episode or several times a day. Pathophysiology is related to cerebral hypoperfusion; carotid stenosis and maneuvers, such as arising from bed as well as hyperextending the neck, lead to decreased cerebral blood flow in watershed territories. Neuroimages are useful in demonstrating carotid-occlusive disease in absence of acute ischemia signs [5].

Aside from acute vascular pathology, patients presenting with sudden hemichorea should also be investigated for

epilepsy. In our case epilepsy was excluded due to clinical manifestations and to the absence of focal cortical involvement on neuroimages.

Another condition that should be considered is nonketotic hyperglycemic syndrome [6, 7]. This is a rare neurologic complication of long-standing diabetes. Under conditions of poorly controlled blood glucose, patients may present hemichorea–hemiballism. Ischemic and nonketotic hyperglycemic HCHB have similar clinical presentation. Radiological investigations provide decisive findings for differential diagnosis. Nonketotic hyperglycemic HCHB presents CT basal ganglia hyperdensity caused by focal mineral deposition (calcium, iron, or manganese) and T1-weighted MRI sequences striatal hyperintensity as a consequence of focal microhemorrhages [6, 7]. Cortical involvement is consistently absent. Regardless of its pathogenesis, HCHB acute phase treatment is based on neuroleptics and/or benzodiazepines administration. Long-term prognosis may vary considerably. In nonketotic hyperglycemic HCHB, symptoms usually disappear with blood glucose normalization. In post-ischemic HCHB minimal functional impairment may persist. Syndromes associated with cortical ischemic localizations are thought to have a better prognosis [3].

Our patient showed clear clinical–radiological signs of focal striatal infarction resulting in sudden onset HCHB. Even without 3-month imaging follow up, the partial syndrome regression can be interpreted either in terms of

post-ischemic tissue recovery or of compensatory neuronal circuits development.

This report remarks the relevance of neuroimaging techniques in cases of acute onset movement disorders.

Patient consent: Patient's informed consent was obtained for publication.

Conflict of interest The authors declare no conflicts of interest.

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