Ref: Ro J Neurol. 2018;17(3)
DOI: 10.37897/RJN.2018.3.9

Infarction of the posterior limb of the internal capsule: The first clinical manifestation of lupus

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ABSTRACT

Isolated cerebral infarction in a small vascular territory such as the posterior limb of the internal capsule rarely represents the first clinical manifestation of systemic lupus erythematosus, and represents a diagnostic challenge, especially if other organ systems are not affected. We report the case of a 28-year-old woman who was admitted to the emergency room with acute onset left-sided hemiparesis. She was diagnosed with acute ischaemic stroke and treated with intravenous thrombolysis, without improvement. The localisation of the lesion in the posterior limb of the internal capsule was based on MRI with DWI sequences, performed after thrombolysis. After extensive workup, a diagnosis of lupus was established. The functional outcome was unfavourable. AChA territory infarction is a special entity, which is increasingly recognised owing to extensive use of MRI in stroke care, with particular clinical characteristics. Cerebrovascular events can be the first clinical manifestation of lupus and contribute to premature morbidity and disability.

Keywords: cerebral infarction, systemic lupus erythematosus, hemiparesis, thrombolysis, vasculitis

INTRODUCTION

The posterior limb of the internal capsule is supplied by the lenticulo-striate branches of the middle cerebral artery and the anterior choroidal artery (AChA). The AChA is a thin artery that originates from the distal segment of the internal carotid artery (ICA), 2-5 mm proximal to the intracranial bifurcation. Additionally, the posterior limb of the internal capsule supplies the medial temporal lobe, the optic radiations, lateral thalamus, the tail of the caudate nucleus, the lateral geniculate body, and the medial part of the pallidum (1). Classical, complete AChA syndrome includes hemiplegia, hemihypoaesthesia and homonymous hemianopia; however, there is important variability in the extent of the lesions, leading to a wide spectrum of neurological symptoms, frequently with a progressive or fluctuating course (2,3).

Before the widespread use of MRI in stroke management, the reported incidence for AChA infarction was low (2.9%). (4) Based on a prospective stroke registry, using a rigorous MRI protocol including at least DWI, FLAIR, gradient echo and TOF images, the incidence of isolated AChA infarction was found to be 8.1% (1). The mechanism of AChA infarction is varied and includes small and large artery disease. Small vessel infarction secondary to occlusion of the small penetrating arteries is an important finding and can be attributed in rare cases to lupus vasculitis (5,6).

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Article History:
Received: 9 August 2018
Accepted: 22 August 2018

Stroke is not rare in patients with documented lupus, with an incidence of 15%. The main mechanisms are the cardioembolism, the antibody-mediated hypercoagulable state, and cerebral vasculitis. Isolated microvascular infarction as the first clinical manifestation of lupus is a very rare finding and represents a diagnostic challenge, especially if only one organ system is affected (7,8).

We report on the case of a 28-year-old woman with acute left-sided hemiplegia secondary to isolated infarction in the posterior limb of the internal capsule, with poor response to i.v. thrombolysis.

CASE REPORT

A 28-year-old woman, with an unremarkable past medical history, who had received oral contraceptives for two years, presented in the emergency department with acute onset headache, nausea, left-sided weakness, and dysarthria, 104 minutes after the beginning of the symptomatology. Her blood pressure was 125/80 mmHg, heart rate 70 bpm. The neurological examination revealed left-sided central facial palsy, left-sided hemiparesis, grade 0 in the upper limb, grade 2 in the lower limb, left-sided hemihypoaesthesiae and mild dysarthria (National Institutes of Health Stroke Scale 12 points). The complete blood count, chemistry profile and coagulation tests were normal. The native cerebral CT scan was normal. The patient had no contraindication for i.v. thrombolysis; this was performed 172 minutes after the onset of the symptomatology, without improvement in the neurological symptoms. A cerebral MRI 1 hour after thrombolysis revealed in the DWI sequences an acute infarction in the posterior limb of the right internal capsule. FLAIR images were normal (Fig. 1). A control cerebral CT scan 24 hours after thrombolysis also showed an infarction in the posterior limb of the internal capsule (Fig. 2). Antiplatelet therapy with 100 mg aspirin qd was initiated.

An extensive workup was performed for the aetiology of the stroke. Carotid and cardiac ultrasound and ECG were normal. The lipid profile, thyroid function, and thrombophilia panel were normal, and infectious markers were negative (HIV, TPHA, hepatitis C). Of the autoimmune markers tested, antinuclear antibodies (40.7 IU/ml; normal range 0-23 IU/ml) and anti-dsDNA antibodies (70 U/ml, normal range 0-40 U/ml) were elevated. Antiphospholipid antibodies, lupus anticoagulant, anti-histone antibodies, and the Coombs test were negative, and the beta-2 microglobulin level and serum protein electrophoresis were normal. A rheumatological consultation established a diagnosis of systemic lupus erythematosus. Treatment with Plaquenil was introduced.

Rehabilitation therapy was initiated two weeks after the onset. Motor function improved to grade 4 in the lower limb and grade 2 in the upper limb. The patient was discharged from the hospital with instructions to continue rehabilitation therapy. At

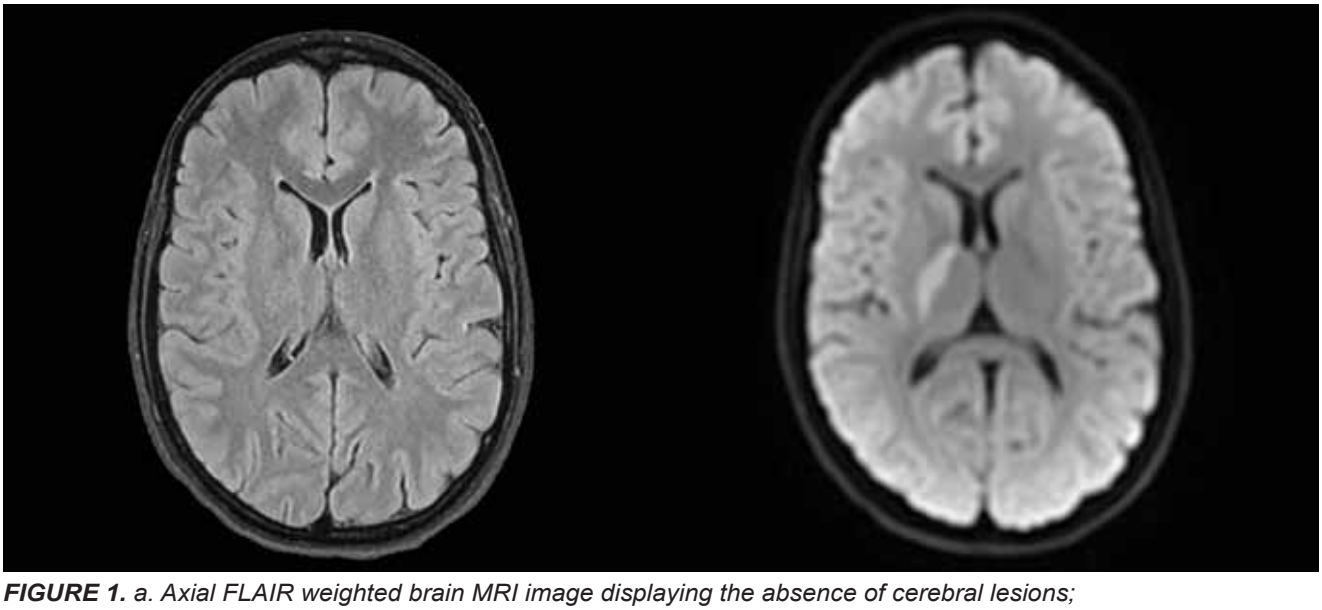

FIGURE 1. *a. Axial FLAIR weighted brain MRI image displaying the absence of cerebral lesions; b. Axial diffusion-weighted image, revealing an acute lesion in the posterior limb of the right internal capsule*

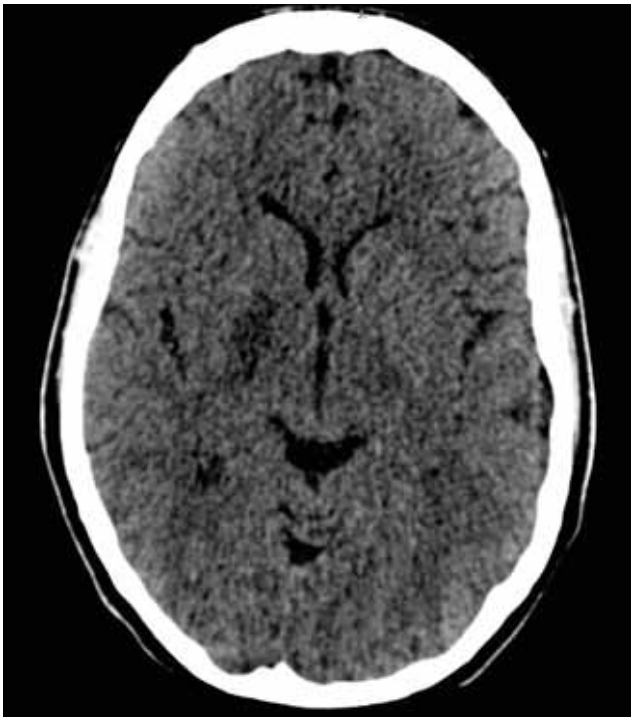

FIGURE 2. *Axial non-enhanced CT scan showing a hypodense lesion at the level of the posterior limb of the right internal capsule*

the three-month control examination, the motor deficit had not improved significantly.

DISCUSSION

The prevalence of AChA infarction in patients with acute ischaemic stroke, based on prospective stroke registries and using state-of-the-art imaging methods, is remarkably higher than 8% (1,9). A clinical presentation with the classical syndrome is rare. Frequent symptoms include motor deficits, followed by sensory dysfunction. Hemianopia, ataxic hemiparesis, isolated ataxia, or dysarthria are very rare findings. The high prevalence of motor deficits is closely associated with involvement of the posterior limb of the internal capsule (1).

In a large case series published by Ois et al., when comparing AChA territory strokes with hemispheric one, they found that AChA infarct patients were younger, with a male predominance, more often diabetic, and the majority of them were not treated previously with antithrombotic medication (9).

There are contradictory data in the literature regarding the underlying aetiology of AChA infarctions. In patients with involvement of both the subcortical and mesiotemporal territories of the AChA, the small-vessel occlusion mechanism in unlikely (10).

In the case of small AChA infarctions, the association with carotid stenosis is more frequent and the cardioembolic sources are rare compared with small deep infarcts in other vascular territories. Compared with hemispheric infarcts, patients with AChA infarcts have a lower rate of significant arterial stenosis and cardioembolism (9). Ois et al. found an independent relationship between the size of the infarct and the pathophysiological mechanism. AChA infarctions > 20 mm were significantly associated with a cardioembolic source and significant carotid stenosis, and the prognosis was worse in this group (9).

A progressive course is commonly seen in AChA infarctions, and early neurological deterioration occurs in 60% of patients, leading to worse functional outcomes. The frequency of a progressive course is higher compared with other infarct sub-types (3,11,12). The mechanisms and predictive factors regarding the background of deterioration have not been clarified.

Despite the frequent occurrence of progressive evolution, patients with AChA infarctions have lower mortality rates and lower recurrence rates compared with patients with hemispheric infarctions, but the functional outcome is unfavourable, with a high rate of dependent status at three months (9). There are contradictory data in the literature regarding the therapeutic response to intravenous thrombolysis. In a case series reported by Wu et al. on15 thrombolysed patients, only one (6.7%) presented with progression (13). They concluded that thrombolytic therapy in these cases reduces the risk of stroke evolution and improves the functional outcome. However, in a larger case series (21 thrombolysed cases) published by Chausson et al., 12 of 21 stroke patients continued to progress. There was no symptomatic haemorrhagic transformation observed in either of these studies (1).

The involvement of the nervous system in lupus varies from 18-70%, depending on definitions and the diagnostic methods applied. The most frequent neuropathological findings in the CNS, secondary to SLE, are small vessel cerebral vasculopathy and microinfarcts (14). Lupus leads to a two-fold increase in the risk of ischaemic stroke; the relative risk is highest among patients younger than 50

years of age (15). However, only one case with AChA infarction secondary to lupus has beenpublished in the literature. The authors used state-of-the-art imaging methods, i.e. 3D-reconstructed MRA images as well as conventional angiography, to demonstrate classical vasculitic changes at the level of the affected AChA, observed as narrowing and ectasia and described as a beading pattern (6).

There is limited experience with intravenous thrombolysis in cases of acute ischaemic stroke secondary to vasculitis. These patients might be at higher risk for haemorrhagic transformation; however, there are some case reports published in the literature with large vessel acute ischaemic stroke secondary to lupus with excellent outcomes after thrombolysis (16,17).

CONCLUSIONS

Stroke in young patients requires a different diagnostic workup and management, taking into account the broad spectrum of underlying causes and pathophysiological mechanisms. AChA territory infarction is a special entity with protean clinical manifestations and potentially worse functional outcomes. MRI with DWI sequences is an important tool for the correct diagnosis and localisation of the lesions. In patients with SLE, cerebrovascular events can be the first clinical manifestation and contribute to premature morbidity and disability.

Acknowledgement and financial support

This study was supported by the internal research grant of the University of Medicine and Pharmacy Tg. Mures (15609/8/29.12.2017).

Conflict of interest: none declared

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