Acta Neurologica Belgica (2020) 120:1477–1479
https://doi.org/10.1007/s13760-020-01427-x

LETTER TO THE EDITOR

Idiopathic internal carotid artery vasospasm successfully treated with balloon angioplasty

Emmanuelle Levecque[1] · Pierre Goffette[2] · Thierry Duprez[3] · André Peeters[1]

Received: 31 March 2020 / Accepted: 30 June 2020 / Published online: 25 July 2020

Keywords Stroke in young adults · Idiopathic stroke · Cervical internal carotid artery vasospasm

Introduction

Idiopathic vasospasms of the extracranial internal carotid artery (ICA) are a rare cause of cerebral ischemia of potentially underestimated occurrence. Only a few observational papers have reported the condition which could be associated with altered autonomic ICA innervation [3] and triggered by mechanical manipulation, drugs or migraines [3]. We describe a young man presenting with bilateral extracranial ICA vasospasms treated with endovascular procedure leading to a very good outcome.

Case history

A healthy 21-year-old man was admitted for right hemiparesis for 24 h with worsening NIH Stroke Scale from 6 to 10. He had no history of migraine, no traumatic event and no family history of stroke, cardiac or neurodegenerative diseases. He does not take chronic treatment. He is not addicted (tobacco, alcohol, drug). Brain CT and CT angiography (CTA) demonstrated focal occlusion of extracranial segments of both internal carotid arteries (ICA). Bilateral dissection was initially hypothesized but common features of the condition (vessel enlargement, parietal hematoma, endoluminal flap, etc.) were absent. He subsequently underwent digital substraction angiography (DSA) which confirmed CTA findings (Fig. 1a, b) and demonstrated collateral circulation mainly supplied by the external carotid arteries and the posterior circulation at a lesser degree. No major intracranial vessel was occluded. A balloon angioplasty without stenting of the symptomatic left ICA was performed in the same session (Fig. 1c, d). The next day, brain MRI showed bilateral watershed infarction prominently involving the left side (Fig. 2a). The patient was treated with aspirin and clopidogrel. Comprehensive screening for risk factors of vasospasm, embolism, and vasculitis remained negative.

The etiological balance was negative: the cardiac telemetry did not reveal any arrhythmia, routine serum analyses, autoimmune evaluation and advanced haematological tests were unremarkable and the toxicological screening was negative.

Follow-up brain MRI demonstrated a significant subsidence of ischaemic lesion on diffusion-weighted images (Fig. 2b). Four days later, Doppler ultrasound and MR angiography demonstrated complete restoration of blood flow within both ICAs (Fig. 3a, b).

The patient completely recovered with a NHSSS of 0 at discharge.

Discussion

Vasospasm of ICAs as a cause of stroke should be considered in patients with recurring ischemic events in the absence of any other causative factor, i.e. in patients with

✉ André Peeters
andre.peeters@uclouvain.be

Emmanuelle Levecque
emmanuelle.levecque@uclouvain.be

Pierre Goffette
pierre.goffette@uclouvain.be

Thierry Duprez
thierry.duprez@uclouvain.be

[1] Stroke Unit, Department of Neurology, Cliniques Universitaires Saint-Luc, Avenue Hippocrate 10, 1200 Brussels, Belgium

[2] Interventional Radiology Unit, Department of Radiology, Cliniques Universitaires Saint-Luc, Brussels, Belgium

[3] Neuroradiology Unit, Department of Radiology, Cliniques Universitaires Saint-Luc, Brussels, Belgium

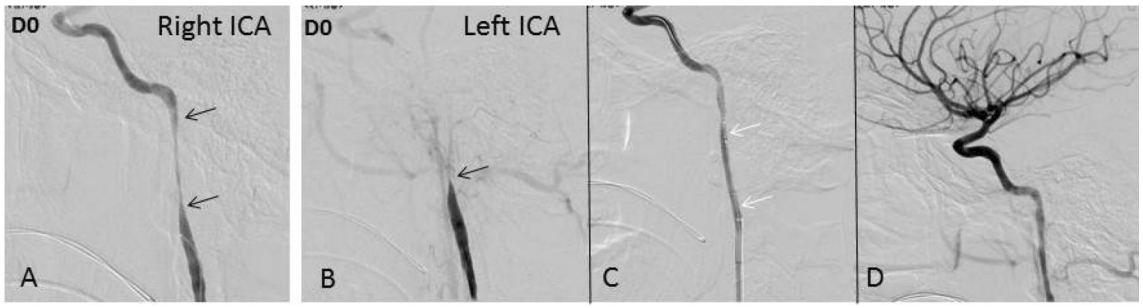

Fig. 1 Digitally subtracted angiography (DSA) with therapeutic intervention at day 0. **a** focal stenosis (between arrows) of extracranial segment of right ICA (lateral view). **b** Occlusion (arrow) of extracranial segment of left ICA (lateral view). **c** Blood flow restoration after easily passing the microcatheter through the stenosis. **d** Result after balloon angioplasty

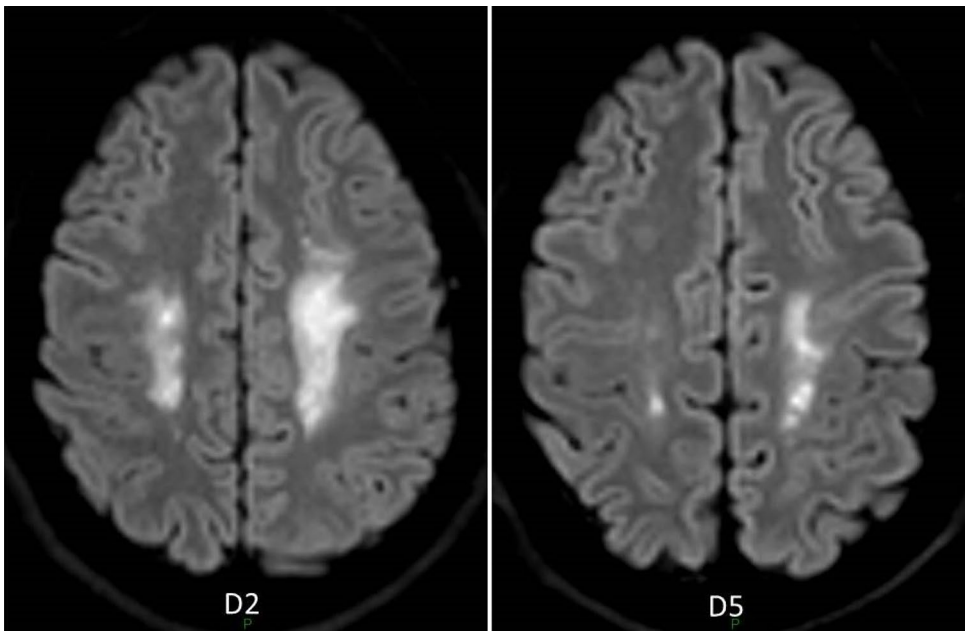

Fig. 2 Diffusion-weighted (DW) monitoring of brain ischemic lesions. **a** Axial transverse view obtained at D2 showing heterogeneous area of restricted freewater diffusion within both centrum semi-ovale. **b** Follow-up view in similar slice location obtained at D5 demonstrating dramatic lesions' subsidence

inconsistent duplex sonographic findings and/or when arterial dissection is hypothesized but no mural hematoma can be detected at MRI [2]. The stenosis looks like a segmental filiform narrowing or a distal carotid bulb that usually resolves spontaneously after 24–72 h. Huisa and Roy in their PubMed search found 11 articles comprising 12 cases. All patients had confirmed recurrent alternating cervical ICA stenosis with the exception of 1 fatal case with no follow-up. The average age at presentation was 32 years. Patients were usually female (69%) and had a history of migraines before developing focal neurologic deficits [1]. Disorders potentially causing vascular stenosis include cerebral vasoconstriction syndrome featured by vasospasm of almost always intracranial arteries with thunderclap headache. Primary angiitis of the central nervous system associated with granulomatous or lymphocytic vasculitis affects the leptomeningeal and cortical arteries and responds to steroids. Arterial dissection often occurs after traumatic events [4]. The fibromuscular dysplasia (FMD) is a pathological process with abnormal connective tissue, rendering the vessel wall vulnerable to repetitive vasospasm. It is a nonatherosclerotic non-inflammatory vascular disease that most commonly affects the renal and carotid arteries. Arteriography is the gold standard for defining the features of FMD, and many cases are incidental findings from arteriography. The most common findings are focal stenosis which appears smooth and band-like and multifocal, giving the "string of pearls" appearance on imaging. The pathophysiology of ICA vasospasm is not yet understood. Moeller et al. explored the possible pathomechanism of vasospasms; they performed a cold pressor test and they conclude that altered autonomic innervation at the affected

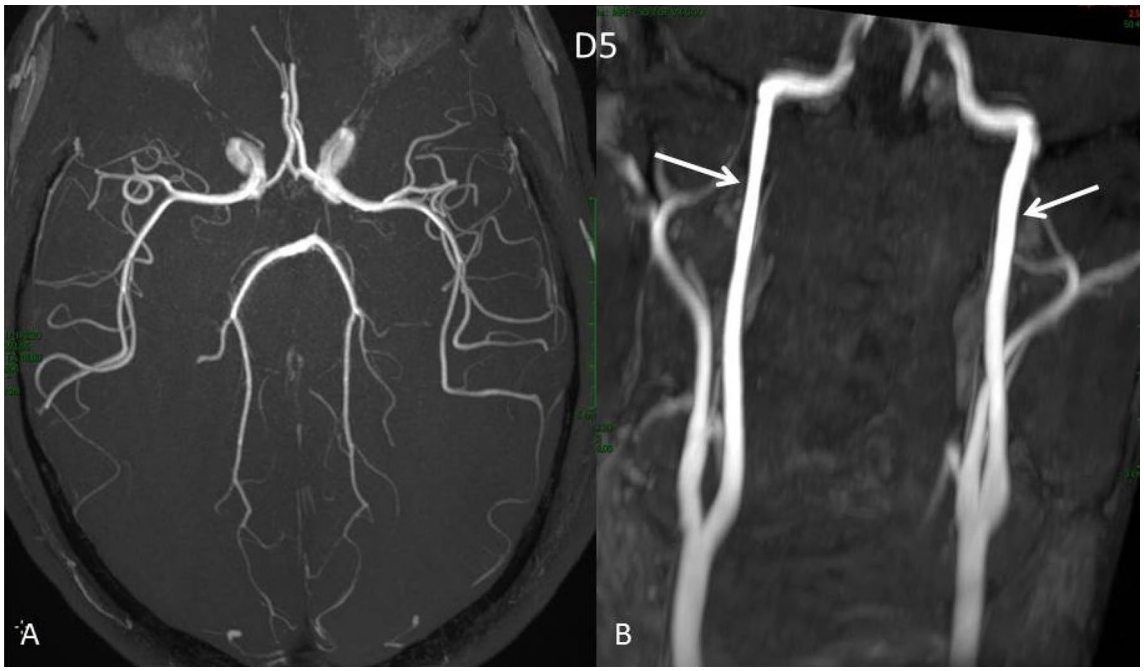

Fig. 3 MR angiograms at discharge. **a** Normal intra-cranial angiogram. **b** Normalized angiogram of both extra-cranial segments of ICAs (arrows)

intermediate ICA segment with upregulated sensitivity to sympathetic vasomotor stimuli contributed to vasospasm. The restriction of the vasospasms to the intermediate ICA portion might be due to an embryologic distinct development of the segment from the cranial portion of the embryonic dorsal aorta and a similarly distinct development of sympathetic vasomotor innervation in close correlation with the vascular system. Provided there was selective overexpression of sympathetic nerve terminals or vasomotor receptors in the cranial portion of the embryonic dorsal aorta, only the intermediate ICA segment would be more prone to vasospasm responses in face of sympathetic stimuli [3]. Treatment of isolated extracranial ICA vasospasm is not standardized [3]. Calcium antagonists alone seem to be insufficient in the treatment of vasospasm of the ICA. In the patient described by Arning et al. combined treatment with calcium antagonists and oral corticoids led to a reduction in the frequency of vasospasm [2].

This undescribed condition of idiopathic bilateral vasospasm of ICAs successfully treated with balloon angioplasty highlighted the need for (1) being aware of falsely "straightforward" diagnosis of ischaemic stroke due to dissection in a young; (2) considering the rare condition of 'idiopathic' vasospasm, mainly if radiological features and risk factors for dissection are missing; (3) treating such patients by the appropriate endovascular intervention.

Compliance with ethical standards

Conflict of interest The authors declare that they have no competing interests.

Ethical approval This article does not contain any studies with human participants (or animals) by any of the authors.

Informed consent For this type of study formal consent is not required.

References

1. Huisa BN, Roy G (2014) Spontaneous cervical carotid artery vasospasm: case report and literature review. Neurol Clin Pract 4(5):461–464
2. Janzarik WG, Ringleb PA, Reinhard M, Rauer S (2006) Recurrent extracranial carotid artery vasospasms: report of 2 cases. Stroke 37:2170–2173
3. Moeller S, Hilz MJ, Blinzler C, Koehn J, Doerfler A, Schwab S, Köhrmann M (2012) Extracranial internal carotid artery vasospasm due to sympathetic dysfunction. Neurology 78(23):1892–1894. https://doi.org/10.1212/WNL.0b013e318258f7ab
4. Sawa NN, Kataoka H, Ueno S (2013) Poor outcome associated with probable bilateral extracranial ICA vasospasm. Case Rep 2013:bcr2013009767