

CASE REPORT

Ocular ischaemia due to a spontaneous carotid artery dissection

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SUMMARY

Internal carotid artery dissection (ICAD) is caused by the disruption of the tunica intima, with the formation of an intramural haematoma that can cause stenosis or occlusion of the artery's lumen, leading to reduced blood flow and secondary thrombus formation. Up to two-thirds of patients with ICAD show ophthalmological symptoms or signs, which are, frequently, the first manifestations of this clinical condition, often preceding for weeks the neurological signs of cerebral infarction. Central retinal artery occlusion (CRAO) is a rare complication of ICAD, secondary either to haemodynamic compromise, with ocular hypoperfusion and reverse flow within the ophthalmic artery, or to thromboembolic events, in rarer cases. We report a case of CRAO secondary to a spontaneous ICAD, in an otherwise healthy middle-aged patient.

BACKGROUND

Internal carotid artery dissection (ICAD) can cause cerebral infarction in up to two-thirds of patients.¹ It accounts for up to 2.5% of first strokes in population below the age of 70 years,² with 70% of patients between the ages of 35 and 50 years.³ The most common clinical manifestations of ICAD are headache, neck pain, tinnitus, focal neurological deficits, Horner syndrome, carotid bruit and cranial nerve palsies.⁴ Up to two-thirds of patients with ICAD show ophthalmological symptoms or signs,⁵ which are frequently the first manifestations of this clinical condition, often preceding for weeks the neurological signs of cerebral infarction. The most frequent ocular sign of ICAD is ipsilateral painful Horner syndrome, which is caused by ischaemia or compression of the pericarotid sympathetic fibres. Other ocular manifestations are positive phenomena and scintillations, oculomotor nerve palsies, decreased visual acuity (VA) due to ischaemic optic neuropathy, ocular ischaemic syndrome or central retinal artery occlusion (CRAO).³ We report a case of CRAO and ischaemic optic neuropathy secondary to a spontaneous ICAD, in an otherwise healthy middle-aged patient.

CASE PRESENTATION

A healthy 50-year-old female patient, without medical history, presented with severe headache, global aphasia, right hemiplegia and right hemihypesthesia of sudden onset 5 hours earlier. Seven days before onset, she reported an episode of

headache and aphasia with spontaneous resolution. On physical examination, the patient's vital signs were: blood pressure, 145/86 mm Hg; heart rate, 75 beats/min and regular; respiratory rate, 18 breaths/min; and oxygen saturation was 99% on room air. Brain CT (figure 1) showed apparent loss of cortico-subcortical differentiation in temporo-insular and lenticulo-capsular regions. CT angiography was suggestive of occlusive dissection of the left internal carotid artery (LICA) (figure 2A).

Angiography (figure 2B) documented a tandem occlusion of the LICA cervical and supraclinoid segments, due to probable dissection. During angiography, aspiration thrombectomy was performed, with embolisation of a small thrombus, originated from the dissected area, to the distal posterior opercular branch. Thrombolytic agent recombinant tissue plasminogen activator was used with grade 2b reperfusion in the thrombolysis in cerebral infarction grading system (figure 2C).

The patient was admitted to the cerebrovascular unit. Brain MRI 48 hours after onset showed left acute ischaemic corticonuclear infarction of the insular and temporo-parietal cortex and lentiform and caudate nucleus, in the territory of the middle cerebral artery (MCA) (figure 3A,B).

Magnetic resonance angiography identified signs of arterial dissection extending from the post-bulbar segment of the LICA to the skull base, with homogeneous reduction of LICA calibre, but without significant focal stenosis (figure 3C). Transcranial doppler ultrasonography showed permeable MCA with haemodynamic changes compatible with type 4 reperfusion in the thrombolysis in brain infarction grading system. Carotid artery doppler ultrasonography showed moderate stenosis (50%–60%) of proximal LICA due to dissection, and severe stenosis (70%–80%) in the distal LICA. A thorough laboratory work-up was carried out, which was unremarkable (figure 4).

OUTCOME AND FOLLOW-UP

After partially regaining language function, 48 hours after the therapeutic intervention, the patient complained of decreased VA on her left eye (LE). It was not possible to assess VA due to the patient's poor collaboration. On inspection, she had normal photopupillary reflexes bilaterally, normal ocular motricity, without diplopia.

On fundus examination, the LE showed retinal oedema with a cherry-red spot macula, consistent



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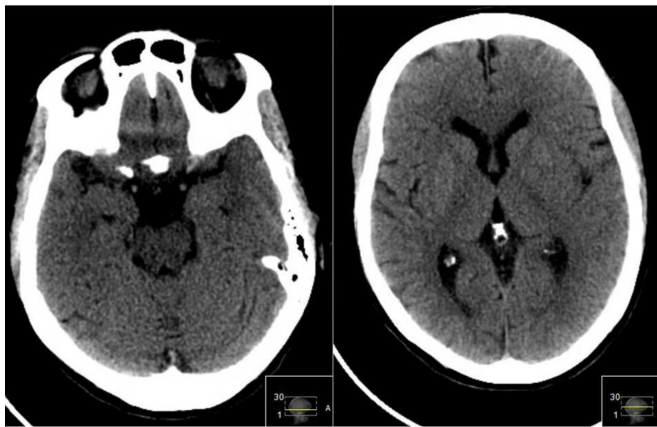


Figure 1 Brain CT showing temporo-insular and lenticulo-capsular loss of cortico-subcortical differentiation, translating an acute ischaemic lesion of the left middle cerebral artery territory.

with central retinal artery occlusion (CRAO), but no visible thrombus. The left ophthalmic artery was permeable on CDUS, with normal flow velocity, amplitude and direction, however, with an increased resistance index, due to a probable distal lesion. Three days later, CDUS showed normalisation of resistance index in the ophthalmic artery, suggesting haemodynamic improvement.

The patient showed progressive improvement on serial ultrasound studies and neurological function and was discharged 6 days after admission, on statin and antiaggregant therapy and indication for speech and language therapy. She was observed at the neuro-ophthalmology department, 2 months after the stroke, for follow-up. Best corrected VA was 20/20 in the right eye and 20/25 in the LE, a slightly reduced photopupillary reflex on the LE, normal ocular motricity and unremarkable biomicroscopy. Fundus examination revealed a slight pallor of the optic disc in

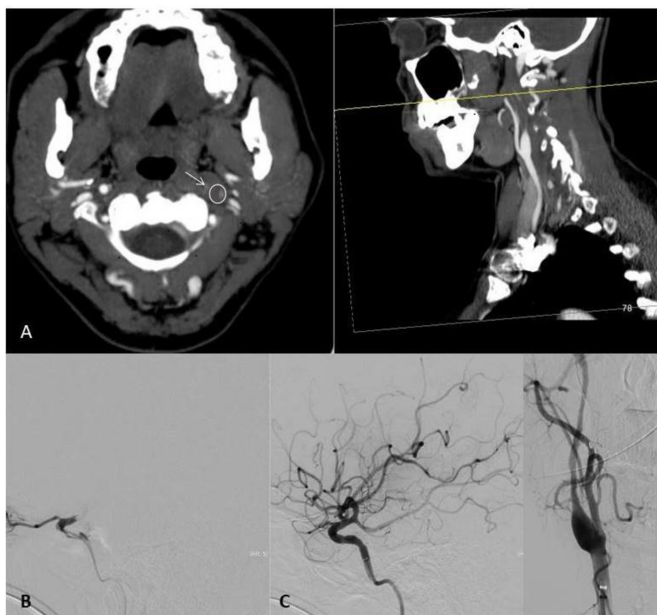


Figure 2 (A) CT angiography suggestive of occlusive dissection (indicated by the arrow and circle) of the left internal carotid artery (LICA). (B) Digital subtraction angiography showing a tandem occlusion of the cervical and supraclinoid segment LICA, due to probable dissection. (C) Reperfusion after percutaneous aspiration thrombectomy.

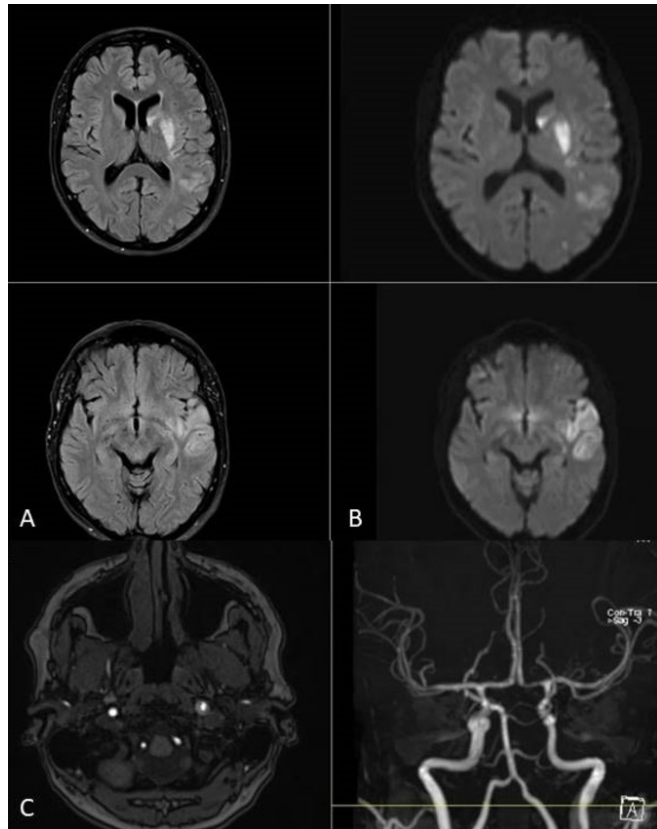


Figure 3 Brain MRI 48 hours after onset: (A). Fluid Acquisition Inversion Recovery (FLAIR) and (B) diffusion showing acute ischaemic infarction of the left middle cerebral artery territory, including the lentiform and caudate nucleus and temporal and parietal cortex (not included in this image). (C) Magnetic resonance angiography identifying signs of arterial dissection extending from the post-bulbar segment of the left internal carotid artery to the skull base, with double lumen signalling.

the LE (figure 5A). Fundus fluorescein angiography showed a discrete increase in foveal avascular zone in the LE (figure 5B). Optical coherence tomography (OCT) of the optic disc showed a decrease in nerve fibre layer thickness in the temporal, superotemporal and inferonasal peripapillary segments on the LE (figure 6B). Macular OCT revealed atrophy of the internal retinal layers, temporal to the fovea (figure 6A), consistent with a nasal campimetric defect on automated static perimetry (figure 7).

DISCUSSION

ICAD are classified as traumatic or spontaneous. Traumatic ICAD may be caused by minor trauma with neck hyperextension or rotation with shearing forces, when practicing athletic sports, valsava manoeuvres, coughing or vigorous nose blowing.⁴ Spontaneous ICAD may be associated with systemic hypertension, pheochromocytoma, Marfan syndrome, Ehler-Danlos syndrome, cystic medial necrosis and fibromuscular dysplasia.⁴ Atherosclerosis has not been reported as a risk factor for ICAD, as it is less frequent in the elderly atherosclerotic population.³

Histologically, ICAD is caused by the disruption of the tunica intima, leading to the accumulation of blood within the layers of the artery wall. This intramural haematoma can spread along the vessel and cause stenosis or occlusion of the artery's lumen leading to reduced blood flow. Virchow's triad events within the false lumen lead to secondary thrombus formation that can lead either

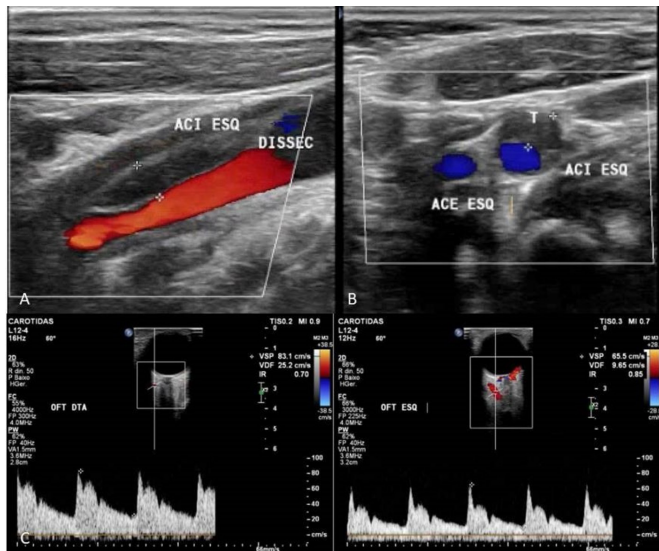


Figure 4 (A) Carotid artery doppler ultrasonography (CDUS) showed moderate stenosis (50%–60%) of proximal left internal carotid artery (LICA) with an intramural haematoma due to probable dissection, and severe stenosis (70%–80%) in the distal LICA. (B) The left ophthalmic artery was permeable on CDUS, with normal flow velocity, amplitude and direction, however, with an increased resistance index (0.85 contrasting with 0.7 on the right eye), due to a probable distal stenosis/occlusion.

to artery occlusion or embolisation to distal vessels.⁶ CRAO is a rare complication of ICAD, and its pathophysiology is believed to be, primarily, from haemodynamic compromise, with ocular hypoperfusion and reverse flow within the ophthalmic artery, to favour the supply of the brain. This reverse flow phenomenon is the reason why ocular embolism is a rare cause of CRAO in ICAD.^{3,5} Only two cases of CRAO due to embolic events have been reported,^{3,7} with visible thrombus within retinal vasculature. Nonetheless, the most frequent site of emboli in CRAO is thought to be the lamina cribrosa, and therefore, not seen in funduscopy or angiography.⁸ In the reported case, although no thrombus was found on retinal vasculature, the CDUS revealed a permeable left ophthalmic artery, with normal flow velocity, amplitude and direction but with an increased resistance index, due to a probable distal stenosis or occlusion. This finding suggests thromboembolic phenomena as the causative event of the CRAO in this patient, probably secondary to the embolisation of a thrombus originated from the dissected area during the percutaneous aspiration thrombectomy. Immediate fibrinolytic therapy is the probable reason for this patient's good outcome.

In a total of 15 reported cases of CRAO secondary to ICAD described in the literature, 10 cases did not involve previous trauma or head hyperextension and rotation.^{8,9} For this reason, it is of vital importance to include ICAD in differential diagnosis

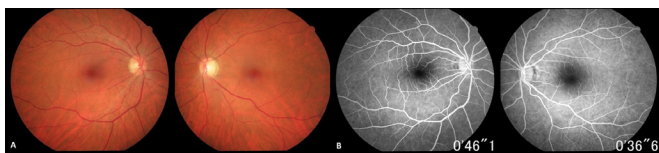


Figure 5 (A) Colour photograph of the ocular fundus, showing a discrete pallor of the left optic disc. (B) Fundus fluorescein angiography showing a discrete increase of foveal avascular zone diameter on the left eye.

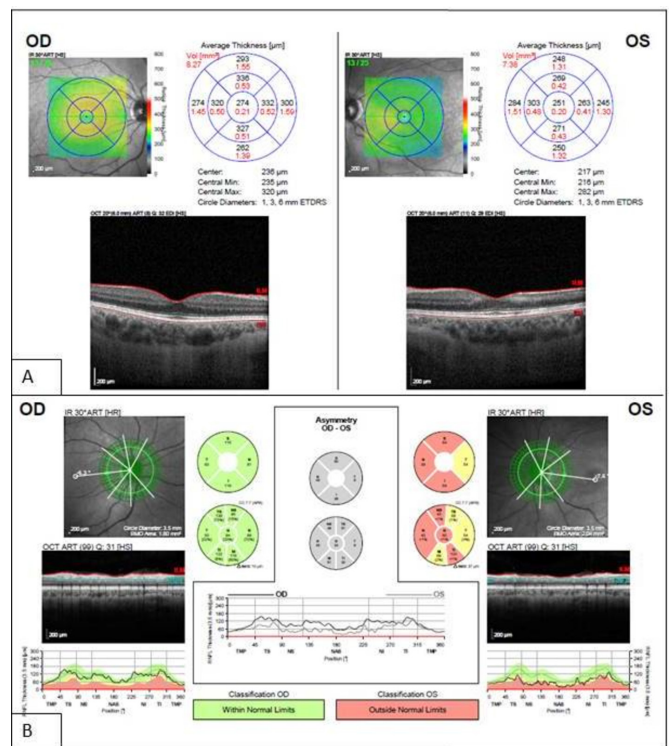


Figure 6 (A) Optical coherence tomography (OCT) of the macula revealed a decrease in retinal thickness temporal to the fovea due to atrophy of the internal retinal layers. (B) OCT of the optic disc showed a decrease in nerve fibre layer thickness in the temporal, superotemporal and inferonasal peripapillary segments on the left eye.

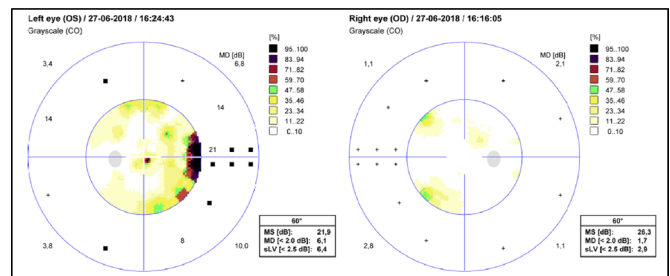


Figure 7 Automated static perimetry (*Octopus*) showed a nasal scotoma on the left eye visual field.

Learning points

1. Up to two-thirds of patients with internal carotid artery dissection (ICAD) show ophthalmological symptoms or signs, which are frequently the first manifestations of this clinical condition, often preceding for weeks the neurological signs of cerebral infarction.
2. Virchow's triad events within the false lumen lead to secondary thrombus formation that can lead either to artery occlusion or embolisation to distal vessels.
3. It is of vital importance to include ICAD in differential diagnosis of a central retinal artery occlusion or anterior ischaemic optic neuropathy, in young- and middle-aged patients, even in the absence of previous trauma or mechanical neck stress.

of a CRAO, in young and middle aged patients, even in the absence of previous trauma or mechanical neck stress.

Contributors SF: evaluated the patient and wrote the draft of the case report. CPdC: chose the neurological images and described them, to be included in the text and images legends. RPP: contributed on literature search and revising the paper. JTF: evaluated the patient, and contributed on literature search and revising the paper. All authors approve the version that is been submitted.

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