Ischemic Optic Neuropathy Associated With Internal Carotid Artery Dissection

Valérie Biousse, MD; Monique Schaison, MD; Pierre-Jean Touboul, MD; Jacques D’Anglejan-Chatillon, MD; Marie-Germaine Bousser, MD

Background: Ischemic optic neuropathy (ION) is an infarction of the anterior or, less frequently, posterior part of the optic nerve, usually due to a disease of small arteries supplying the optic nerve. Carotid stenosis or occlusions are rare causes, and among them, carotid dissections have been so far reported in only 5 cases.

Methods: We describe 4 patients with ION (2 anterior and 2 posterior) due to internal carotid artery dissection of a consecutive series of 110 patients with internal carotid artery dissection (3.6%).

Results: None of the patients had signs of central retinal artery occlusion or ischemic ocular syndrome. Ischemic optic neuropathy occurred after a mean of 5.3 days (range, 3-8 days) following the first symptom, which was headache in 1 patient, transient monocular blindness in 2, and hemispheric transient ischemic attack in 1. One patient had associated Horner syndrome, and 2 had severe ipsilateral headache and orbital pain. None of the patients developed a cerebral infarction. These features differ from those observed in “classic” nonarteritic anterior ION and might therefore point to carotid dissection.

Conclusion: Ischemic optic neuropathy may occur as an early sign of carotid dissection: young age, previous transient monocular blindness, an association with pain, Horner syndrome, or hemispheric transient ischemic attacks are suggestive of this cause and should prompt confirmatory investigations.

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RESULTS

Ischemic optic neuropathy was noted in 4 patients (3.6%): 3 women and 1 man (mean age, 44 years; range, 33-51 years). The dissection was spontaneous in all 4 patients. Their clinical presentation is summarized in the Table.

Among these 4 patients, 2 had AION and 2 had PION without any sign suggestive of central retinal artery occlusion or ischemic ocular syndrome. The permanent visual loss related to ION was never inaugural of the dissection: it occurred after a mean of 5.3 days (range, 3-8 days) following the first symptom, which was headache in 1 patient, transient monocular blindness in 2, and hemispheric transient ischemic attack (TIA) in 1. Later signs included Horner syndrome in 1 patient, severe ipsilateral headache and orbital pain in 2, and hemispheric TIAs in 1. None developed cerebral infarction. In 2 patients (patients 1 and 3), the visual loss was extremely severe, and 2 others (patients 2 and 4) had a permanent visual field defect with normal central visual acuity.

All patients had a tight stenosis of the extracranial ICA (80%-95%) extending up to the supraclinoid segment in 1. A retrograde filling of the ophthalmic artery was observed in 2 patients. All patients except 1 (patient 4) were treated with heparin sodium (followed by warfarin) in addition to strict bed rest and treatment to increase blood pressure when hemodynamic symptoms (such as postural transient monocular blindness and/or TIAs) were present. Angiograms and/or magnetic resonance angiography were repeated and showed ICA recanalization in all patients.

REPORT OF CASES

CASE 1

A previously healthy 33-year-old woman suddenly had transient paresthesia in the left hand. The next day, she complained of a severe diffuse headache, which lasted about 1 hour; she also had a transient monocular inferior visual field loss in the right eye, which resolved spontaneously in a few minutes, but recurred the next day. Four days later, she had a sudden complete visual loss in the right eye associated with a severe orbital headache.

Ophthalmologic examination showed a visual acuity of 20/15 OS but only light perception in the superior temporal quadrant of the right eye. The left pupil was normal, and the right pupil was larger than the left, and only minimally reactive to direct or consensual light stimulation. There was a right afferent pupillary defect. Funduscopic examination of the right eye showed optic disc edema, with dilated veins and a few peripapillary hemorrhages without retinal edema, cherry red spot, or intravascular embolic material. Goldmann visual field disclosed a small island in the superior temporal field of the
right eye (Figure 1). These findings were suggestive of AION. Findings of the left eye and the remainder of the neurologic and general examination were normal.

Cerebral angiogram showed a long irregular 95% stenosis of the infrapetrous portion of the right ICA, typical of extracranial ICAD, with retrograde filling of the ophthalmic artery from the right external carotid artery. She was treated with heparin followed by warfarin for 3 months. The ICA normalized but vision did not improve and disc atrophy developed.

**CASE 2**

A 41-year-old man complained of acute “cloudy vision” of the superior visual field of the right eye, which lasted 24 hours and resolved incompletely. Three days before, he had a right-sided headache and periorbital pain associated with drooping of the right upper lid and miosis.

Visual acuity was 20/20 OD and 20/15 OS with a permanent “cloud” on the superonasal visual field of the right eye. Pupil examination disclosed a right Horner syndrome. On the right side, funduscopic examination showed mild optic disc edema without retinal or vascular abnormalities. Goldmann visual field showed a superior altitudinal defect (Figure 2). These findings were suggestive of AION. The left eye was normal, as were the results of the remainder of the general and neurologic examination.

Carotid duplex scanning and cerebral angiogram showed a 80% stenosis of the cervical portion of the right ICA extending up to its suprACLidoid segment, and a reduction of blood flow within the ophthalmic artery. The left ICA was normal with cross-filling of the anterior and middle cerebral artery. Heparin treatment was started, followed by warfarin. The headache completely resolved within 2 weeks. Five months later, cerebral angiogram was normal, the Horner syndrome had disappeared and visual acuity was normal. However, the visual field defect persisted and there was a mild segmental disc pallor in the right eye.

**CASE 3**

A 51-year-old woman had 4 brief episodes of transient monocular blindness in the left eye in 1 day. Results of ophthalmologic examination performed on the same day were normal. Two days later while standing up, she had a transient right hemiparesis associated with aphasia that resolved in 15 minutes. These symptoms recurred every time she tried to stand up.

On admission, ophthalmologic and neurologic examination results were normal except for decreased sensation in the left fronto-orbital region. A brain computed tomographic scan was normal. Carotid duplex scanning and cerebral angiogram showed a 95% stenosis of the whole cervical segment extending into the petrous portion of the left ICA with a retrograde filling of the ophthalmic artery from the external carotid artery, and a low flow in the middle cerebral artery. Treatment with heparin was started in addition to strict bed rest. However, 3 days later, while raising her head, she suddenly had a transient inferior visual field loss in the left eye, followed by a complete visual loss. Ophthalmologic examination showed no light perception in the left eye, and a dilated nonreactive pupil to direct or consensual light stimulation. Optic disc, retina, and retinal arteries were normal. This was suggestive of PION. The following days, she experienced multiple transient episodes of right hemiparesis with dysphasia, occurring whenever she tried to raise her head. Treatment was initiated to increase blood pressure. Cerebral angiogram was normal 3 months later, but the blindness in the left eye persisted with optic atrophy.

**CASE 4**

A 51-year-old woman with a history of hypertension suddenly had a right-sided monocular visual loss that partially resolved in a few minutes. It was followed by tingling and numbness in both hands and a brief loss of consciousness.

Recurrent episodes of transient hand weakness and numbness on either side occurred during examination, associated with nausea and tinnitus of the right ear. There was a permanent blurring of vision of the superior part of the visual field of the right eye. Reflexes were brisk with a bilateral extensor response. Funduscopic examination results were normal. There was no detailed ophthalmologic examination at that time.

### Table: Cases 4-7

<table>
<thead>
<tr>
<th>Other Symptoms or Signs</th>
<th>Presumed Mechanism</th>
<th>Inaugural Symptom or Sign of the Dissection</th>
<th>Time Between First Symptom and ION or Cerebral Infarction, d</th>
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<tr>
<td>Recurrent Dilation TIA</td>
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<td>Recurrent Dilation</td>
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<td>Normal Infarction</td>
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<td>1 d between pain and PION:</td>
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<td>2 d between PION and stroke</td>
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A computed tomographic scan of the brain was normal. Cerebral angiogram showed an irregular stenosis of the intracranial segment of the right vertebral artery and a 90% stenosis of the infrapetrous segment of the right ICA, with a small ICA aneurysm in the left cavernous sinus. She was treated with aspirin and had no further neurologic episode. Some blurring of vision persisted in the right eye. Ophthalmologic examination 8 months later showed normal visual acuity in both eyes with normal pupils. Funduscopic examination showed segmental inferior disc pallor in the right eye without any abnormality of the vessels or retina. Confrontation visual field and Amsler grid suggested a persistent altitudinal defect in the superior visual field of the right eye. Ocular signs both initially and at follow-up were suggestive of PION.

This study describes 4 cases of ION (2 AION and 2 PION) consequent to spontaneous dissection of the ipsilateral ICA of a consecutive series of 110 patients with ICAD (3.6%). The diagnosis of ION was based on sudden monocular visual loss in the absence of retinal and vascular abnormalities on fundus examination, associated with a sectorial visual field defect, and either disc edema (AION) or a normal disc (PION) initially. Of course, it does not preclude choroidal ischemia. However, the follow-up funduscopy that showed optic disc pallor without retinal pigment changes is more suggestive of ION.

Five cases of presumed ION have been reported in patients with ICAD (Table).12-16 but the diagnosis is probably overlooked particularly when visual acuity remains good, or when there is a severe stroke with dysphasia or disorder of consciousness. Our patients were specifically asked about the presence of visual symptoms and this probably reflects the high frequency of ION in our series. Newman et al13 reported 1 case of AION related to ophthalmic artery occlusion. Rivkin et al16 described a man who presented with a painful PION followed 2 days later by a massive cerebral infarct. McNeill et al14 described a 48-year-old woman who had 14 episodes of “monocular visual blurring” exacerbated by “bending or stooping” with “an indistinct temporal disc margin on funduscopy” as the only signs of a left extracranial ICAD. The 2 other cases are merely mentioned but without any ophthalmologic details in 2 large series of ICAD, one of 30 patients with stroke,13 and one of 23 patients with cranial nerve palsy.12

Some clinical features of ION in patients with ICAD differ from those observed in “classic” nonarteritic AION. (1) The mean age is younger in ICAD: 44 years (range, 33-51 years) vs 60 years (range, 11-90 years).3 (2) Severe ipsilateral orbital pain is a prominent symptom of ICAD: it was present in 2 of our patients and in 2 of the 3 detailed published cases.15,16 and preceded ION by 3 to 5 days. Indeed, cephalic pain is the most frequent inaugural symptom in ICAD (up to 95% of patients) and is usually unilateral, ipsilateral to the dissection.17 Pain is so moderate and rare (8%-12% of cases)18,20 in nonarteritic AION that it is a major criterion to differentiate AION from inflammatory optic neuritis,4,18,20 and that it was considered as an exclusion criterion for the recent Ischemic Optic Neuropathy Decompression Trial.4,18 Thus, ION associated with severe cephalic pain in a young patient should immediately point to ICAD. In older patients, the presence of pain associated with ION should raise the hypothesis of giant cell arteritis; however, the usual presence of systemic symptoms and the elevation of the sedimentation rate easily point to this diagnosis. (3) Previous recurrent episodes of transient monocular blindness were reported by 3 of our patients and by 2 published cases.14,15 They occurred from 3 to 8 days (mean, 5.3 days) before permanent visual loss related to ION. By contrast, they were reported by only 5% of patients with nonarteritic AION included in the Ischemic Optic Neuropathy Decompression Trial.4 (4) Horner syndrome is so common in ICAD (up to 58% of cases)11 that its association with ION, such as in our second patient, should immediately point to the diagnosis of ICAD.

It is likely that the mechanism of ION in ICAD is the reduction of ocular blood flow caused by a sudden decrease in the caliber of the true lumen of the ICA in our patients. Indeed, our 4 patients had a severe stenosis of the ipsilateral ICA associated with decreased flow in the ophthalmic artery, which was reversed in 2 patients. In patient 3, as well as in the patient described by McNeill et al,14 the visual symptoms were related to changes of posture, which is highly suggestive of a hemodynamic mechanism. Moreover, no case has been reported with associated visible retinal emboli. None of our patients had signs of venous stasis retinopathy or ischemic ocular syndrome, which are a hallmark of chronic hypoperfusion of the eye. Preserved supply through choroidal anastomoses during the early phase and rapid re- canalization of the ICA in all our patients may explain the relative retinal sparing. However, in patients 1 and

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3, an ipsilateral pupillary dilation with poor or absent reactivity to direct or consensual light stimulation was observed. Newman et al have reported the same phenomenon in ICAD and have suggested that it may reflect ischemia to the ciliary ganglion or to the iris, suggesting a prolonged hyperperfusion of the eye. Indeed, these 2 patients had a severe visual loss and hemispheric TIAs. It also has been suggested that morphologic factors of the disc such as small “crowded” disc and small or absent cup might play a role in the pathogenesis of nonarteritic AION. It is possible that such an anomaly of the optic disc may be a predisposing factor for ION in ICAD. However, we do not have this information either in our patients or in the 5 previously reported cases.

Despite the absence of treatment of proven efficacy for both ICAD and ION, we administered heparin to 3 of our patients with ION to decrease the risk of subsequent ischemic stroke, which is present up to 1 month after the first symptom of dissection. Because ION was presumed to be related to decreased perfusion pressure, we sought to improve perfusion to the eye and brain by favoring strict bed rest, by eventually stopping antihypertensive agents, and, in 1 patient who had recurrent hemodynamic symptoms, by raising blood pressure. This treatment did not restore vision in the 2 patients with severe visual loss and its potential preventive effect on further cerebral ischemia is impossible to assess. However, in contrast to 2 of 3 previously reported cases, none of our patients developed a cerebral infarct.

Internal carotid artery dissection is a possible, though rare, cause of ION. It should particularly be considered when associated with pain, recurrent transient monocular blindness, and Horner syndrome in a young patient. In such cases, clinicians should urgently perform noninvasive vascular explorations (ultrasound, magnetic resonance imaging, and magnetic resonance angiography) and initiate treatment to try to prevent a cerebral infarction, the major complication of ICAD.

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Corresponding author: Valérie Biousse, MD, Neurology Department, Hôpital Lariboisière, 2 rue Ambroise Paré, 75010 Paris, France (e-mail: biousse@ccr.jussieu.fr).