Temporal Arteritis-Like Presentation of Carotid Atherosclerosis

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SUMMARY A 68 year-old woman presented with a two-week history of amaurosis fugax, ipsilateral fronto-temporal headache and jaw claudication suggesting carotid giant cell arteritis. However, this syndrome proved to be due to atherosclerosis causing complete occlusion of the external carotid artery at its origin and narrowing of the internal carotid artery. Combined external and internal carotid endarterectomy relieved the symptoms. The symptom complex of temporal arteritis may be rarely mimicked by carotid atherosclerotic occlusive disease.

THE CONSTELLATION OF NEW-ONSET HEADACHE, ipsilateral visual impairment and jaw claudication in an elderly person is considered virtually diagnostic of carotid giant cell arteritis (GCA) usually referred to as temporal arteritis. The diagnosis is supported by high ESR and confirmed by superficial temporal artery (STA) biopsy and urgent and long-term therapy with corticosteroids is instituted to prevent blindness and other neurological sequelae.1 We have recently encountered a patient with this clinical syndrome in whom the underlying disease was not GCA but atherosclerosis of the external and internal carotid arteries (ECA, ICA).

Report of a Case
A 68 year-old black woman complained of two episodes of dimming of vision in the right eye over the 2 weeks prior to admission. Each episode came on abruptly and resolved in 15–20 minutes and was described as a shade coming over the eye. After the second episode she noted a moderately severe throbbing pain above the eye and the temple which lasted several hours, responded to aspirin but recurred on and off. During these 2 weeks, she experienced pain in the right side of the jaw on chewing usual food. The dull ache began in the lower jaw and spread to the upper jaw and the temple. It was severe enough to stop eating which relieved the pain. There were no symptoms indicating dysfunction of the cerebral hemispheres or brainstem, and she denied malaise, body aches and pains, anorexia or weight loss. She had a history of treated mild hypertension and severe coronary atherosclerosis causing unstable angina for which a coronary artery bypass graft had been performed 2 years earlier. At that time an asymptomatic bruit over the left carotid artery had been noted.

On examination the blood pressure was 130/70mm Hg. There was a high-pitched holosystolic bruit maximal over the bifurcation of the right common carotid artery (CCA) but the previously noted left carotid bruit was not heard. The carotid arteries were not tender in the neck. The right superficial temporal and the facial artery pulses were absent without tenderness, thickening of the vessel wall or changes in the overlying skin. These pulses were present on the left. Examination of the mental state, cranial nerves and motor, sensory, reflex and gait functions revealed no abnormalities. In particular, corrected visual acuity, visual fields and the appearance of the ocular fundi were normal. There was no Horner’s syndrome.

The ESR was less than 20mm on repeated testing. Complete blood count and blood sugar were normal. On angiography, the right external carotid artery was completely occluded at its origin while the distal common carotid and the origin of the internal carotid arteries showed tight focal stenosis (fig. 1). The right ophthalmic artery was patent. On the left there was mild stenosis at the origins of the internal and external carotid arteries. The right middle cerebral artery did not fill by cross-flow from the left carotid injection.

An endarterectomy was performed on the right common carotid artery and extended into the external and internal carotid arteries using an indwelling shunt. A considerable amount of atherosclerotic material was removed from the ECA until there was good back bleeding. This material, on microscopic examination, consisted of calcified atheromatous plaque with superimposed organised and partially revascularised thrombus.

The patient experienced cessation of jaw claudication as soon as she resumed eating solid food after the surgery. The superficial temporal and facial pulses, previously impalpable, returned. In a follow-up period...
FIGURE 1. Subtraction film after right common carotid injection demonstrates stenosis at the origin of the ICA and the arrow points to total occlusion of the origin of the ECA.

of 15 months she remained asymptomatic and had been receiving aspirin.

Discussion

The clinical picture of our patient suggested GCA. Transient monocular blindness lasting minutes to hours indistinguishable from amaurosis fugax of carotid atherosclerosis, has been reported in GCA and in some cases heralds permanent loss of vision in one or both eyes.2-3 Intermittent claudication of the jaw is considered to be pathognomonic of GCA.4 Polymyalgia rheumatica may be absent or appear later in the course of the disease. ESR of less than 40mm has been reported in about 30% of biopsy-proven cases of GCA.5 However, these negative features combined with a prominent bruit over the appropriate carotid bifurcation, the history of an asymptomatic bruit over the other carotid artery, and the complete absence rather than segmental decrease of facial and superficial temporal artery pulses raised the possibility of atherosclerotic occlusive disease of the ECA an the ICA. Angiography, surgery, histopathology of the vessel and the course after endarterectomy confirmed this diagnosis.

Jaw claudication, most often due to GCA, has rarely been reported with amyloidosis and arteritis of the facial artery.6 Jaw pain or claudication occurred in 3 of 88 patients who were suspected of having GCA but who had negative temporal artery biopsies and were followed long-term. However, the final etiology of the claudication in these 3 patients was not stated.7 A case of hemifacial pain for 4 weeks preceding contralateral hemiplegia due to total occlusion of the ICA and the facial artery has been reported but its etiology was not mentioned and intermittent claudication of the jaw was not described.8 In our patient the headache and jaw claudication are attributable to ECA occlusion and the amaurosis fugax to the ICA stenosis. We could find no previous reports of jaw claudication from atherosclerotic occlusion of the ECA or of the symptom complex of headaches, jaw claudication and amaurosis fugax from combined atherosclerosis of the ECA and the ICA.

The manifestations of ECA atherosclerosis are not well known although atherosclerosis is as common in the ECA as in the ICA.9 It is usually asymptomatic but is a source of bruits over the carotid bifurcation.10 ECA disease may cause retinal or hemispheric TIAs when the ipsilateral ICA is occluded.11-15 Embolism from the ECA atheroma through the anastomoses with the ophthalmic and middle cerebral arteries or a critical fall of the perfusion pressure in these channels have been the proposed mechanisms of the symptoms which are reported to be relieved by external carotid endarterectomy. Exceptionally, ECA atherosclerosis causes TIAs even when the ICA is not narrowed: A patient experienced recurrent amaurosis fugax with a completely occluded ECA but with an ICA which was patent as a result of an earlier endarterectomy.16 The TIAs were felt to be caused by emboli from the ECA stump entering the ICA circulation. In another patient with severe ECA atherosclerotic stenosis and fully patent ICA, recurrent amaurosis fugax was considered to be due to emboli through the ophthalmic artery which had an anomalous origin from the middle meningeal branch of the ECA.17 External carotid endarterectomy abolished the symptoms in both cases.

GCA affects the external and the internal carotid arteries in a pattern different from atherosclerosis. In the ECA system, the individual branches, especially the STA, rather than the origin of ECA are affected. In the ICA the ophthalmic artery and its posterior ciliary branches are most frequently and most severely affected. In the rare cases where the ICA stem is involved, the cavernous portion bears the brunt of the burden. An exceptional case of bilateral occlusion of ICAs 2cm distal to their origins has been described.19 We believe our case underscores the need for a positive diagnosis to be established in patients presenting with new onset headaches, amaurosis fugax and jaw claudication before embarking on long-term corticosteroid therapy for presumed GCA. The Mayo Clinic group recommends that when temporal arteritis is suspected it should be confirmed by adequate STA biopsies because none of the clinical features despite persistently high ESR is pathognomonic of GCA.6-7 Only 8 of 88 such biopsy-negative patients developed GCA in a median follow-up of 70 months.7 In such instances, alternative diagnoses should be sought, among them atherosclerosis of the ECA and the ICA. Long history of retinal or cerebral TIAs, carotid bifurcation bruits, complete absence of facial and superfi-
cial temporal artery pulses rather than their segmental loss, coexisting atherosclerosis elsewhere, hypertension and diabetes provide supportive evidence for atherosclerotic etiology. In selected cases as in our patient, carotid angiography rather than STA biopsy may be the most appropriate confirmatory test. Angiography may reveal segmental arteritic lesions even in GCA but biopsy remains its definitive test. We also suggest that in the older patients presenting with atypical facial pain or lower half headache syndrome along with other evidence of arteriopathy, ECA atherosclerotic occlusive disease be added to the differential diagnosis.

References
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