Pearls & Oy-sters: Resolution of hemichorea following endarterectomy for severe carotid stenosis

I. Galea, PhD
F. Norwood, PhD
M.J. Phillips, PhD
C. Shearman, MS
P. McMonagle, PhD
W.R. Gibb, MD

Address correspondence and reprint requests to Dr. Ian Galea, Neurology Department, Wessex Neurosciences Centre, Mailpoint 101, Level B, Southampton University Hospital Trust, Tremona Road, Southampton SO16 6YD, UK
I.Galea@doctors.org.uk

We report three patients with hemichorea who were found to have contralateral atherosclerotic carotid artery stenosis. Their details are compared in the table. All patients had carotid duplex ultrasonography because they exhibited neurologic signs or symptoms suggestive of cerebrovascular ischemia in addition to chorea.

Patient A developed right hemichorea; 9 months later she experienced three episodes of right limb weakness and speech disorder lasting 5 minutes, suggestive of transient ischemia. The severity of chorea increased and it mildly affected the left side. Neurologic examination revealed predominantly right-sided chorea and mild orofacial dyskinesia without other neurologic signs. The following blood tests were normal: full blood count, inflammatory markers, coagulation studies, renal, glucose, bone, liver, and thyroid biochemistry, immunoglobulin electrophoresis, anticardiolipin antibodies, and antinuclear antibodies screen (including extractable nuclear antigens). CT of the head was normal. Carotid duplex ultrasonography revealed 90% stenosis in the left internal carotid artery.

Patient B developed right hemichorea and 3 months later expressive dysphasia and right facial and hand weakness; this precipitated admission. In addition to chorea, there was right visual inattention and upper motor neuron weakness affecting the right face and hand with normal reflexes and plantar responses. The same blood tests as for patient A were normal; in addition, treponemal serology was consistent with past infection. MRI of the brain showed a left-sided parietal lobe infarct which was limited to the gray matter of the posterior parietal lobe suggestive of watershed infarction. Carotid duplex ultrasonography revealed 90% stenosis in the left internal carotid artery.

Patient C presented with right-sided upper limb cortical sensory loss (dysgraphesthesia, astereognosis, and joint position sense loss) and within a month had developed right hemichorea. A blood test panel as described above was normal. In addition, the following investigations were normal or negative: treponemal serology, copper, ceruloplasmin, antistreptolysin titers, rheumatoid factor, and anticytoplasmic nuclear antibodies. CSF examination was performed: cell count, protein, and glucose were within normal limits, herpes simplex virus PCR was negative, and oligoclonal bands were absent. MRI of the brain showed a left-sided parietal lobe infarct which involved the post-central gyrus and the underlying parietal white matter. Carotid duplex ultrasonography revealed 90% stenosis in the left internal carotid artery.

All three patients proceeded to carotid endarterectomy with complete resolution of the chorea. Neurologic signs in patient B resolved, but right-sided cortical sensory loss persisted in patient C.

DISCUSSION New onset chorea frequently triggers referral to neurology departments. Its several causes include metabolic, vascular, structural, autoimmune, and genetic.[1] Diagnostic workup therefore usually includes a detailed family and drug history, a general and neurologic examination, selected blood tests, molecular genetic testing, and neuroimaging. However, carotid duplex ultrasonography is not a usual investigation and is not mentioned as a potential diagnostic test in recent reviews of chorea.[1,2] In our cases, we cannot exclude the possibility of coincidence to explain the association of carotid artery stenosis and chorea, but we present evidence of an association and discuss possible pathophysiology.

There are few published series examining the vascular causes of new onset chorea. A recent study found various ischemic, and one hemorrhagic, lesions in 21 of 51 (41%) consecutive cases of sporadic chorea.[3] Some of these lesions were in basal ganglia or thalami but they were thought to be the cause of chorea in only two patients. Carotid duplex ultrasonography was not reported so it is possible that some had undetected carotid artery stenosis.

From Wessex Neurosciences Centre (I.G., P.M., W.R.G.) and Department of Vascular Surgery (M.J.P., C.S.), Southampton General Hospital; and King's College Hospital (F.N.), London, UK.

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Table Characteristics of the three cases

	Patient A	Patient B	Patient C
Age	72 years	75 years	73 years
Gender	Female	Male	Male
Semiology of chorea at onset	Right arm > leg	Right arm & leg	Right arm & leg
Duration of chorea before CEA	17 months	3 months	1 month
Time course of onset	Acute	Acute	Subacute
Medication at presentation	Aspirin, ramipril, simvastatin	Aspirin, nebivolol, lansoprazole	Thyroxine, nifedipine
Family history of movement disorders	None	None	None
Other neurologic signs	None	Right cortical sensory loss, right UMN weakness face & hand	Right cortical sensory loss
Carotid bruit	None	None	Left carotid bruit
Blood test panel	All normal	All normal	All normal
CVD risk factors	IHD, HT, PVD, smoker	HT, ex-smoker, FH	HT, FH
Carotid stenosis on duplex ultrasonography	90% left ICA, 25–50% right ICA	90% left ICA, <25% right ICA	90% left ICA, <25% right ICA
Brain imaging	CT normal	MRI: left posterior parietal gray matter infarct	MRI: left anterior parietal lobe infarct
Response to antichoreic agent	Partial to haloperidol	Not tried	Not tried
Recovery of chorea after carotid endarterectomy	Full	Full	Full
Time to resolution of chorea	6 months	2 weeks	Immediately
Follow-up period since carotid endarterectomy	4 years	5 years	1 year

CEA = carotid endarterectomy; UMN = upper motor neuron; CVD = cerebrovascular disease; IHD = ischemic heart disease; HT = hypertension; PVD = peripheral vascular disease; FH = family history; ICA = internal carotid artery.

In patients presenting to hospital with stroke, the incidence of chorea following the onset of stroke is small, with figures of 0.4%[4] and 1.3%[5] in studies of 2,500 and 1,500 patients. Radiologic evidence of infarction in basal ganglia or thalami in some of these cases was likely to be relevant to the cause of chorea, but information on carotid duplex ultrasonography was not presented.

Our cases suggest an alternative relationship between chorea and vascular disease. Chorea preceded symptoms of left hemisphere ischemia (dysphasia and right hemiparesis) in the first two cases. In the third case, chorea followed signs of cortical sensory loss. There was no radiologic evidence of basal ganglia or thalamic infarction but carotid duplex ultrasonography revealed 90% stenosis in the left carotid artery in all cases and the chorea resolved following left carotid endarterectomy. In the third case, carotid endarterectomy resulted in complete resolution of the chorea but not of the cortical sensory loss, indicating that the parietal infarct was not causative.

Carotid endarterectomy reduces the absolute risk of stroke in symptomatic[6] and more modestly in asymptomatic[7] carotid artery stenosis, with an absolute risk reduction of 16% and 7% over 5 years. It is therefore likely that patients presenting with chorea who have carotid artery stenosis will also benefit from a reduction in future stroke risk as a result of revascularization. In our patients, early recognition of chorea as a marker of possible cerebrovascular ischemia may have prevented subsequent ischemic events. We suggest vascular imaging (carotid duplex ultrasonography, MR or CT angiography) in patients presenting with unilateral chorea irrespective of the presence or absence of symptoms, signs, and imaging evidence of cerebrovascular ischemia. This should also be considered in patients presenting with asymmetric and generalized chorea, as unilateral structural lesions may cause bilateral chorea presumably due to a proportion of corticospinal tract fibers running ipsilaterally, and bilateral stenosis is also possible.

Carotid occlusive disease risks blood flow in lenticulostriate branches of the middle cerebral artery. Ischemia in this basal ganglia territory presents the most likely reason for chorea, and a similar mechanism may be relevant to other vascular causes such as cardiolipin antibody syndrome and polycythemia. In patient A, chorea was the only clinical sign while in the other two patients chorea dominated the clinical

picture, accompanied by subtle neurologic signs. This may be due to striatal neurons being particularly vulnerable to ischemia.[8]

In all patients, there was complete resolution of chorea after revascularization, suggesting chorea was caused either by embolization or hypoperfusion. Embolization is unlikely given the normal imaging of the basal ganglia and thalamus, and the reversibility of the chorea in our patients. Striatal hypoperfusion is more likely and in two recently reported cases similar to ours, cerebral blood flow studies supported this explanation.[9] Surgical revascularization was accompanied by cessation of chorea and normalization of cerebral blood flow on SPECT.[9]

It is interesting that the speed of recovery of chorea was inversely proportional to the duration of chorea before surgery (table). The delayed recovery in patients waiting longer for surgery suggests a degree of synaptic plasticity during the period of hypoperfusion. Interestingly, in vitro studies have shown that transient ischemia may have both acute and long-term effects on the excitability of GABAergic striatal neurons.[10]

Carotid artery stenosis is an underrecognized association with new onset chorea. Vascular imaging by carotid duplex ultrasonography or MR/CT angiography is recommended since its detection may allow treatment of chorea and prevent cerebral infarction. Further research with blood flow studies is warranted.

Clinical pearls. Carotid artery stenosis should be considered in the differential diagnosis of unilateral chorea, even in the absence of preceding presentation with stroke or transient ischemic attacks.

Carotid endarterectomy may reduce contralateral chorea in patients with critical carotid artery stenosis.

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