Transient Aortic Arch Syndrome with Dysphasia Due to Ergotism


SUMMARY A 55-year-old woman with an aortic arch syndrome of acute onset and dysphasia, resulting from the excessive administration of ergot derivative, is described. Complete resolution of symptoms and return of upper limb and carotid artery pulses to normal occurred within four days of cessation of the ergot derivative. Lower limb involvement was conspicuously absent in this case.

Stroke, Vol 14, No 5, 1983

ARTERIOSPASTIC DISEASE resulting from the excessive use of medicinal ergot preparations typically involves the lower limbs or less commonly, the mesenteric vessels. Although upper limb involvement has been described previously,† selective upper limb involvement with complete sparing of the lower limbs has been only rarely reported.3-5 To our knowledge, there have been only four angiographically documented cases of ergot-induced carotid arteriospasm.5-8

We report the case of a 55-year-old woman with transient, severe, bilateral brachial and internal carotid artery stenoses and expressive dysphasia due to the excessive use of ergotamine tartrate suppositories, but with no evidence of lower limb arteriospasm.

Case Report

A 55-year-old woman was referred with a three day history of dizziness and headache of sudden onset. Physical examination revealed absent pulses and unrecordable blood pressure in both upper limbs, which were cool with mild cyanosis of the hands and loud bilateral carotid bruits. The left carotid pulse was not palpable and the bruit on this side had both systolic and diastolic components. The lower limbs were warm and well-perfused and all lower limb pulses were normal. There were no abdominal or femoral bruits. The carotid examination, electrocardiogram and chest X-ray were normal. In particular, there was no evidence of mediastinal enlargement. There was no clinical or laboratory evidence or rheumatoid arthritis, ankylosing spondylitis or other rheumatic disorder. The erythrocyte sedimentation rate was 30 mm/hour. Neurological examination was initially normal, but on the second hospital day expressive dysphasia was noted without other neurological deficit.

Doppler orbital blood flow studies showed reversed flow on the right side and low amplitude flow on the left side. Oculoplethysmographically determined ophthalmic artery pressures were 70 mm Hg on both sides. Arch aortography revealed that the aorta and proximal brachiocephalic vessels were normal, but selective angiography demonstrated bilateral internal carotid artery stenoses (fig. 1) and complete occlusion of the left brachial artery (fig. 2). All these stenoses were noted to be long, smooth and tapering. Angiograms of the right brachial artery were not performed because the right brachial artery was palpable at the time of angiography, as was the left carotid pulse, suggesting that the angiographic appearances underestimate the severity of the carotid stenoses present at the time of admission.

On the third hospital day, the expressive dysphasia resolved completely, no carotid bruits were audible and all upper limb pulses were palpable. By the fourth hospital day, the upper limb pulses were normal. Doppler orbital blood flow studies repeated one week after admission were normal and demonstrated brisk augmentation following superficial temporal artery compression. The ophthalmic artery pressures were 104 mmHg on the right side and 102 mmHg on the left side. Brachial artery systolic pressures were both 120 mmHg.

Despite the selective involvement of branches of the aortic arch, completely sparing branches of the descending aorta and particularly the lower limb vessels, the angiographic appearances and the patient’s rapid and complete recovery were strongly suggestive of an arteriospastic disorder. The patient did in fact, give a history of recurrent headaches for many years and had taken an oral ergotamine tartrate preparation some years previously, but strenuously denied any current use of such preparations, including suppositories.

The patient was discharged one week after admission, at which time physical examination was normal. Subsequent enquiries by the patient’s local medical officer revealed that she had procured, without his knowledge, eighteen ergotamine tartrate (2 mg) suppositories during the two weeks prior to the onset of her symptoms. The patient subsequently admitted to using all eighteen suppositories.

Discussion

This is, to our knowledge, the first reported case of an aortic arch syndrome due to ergotism. Neurological symptoms have been reported during epidemics of ergot poisoning in Europe due to the ingestion of ergotised grain products,4 but these have consisted of psychic disturbances, convulsions, muscular spasms or dysesthetic sensations of intense heat and cold, or burning pains ("St. Anthony’s fire"), rather than focal cerebral neurological deficits. These symptoms have occurred most commonly in malnourished subjects, suggesting the possibility of a coincident vitamin defi-
FIGURE 1. Lateral and antero-posterior projection of left (A and B) and right (C and D) common carotid arteriograms. The left common carotid arteriogram reveals a 70% stenosis of the left internal carotid artery in the high cervical region and a further stenosis as the artery enters the carotid canal (arrows). The right common carotid arteriogram reveals a very long stenosis of the right internal carotid artery, extending from the high cervical region to the entrance to the carotid canal (arrows).
ERGOT-INDUCED AORTIC ARCH SYNDROME/Feneley et al
813

FIGURE 2. Left brachial arteriogram demonstrating a long, tapering stenosis and complete occlusion of the left brachial artery with extensive collateral circulation.

Richter and Banker\textsuperscript{9} and Brohult, Forsberg and Hellstrom\textsuperscript{10} each reported a case of medicinal ergot poisoning with a focal cerebral neurological deficit and an angiographically demonstrated right internal carotid artery stenosis. Senter, Lieberman and Pinto\textsuperscript{11} described a patient with bilateral internal carotid artery stenoses very similar to those found in our patient, due to ergotism. This patient also had arteriospastic involvement of both the upper and lower limbs. Bilateral papillitis of the optic nerves has been reported following the use of ergotamine tartrate suppositories.\textsuperscript{12} While selective ergot-induced arteriospasm of the upper limbs, sparing the lower limb vessels, has been rarely reported,\textsuperscript{2-7} the combination of upper limb arteriospasm and carotid arteriospasm without lower limb involvement, producing the clinical picture of an aortic arch syndrome, has not previously been described.

Although arteriospastic disease has been reported following the oral administration of medicinal ergot preparations, it much more commonly follows the excessive administration of ergot derivatives by rectal suppository for migraine headache, or by injection.\textsuperscript{4} This reflects the fact that the liver is the primary site of detoxification of ergot alkaloids,\textsuperscript{4} so that higher serum levels are attained when these compounds are absorbed directly into the systemic, rather than portal circulation and also, in the presence of impaired hepatic function. The rate and possibly extent of intestinal absorption of ergotamine are enhanced by the simultaneous administration of caffeine.\textsuperscript{13}

Gangrene of the extremities has been reported in both humans and animals following the ingestion of ergotised grain products, but is quite uncommon following medicinal ergot poisoning, perhaps due to the intermittent nature of medicinal ergot administration.\textsuperscript{4} The animal experiments of Lewis\textsuperscript{14} suggested that secondary thrombosis of the arteriospastic vessels was necessary before gangrene ensued. Intravenous infusion of phenoxybenzamine\textsuperscript{15} and sodium nitroprusside\textsuperscript{4, 16} have been found to be effective in reversing the arteriospasm produced by ergotism.

The sudden onset of an aortic arch syndrome in a 55-year-old patient was consistent with aortic dissection, but the absence of pain and normal chest X-ray made this diagnosis unlikely and aortic dissection was finally excluded angiographically. The angiographic features were unlike those of atherosclerotic disease, Takayasu’s disease, or cranial arteritis. The erythrocyte sedimentation rate of 30 mm/hour also made cranial arteritis and Takayasu’s disease unlikely diagnoses and Takayasu’s disease was further excluded by the patient’s age.

Although the arch aortogram excluded aortic dissection, the possibility of internal carotid artery dissection was considered, in view of the sudden onset of the syndrome. The angiographic appearance of the right carotid lesion was consistent with arteriospasm in our view, rather than dissection. The appearance of the left carotid lesion was also consistent with arteriospasm, though the possibility of fibromuscular dysplasia was considered; certainly, there was no evidence of dissection. It should be emphasized that the carotid pulse had returned at the time of angiography, suggesting that the angiographic appearance does not reflect the severity of stenosis present at the time of admission and further supporting the diagnosis of arteriospasm. In addition, the appearance of the brachial artery was typical of an arteriospastic lesion.

The possibility of a non-spastic carotid arterial lesion could have been completely excluded by the demonstration of normal carotid arteries at repeat angiography following recovery. However, it was felt that the complete resolution of the patient’s signs, associated with normalisation of the carotid Doppler and oculoplethysmographic studies provided adequate evidence of the arteriospastic nature of the problem and further cerebral angiography, with its finite (albeit small) risk, was felt to be unwarranted.

While it is conceivable that differential effects of ergot on the intracranial and extracranial circulation might reduce the reliability of carotid Doppler studies, the oculoplethysmographic determinations would not be so affected. Further, the final carotid Doppler studies and oculoplethysmographic determinations were
performed at least one week after the cessation of the ergot preparation.

In conclusion, the sudden onset, angiographic appearance and rapid resolution of the syndrome (following cessation of the causative agent after hospital admission) in a patient with a previous history of ergot administration for recurrent headache, were entirely typical of ergotism. Despite the patient’s denial of ergot use, there appeared to be no other satisfactory explanation of the clinical syndrome and subsequent enquiries finally confirmed this diagnosis.

We have reported this case in order to demonstrate that, while lower limb arteriospasm is more typical of ergotism, selective upper limb involvement and even carotid arteriospasm with neurological sequelae can result from the excessive administration of medicinal ergot derivatives, notably in the form of suppositories. Ergotism, albeit a rare cause, must now be included in the differential diagnosis of aortic arch syndromes.

References
Transient aortic arch syndrome with dysphasia due to ergotism.
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Stroke. 1983;14:811-814
doi: 10.1161/01.STR.14.5.811

Stroke is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0039-2499. Online ISSN: 1524-4628

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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