Steroid-responsive leucoencephalopathy

Steroid-responsive leucoencephalopathy due to cholesterol embolism

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Occlusion of small deep perforating arteries due to cholesterol embolism

The paper by Andreux *et al*¹ (see p 180) reported the first case of multiple cholesterol emboli responsible for a steroid-responsive encephalopathy.

Showers of cholesterol emboli are dislodged from disrupted, ulcerated carotid or aortic atheromatous plaques. The aortic arch is, however, the main source. In the French Study of Aortic Plaques in Stroke, after adjustment for the presence of carotid stenosis, atrial fibrillation, peripheral arterial disease and other risk factors, aortic plaques ≥ 4 mm thickness were found to be an independent risk factor for recurrent stroke.² Cholesterol embolisation can occlude retinal arteries and/or multiple small deep and leptomeningeal brain arteries 17–585 μ m in diameter. Lacunar infarcts, commonly due to lipohyalinosis, can also be caused by cholesterol emboli occluding the small deep perforating arteries.³ Diagnosis may be difficult, especially in patients presenting fluctuating, but also progressive, cerebral symptoms such as confusion and disorientation. Recurrent amaurosis fugax or transient ischaemic episodes, ischaemic stroke, epilepsy, confusion and encephalopathy are the main neurological findings often associated with inflammatory syndrome.⁴ Involvement of other peripheral organs can help in the diagnosis, notably renal infarcts,

pancreatitis, intestinal infarcts and purple toes, as well as the occurrence of acute neurological deficit after vascular surgery or arterial catheterism. Cholesterol crystals can be identified at funduscopy and skin biopsy. Carotid ultrasonography and transoesophageal echocardiography are required to detect the source of embolism. Surgery is recommended for the prevention of cholesterol emboli released from carotid atherosclerotic stenosis, but there has been no randomised controlled trial comparing antiplatelet agents, anticoagulant treatment and statins when the source is in the aortic arch.

The authors report the case of an extensive leucoencephalopathy, with multiple bilateral hypersignals on fluid-attenuated inversion recovery image-weighted magnetic resonance imaging (MRI) suggesting vasogenic oedema in the cerebral white and grey matter, brain stem, cerebellum and basal ganglia. There was no evidence of cytotoxic oedema or infarcts on diffusion-weighted MRI. Follow-up MRI showed complete resolution of lesions after high doses of steroids. Histopathological examination of the brain showed numerous scattered small cortical and subcortical infarcts and multiple cholesterol emboli occluding small leptomeningeal and perforating arteries infiltrated by reactive lymphocytes. The

physiopathological mechanism of this reversible leucoencephalopathy remains unclear. On the basis of histological examination and the dramatic response to corticosteroids, the authors' hypothesis is a localised arterial inflammation at the site of cholesterol occlusion, but endothelial vascular reaction disrupting the blood–brain barrier is another possible explanation, as focal arterial spasm around embolic occlusions and damaging the arterial wall has been reported.⁵ This case requires confirmation, but neurologists should be aware that cholesterol emboli can be responsible for acute encephalopathy likely to respond to corticosteroid treatment. A limiting factor is, however, the inability of steroids to recanalise the arterial cholesterol occlusion and prevent cerebral infarcts. The only expected potential effect, as suggested by the authors, is the reduction of vasogenic oedema underlying the clinical syndrome of encephalopathy.

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REFERENCES

- 1 Andreux *et al*. *J Neurol Neurosurg Psychiatry* 2007;**78**:180–2.
- 2 Di Tullio MR, Sacco RL, Homma S. Atherosclerotic disease of the aortic arch as a risk factor for recurrent ischemic stroke. The French Study of aortic plaques in stroke group. *N Engl J Med* 1996;**334**:1216–21.
- 3 Laloux P, Brucher JM. Lacunar infarctions due to cholesterol emboli. *Stroke* 1991;**22**:1440–4.
- 4 Ezzeddine MA, Primavera JM, Rosand J, *et al*. Clinical characteristics of pathologically proved cholesterol emboli to the brain. *Neurology* 2000;**54**:1681–3.
- 5 Brucher JM, De Smet YD, Gonsette RE. Rupture of the arterial wall in two cases of recent cerebral embolism: morphological expression of cerebral vasospasm? *J Neurol* 1986;**233**:267–0.