Comment

This patient had subacute encephalitis, the commonest neurological manifestation of AIDS,^{2,3} which is characterised initially by mild cognitive changes and psychomotor retardation. Analysis of cerebrospinal fluid may show a mild pleocytosis, a rise in protein concentration, and a lowered glucose concentration.

The prevalence of ophthalmological signs in AIDS is at least 50% and is higher in those studied at necropsy.^{4,5} The commonest findings are microaneurysms, haemorrhages in the nerve fibre layers, and focal non-perfusion of small retinal vessels, producing cotton wool spots. These abnormalities are probably due to a microvascular retinopathy. Other intraocular complications include cytomegalovirus retinopathy, Roth's spots, and ocular candidiasis. Our patient's ophthalmological signs were caused by ischaemia of the optic nerve head from affected posterior ciliary arteries. The localised areas of fluorescein staining of the retinal veins suggest that the retinal circulation was also affected.

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Disseminated cholesterol embolism presenting as neuromyelitis optica

Embolisation of cholesterol crystals from eroded aortic atherosclerotic plaques may result in various clinical manifestations affecting the kidney, the skin, the gastrointestinal tract, the heart, the retina, and the brain.¹

We report a case that suggests that neuromyelitis optica should be included in the clinical range of cholesterol embolism.

Case report

An 80 year old woman was admitted because of paraplegia and urinary incontinence of recent onset. Six months earlier she had experienced a transient loss of vision in the left eye, which had been attributed to optic neuritis. Physical examination showed dehydration, petechiae on the toes, flaccid paraplegia with bilateral Babinski's sign, and a well defined loss of sensation in response to heat at the level of T 9. Funduscopic examination yielded normal results. The results of biochemical tests showed an increased erythrocyte sedimentation rate (117 mm in the first hour) and renal insufficiency (blood urea nitrogen concentration 36.7 mmol/l, serum creatinine concentration 295 µmol/l). Antinuclear antibody was present at a titre of 1/320; antibodies to double stranded deoxyribonucleic acid and smooth muscle were absent. The test for lupus erythematosus yielded negative results, and there were no clinical or serological signs of systemic lupus erythematosus. No proteinuria was found, urine sediment was normal, and renal function improved after rehydration. Analysis of cerebrospinal fluid yielded unremarkable results, and myelography excluded medullary compression. Visual evoked potentials showed increased latency on the left side.

Neuromyelitis optica was diagnosed, and the patient received prednisolone 1 mg/kg/day without improvement of her neurological state. She died in a marasmic state four weeks later. At necropsy the thoracic and abdominal aorta showed ulcerated atheromatous plaques covered by friable material. The spinal cord showed extensive necrotic lesions at the thoracic level. Several distal branches of the anterior and posterior spinal arteries had been obliterated by needle shaped, crystalline clefts characteristic of cholesterol emboli (figure). In addition, hyaline thickening of small arterioles was noted. No specific changes were found in the brain except an area of demyelination in the optic chiasma. The eyes were not available for pathological studies. Examination of other tissues

Spinal cord at thoracic level (Luxol fast blue), showing extensive necrotic lesions of white and grey matter. Arrow indicates a cholesterol embolus in a branch of posterior spinal artery, and inset shows higher power view of cholesterol cleft in a spinal vessel.

showed disseminated cholesterol embolism with arterial reactions at different stages. Moreover, characteristic crystalline clefts were found in small and medium sized arteries of the kidneys, the liver, the stomach, the pancreas, the spleen, the adrenal glands, and the skin and in a mediastinal lymph node.

Comment

Neuromyelitis optica, or Devic's disease, is characterised by the association of optic neuritis with spinal cord lesions. This clinical entity has been described in association with viral infections, tuberculosis, and systemic lupus erythematosus but is most often idiopathic.²

Pathological and clinical data indicate that neuromyelitis optica is usually a demyelinating disease related to multiple sclerosis. Recently, attention has been drawn to peculiar vascular changes observed in some patients with the disease. These changes, reminiscent of those reported in patients with subacute necrotic myelopathy, are characterised by the proliferation of thickened, hyalinised small vessels in areas of severe demyelination.²

In our patient similar changes were found associated with cholesterol embolisation. Atheromatous emboli have previously been implicated in spinal cord infarcts,^{3,4} but, to our knowledge, this is the first description of neuromyelitis optica associated with cholesterol embolism.

Cholesterol embolism usually develops after surgery on a severely atherosclerotic aorta or after major vessel angiography, but, as in our patient, it may also occur spontaneously. The protean clinical manifestations may mimic systemic vasculitis, and laboratory findings may add to confusion as erythrocyte sedimentation rate is often increased, transient eosinophilia is common, antinuclear antibody may be present, and abnormal results of urine analysis are common.⁵

Neuromyelitis optica is viewed as a clinical syndrome with several possible aetiologies.² Our findings indicate that cholesterol embolisation should be considered to be a possible cause of the syndrome, especially in elderly patients with severe aortic atheromatosis.

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