father and sister have similar headaches. Two years ago he discovered that the pain disappeared after a few hours of strenuous cycling.

With the next attack he was far from home and he therefore tried running (in Hyde Park, incidentally), with equal and quicker success. Since then, 15 subsequent attacks were treated by running for about 20 minutes, with immediate relief on all occasions but one (95% confidence interval of cure rate 68–99%). Of the 80-odd previous bouts only one or two ended on the day of onset (95% confidence interval of spontaneous cure rate 1–9%). Concurrent control episodes would have been preferable, but the patient has been unwilling to abstain from his athletic remedy.

I hope these observations will prompt others to try this form of treatment for common migraine. It is cheap, has relatively few side effects, is only mildly addictive, and may even provide additional benefits.

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Cerebral thrombophlebitis in a patient with systemic lupus erythematosus

Sir: We report a pattern of systemic lupus erythematosus (SLE)-associated cerebral vasculitis that, to our knowledge, has not been previously described. Ellis and Verity reviewed the neuropathological findings in 57 cases during the years 1955–1977. Their series did not have a single instance of thrombosis of the large meningeal veins. Johnson and Richardson’s clinical-pathological study between the years 1945–1960 was similar. Other authors confirm the rarity of thrombosis or other lesions of the larger vessels. We describe the clinical course and neuropathological findings in such a case. A 33 year old black woman was diagnosed as having systemic lupus erythematosus based on a history of pericarditis, arthritis, a positive ANA of 1:640, anti-DNA 62 and a positive Smith antigen. She was maintained on naprosyn for 9 months until March 1986 when she presented with a 7 day history of fever, malaise, cough, nausea, vomiting and arthralgia.

Twenty-four hours following admission, tender erythematous macular lesions were noted on her fingertips. Over the next four days the patient remained febrile; blood, urine and sputum cultures were normal. Serum complement levels were low (C3 56 mg/dl, C4 9 mg/dl) and the double stranded DNA (dsDNA) was elevated at 180 U/ml. The patient was begun on prednisone, 60 mg/day for presumed lupus flare. The patient defervesced and the cutaneous lesions resolved. Forty-eight hours later, 6 days following admission, the patient experienced the abrupt onset of a generalised tonic clonic seizure. A computed tomography (CT) scan was obtained which revealed a small left temporal haemorrhagic infarct. She later had two additional tonic clonic seizures.

After the last seizure, the patient became agitated, restless, and began making purposeful movements without apparent awareness of her environment. She did not respond to visual threat. Over the next 12 hours, she developed dyconjugate gaze and bilateral papillodema. A repeat CT scan revealed bilateral temporal hemispheric haemorrhagic infarcts, oedema, and herniation. She died 10 days after admission. A necropsy was performed. The brain weighed 1500 g and was diffusely edematous with bilateral uncal and cerebellar tonsillar herniation. There were focal subarachnoid haemorrhages over the cerebrum, cerebellum, and spinal cord. The dura and dural sinuses were unremarkable. Coronal sectioning revealed multiple haemorrhagic infarcts (fig A). The largest lesions were in the left inferior frontal and left superior temporal lobes and measured about 6 × 4 × 4 cm and 4 × 4 × 3 cm respectively. There was also a 1.5 cm diameter haemorrhagic infarct in the right superior temporal gyrus, and multiple haemorrhagic infarcts in the cerebellum measuring up to 0.5 cm in diameter. The meningeal veins overlying the infarcts were grossly thrombosed.

Sections were processed for histology and stained with haematoxylin-eosin and special stains for microorganisms. There was severe vasculitis of the meningeal veins, with necrosis of the vessel walls, thrombosis, and transmural inflammatory cell infiltrate. The inflammatory cells consisted mostly of polymorphonuclear leukocytes (fig B) with rare lymphocytes and macrophages. Rarely, some of the small meningeal arteries showed mild inflammation of the vessel wall but no evidence of necrosis or thrombosis. There was occasional inflammation, fibrinoid degeneration and thrombosis of small intraparenchymal arterioles, venules, and capillaries. Microinfarcts and small haemorrhages were not seen. There were signs of diffuse cerebral anoxia. Special stains for organisms were negative. All major branches of the arterial system were sampled and were not involved by vasculitis.

This was an interesting and unusual case of cerebral lupus vasculitis involving primarily the large meningeal veins. The catastrophic neurological events occurred despite prednisone therapy. Thrombophlebitis resulted in multiple haemorrhagic infarct, herniation and death.

Central nervous system (CNS) involvement occurs in 50% of patients with SLE. One of the earliest reports of the neuropathological features of central nervous system SLE was that by Berry and Hodges. The brains of five patients were examined: two had insignificant vascular changes, the third showed proliferation of the intimal and elastic layers of the leptomeningeal vessels, the fourth had vascular proliferation with areas of acute fibrinous reaction, and the fifth brain had old and recentencephalomalacia, proliferative vascular lesions, and thrombotic occlusion of the leptomeningeal and parenchymal vessels.

In 1968, Johnson and Richardson reviewed 24 cases of CNS-SLE, 10 of which had significant gross abnormalities. Only three had large intracerebral haemorrhages. None showed cerebellar infarct or haemorrhage. They believed that the central nervous system signs were inflammatory in origin, pointing out that while "perivascular infiltrates" were often noted, true arteritis was rare. Only once did they see arteritis of a branch of the middle cerebral artery. In their experience, even “true vasculitis” with inflammatory cells within a vessel wall was rare (3 of 24 cases), and in no instance was it a prominent or generalised phenomenon. In another study, vasculitis was apparent in only one of six instances, and the inflammatory infiltrate was almost entirely perivascular and localised to the subependymal areas. Although ischaemic infarcts were seen in nearly one-half of the cases reported by Ellis and Verity (80% microinfarcts, 20% large infarcts), associated arterial thrombi were rarely seen. Larger infarcts were found usually in the distribution of the middle cerebral artery. Forty-two percent showed prominent haemorrhage of some form, demonstrable vasculitis being highly correlated with subarachnoid haemorrhage. We found...
Fibrinoid degeneration 

The involvement.

Thrombophlebitis rather than arterial disease, and no microinfarcts with haemorrhage. This reverse pattern of involvement of the small and large vessels is distinctly unusual.

Thrombosis or other lesions of larger vessels are distinctly uncommon at necropsy. However, in the recent literature, Kelley et al reported a case of medium-sized cerebral transmural arteritis, and Scharre et al described a case of arteritis with giant cells involving the middle and posterior cerebral arteries, both in SLE patients at necropsy.

In summary, our patient had an SLE-associated cerebral vasculitis which differed from previous cases in having more inflammation and necrosis of the larger veins than the smaller vessels. Such extensive involvement of the larger meningeal veins and involvement of the cerebellum has not, to our knowledge, been previously documented.

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Fig (A) Coronal section showing haemorrhagic infarcts in left frontal and temporal and right temporal lobes (arrows). The softened area in the centre is due to autolysis. (B) Transmural inflammation, in meningeal vein (upper part of field), consisting mostly of polymorphonuclear leucocytes. The vein is filled by thrombus (lower part of field). Haematoxylin and eosin, × 800.
Letters


Accepted 22 May 1987

Cerebral angiopathy and recurrent strokes following Borrelia burgdorferi infection

Sir: Neurological abnormalities are seen in 11 to 15% of Borrelia burgdorferi infection. We report a woman who developed an angiopathy with recurrent ischaemic events during 3 years following a tick bite, with Borrelia burgdorferi infection.

During spring 1983, this 40 year old woman noticed a tick bite under her left breast, followed by red lesions on the chest and limbs. One year later, she experienced headache associated with weakness and paraesthesia around the mouth and in the left hand. She also complained of vertigo and intermittent dysarthria. She received flunarizine (10 mg per day for 3 months) and the symptoms disappeared. In November 1985 she developed a left ataxic hemiparesis, and CT showed a hypodense area in the right lenticular nucleus suggesting infarction. As blood pressure was 170/120 mm Hg, amiloride and hydrochlorothiazide were introduced. The patient recovered within 4 weeks. Four months later, she complained of blurred vision with numbness in the hands. There was a slight left-sided hemiparesis. Treatment with atenolol (50 mg per day) was begun and the symptoms did not recur. In August 1986, she developed diplopia, confusion and a stumbling gait. The patient showed a skew deviation with the left eye down, superimposed microsaccadic movements on pursuit attempts to the left, and an abduction palsy in the left eye. There was a limitation of vertical gaze, particularly upward. The optokinetic nystagmus quick phase was decreased to the right. There was a slight left-side hemiparesis with increased tendon reflexes, without Babinski's sign. A mild ataxia was present in the left arm with dysdiadochokinesia. Sensory testing was normal. The patient was disoriented in time and place, and periods of irritability alternated with drowsiness. Comprehension and spontaneous speech were impaired. Writing and reading were also affected. There were no apraxia. No meningeal signs were present and the remainder of the physical examination was normal. On CT scan, there was a bilateral hypodense area in the thalamus suggesting a paramedian infarct, and the old lesion in the right lenticular nucleus was still visible, without contrast enhancement (fig 1). The cerebrospinal fluid (CSF) contained 275 cells/mm$^3$ with 82% lymphocytes and 9% plasmocytes. Protein 267 mg/dl (peripheral blood glucose = 3.5 mmol/l). Oligoclonal bands were determined by agarose gel electrophoresis using concentrated CSF (Beckman Immunochimistry Systems). Titres of specific antibodies against Borrelia burgdorferi were determined in the CSF: IgM = 1/4, IgG = 1/128, and in the blood: IgM = 1/32, IgG = 1/256 (positive test: >1/32). Gram stain, routine culture and culture for acid fast bacilli and fungi were negative. Reagin tests to Treponema pallidum were negative in the CSF and blood. The right carotid arteriogram demonstrated segmental narrowing and obstruction of branches of middle and anterior cerebral arteries (fig 2). More distal branches of these arteries received collateral flow in a retrograde fashion via leptomeningeal anastomoses. The same lesions were present in the left side. The thalamic arteries were extremely narrowed bilaterally. The basilar artery and the right superior cerebellar artery (SCA) were small, and there were anastomoses between the SCA and the posterior inferior cerebellar artery, and between the anterior inferior cerebellar artery and leptomeningeal arteries. Laboratory data showed a haemoglobin of 143 g/l and a white blood count of 9800 cell/mm$^3$ with a normal differentiation. The ESR was 6 mm/h. The following tests were negative or normal: serum sodium, potassium, urea, glucose, SGOT, SGPT, alkaline phosphatase, gamma GT, serum protein and electrophoreses, IgA, IgG, IgM, Ig-K, Ig-λ, C3, C4, cryoglobulins, antinuclear antibody and anti-DNA, rheumatoid factors and Kweiss test. Immune complexes were 7,1 (n < 5-6). The blood pressure was normal. A chest radiograph and an electrocardiogram showed no abnormality. A muscle biopsy (M vastus medialis) showed no abnormal fibres or vessels, but direct immunofluorescent studies of a skin biopsy specimen (thigh) revealed antibodies (IgG) in the basement membrane zone at the dermo-epidermal junction. Penicillin G (24 million units per day for 10 days) and prednisone (60 mg per day with tapering over 8 weeks) were begun. The neurological state improved within 3 weeks. The CSF showed 82 cells/mm$^3$ with 92.5% lymphocytes and 1.5% plasmocytes. Protein 123 mg/dl (peripheral blood glucose = 3.4 mmol/l). Intrathecal synthesis of IgG and oligoclonal bands at electrophoresis. Clinical examination 3 months after the end of the treatment showed mild memory disorders and there was a relative palsy of the upward saccades. Specific antibodies against Borrelia burgdorferi were in the CSF: IgG = 1/8, and in the blood: IgG = 1/64. IgM were negative in the CSF and in the blood.

The relation between cerebral infarction and vasculitis due to infectious diseases prompted us to discuss the possible relation between a Borrelia burgdorferi infection and the development of ischaemic events associated with a diffuse cerebral angiopathy in our patient. During 3 years following the tick bite, repeated cerebrovascular accidents involving several vascular territories were observed. The CSF was typical with a lymphocytic pleocytosis, elevated protein, oligoclonal bands and IgG index. Specific antibodies against Borrelia burgdorferi were also present, compatible with an old infection.1 2 The cerebral angiopathy demonstrated on angiography has not been reported previously in Borrelia burgdorferi infections, probably because angiography is not performed currently in this condition, even when relapsing CNS dysfunction is present.3 In our case, the angiographic findings were similar to those observed in meningovascular syphilis or in several non-infectious arteritis.4 5 In our patient, there were neither findings suggesting arterial involvement outside the central nervous sys-

Fig 1 CT shows a hypodense area in the left thalamus and the old lesion in the right lenticular nucleus.
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