A diagnosis of cluster headache was made, the attacks being clearly different from the classic migraine of earlier years which had ceased at the age of 20. He was advised to eliminate alcohol during clusters. Half of an ergotamine tartrate suppository (1 mg) was given 8 to 12 hourly in an attempt to anticipate attacks. Symptoms disappeared quickly and the bout had ended within 2 to 3 weeks in 1978.

He next reported in 1986, having had occasional mild clusters in the intervening 8 years, which he could readily control with ergotamine used for 2 or 3 weeks. However, for 2 weeks the pattern of his headaches had changed.

During this period he had experienced no less than 10 to 14 attacks of similar headache each day (see fig). Seven or eight were at night "waking me like clockwork at hourly intervals" and three or four occurred in waking hours. Attacks were much briefer, many lasted 5 to 10 minutes; the longest persisted for 40 minutes. He described a stabbing and throbbing pain in the left forehead and eye, radiating to the left temple, side of nose, upper jaw and, when severe to the ear and occiput. Associated with this intense pain were localised tenderness and a watering bloodshot eye, congested stuffy nostril and profuse sweating over the same side of the forehead and cheek. He had eliminated alcohol and taken meptazinol, paracetamol and aspirin preparations without relief. Movements of the neck and local pressure had not been noticed to trigger attacks. He had no visual disturbance, and was nauseated in only the most severe episodes, but he had never vomited.

A diagnosis of chronic paroxysmal hemicrania was suspected; however, he was started on 1 mg ergotamine tartrate suppositories 8 hourly as a base line with which to compare the future use of indomethacin. Ergotamine afforded but partial relief for 4 days; his headaches then returned with a vengeance (fig). After 2 weeks he was given indomethacin 25mg, tds, and symptoms rapidly declined in intensity and frequency. The dose was increased to 25mg, qds, and symptoms vanished after 22 days. He has been in total remission, off drugs for 7 months.

His illness has thus shown three phases: classic migraine age 16 to 20 yr, episodic cluster headache age 48 to 57 yr, chronic paroxysmal hemicrania age 58 yr (a single attack so far).

The content of chronic paroxysmal hemicrania attacks is very similar to cluster headache. There is intense, short lived strictly unilateral pain centred on the orbit and temple, associated with a red watering eye and blocked or running nostril. In both syn-

dromes the victim is restless and distraught and will pace the floor or leave the house, in vivid contrast to the enforced immobility which characterises migraine.

The characteristics of chronic paroxysmal hemicrania are: (a) greater frequency of attacks, usually more than 8 per 24 h; (b) briefer duration than cluster attacks 5 to 30 min as compared to 30 to 120 min; (c) a much higher incidence in women; (d) a complete therapeutic response to indomethacin.

Unusual features in our patient are: he is male; the onset of chronic paroxysmal hemicrania age 58 yr is late (mean was 29 in Sjaastad's series;⁷ headaches responded to indomethacin but, did not recur when the drug was stopped. In the light of this case and one other unreported patient a 17 year old girl with an episodic course (JMSP), we suggest a new classification, as in cluster headache there may be two types:

Chronic paroxysmal hemicrania

1 episodic—uncommon, perhaps 5% of cases

2 chronic—common, perhaps 95%.

Cluster headaches

1 episodic, common (90%)

2 chronic, uncommon (10%)

Finally, it is of interest that one patient has had three distinctive and apparently separate headache syndromes at three different periods of his life. The aetiology of both chronic paroxysmal hemicrania and cluster headache remain obscure. There is evidence (reviewed in refs 10, 11) in cluster headache of a peripheral paresis of the sympathetic nervous system, probably located at the third neuron in the pericarotid plexus. This explains the miosis, ptosis, conjunctival injection and nasal stuffiness. Lachrymation and rhinorrhoea, however, raise the question of parasympathetic (cholinergic) hyperactivity, but evidence for this is insubstantial.¹⁰ Similar mechanisms are likely to obtain in chronic paroxysmal hemicrania, but the timing, frequency and therapeutic responses are distinctive.^{11,12} The occurrence of the three disorders at separate times in a 40 year period in one patient strengthens the case for a common susceptibility to "neurovascular headaches"¹³ and perhaps for a central that is, hypothalamic disorder of threshold which dictates the pattern and timing of attacks and the remissions.

SHS PEARCE*

JGC COX

JMS PEARCE

Department of Neurology,

Hull Royal Infirmary,

Hull HU3 2JZ

and The Medical School,*

University of Newcastle upon Tyne,

UK

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Relief of common migraine by exercise

Sir: It is generally believed that exercise aggravates vascular headache of any kind.^{1,2} I wish to report a case in which the effect was quite the contrary.

A 44 year old male physician had been suffering for 6 years from attacks of a dull boring headache in the left temple. The bouts occurred between one and four times a month, usually on waking up in week ends, and lasted until the next morning occasionally for 2 or 3 days. The attacks were often triggered by lack of sleep, a few glasses of red wine, or both. Visual or other prodromes never occurred, and apart from faint nausea there were no associated features. Pressing the superficial temporal artery gave some sort of relief but did not end the attacks; aspirin was ineffective. His

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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.

J VAN GIJN

University Department of Neurology,
Nicolaas Beetsstraat 24,
3511 HG Utrecht,
The Netherlands,

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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 Verity¹ 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 clinicopathological study² between the years 1945–1960 was similar. Other authors confirm the rarity of thrombosis or other lesions of the larger vessels.³ 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 (C_3 56 mg/dl, C_4 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 dysconjugate gaze and bilateral papilloedema. 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 $6 \times 4 \times 4$ cm and $4 \times 4 \times 3$ 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.⁴ One of the earliest reports of the neuropathological features of central nervous system SLE was that by Berry and Hodges.⁵ 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 recent encephalomalacia, 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,³ 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),¹ 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