Letters

Muscle hypoxia in polymyositis

Sir: The pathogenesis of polymyositis is unknown, but there is circumstantial evidence that pathological changes in the intramuscular blood vessels may play a role. Ultrastructural abnormalities have been found in the endothelium of the microvessels, and capillary basement laminae have been reported to be thickened. Capillaries are lost in polymyositis, particularly in the dermatomyositis subtype.[1]

These changes, possibly triggered by immune complex deposits in the vessel walls,[2] may be responsible for the reported decline in muscle blood flow in polymyositis,[3] but it is not known whether they are able to render the muscle ischaemic enough to contribute to the necrotising damage in the muscle tissue.

We measured tissue gas tensions in five adults (four women, one man, age 42–67 years) with 2–7 years history of definite polymyositis as diagnosed by clinical, neurophysiological and muscle biopsy examinations. One of them had skin symptoms suggesting dermatomyositis. All of these patients were ambulatory, and were at a chronic stage of disease with only minimal clinical symptoms (one further patient—with a low resting PO_2 is not included in the results, as her exercise test had to be interrupted because of fatigue). Five healthy volunteers (two women, three men, age 42–68 years) served as controls. The measurements were carried out by means of an implanted Silastic tonometer.[4,5] Under local anaesthesia, a 16 cm long silicone elastomer tube with an external diameter of 1·4 mm and an internal diameter of 1·0 mm was inserted into the medial belly of the gastrocnemius muscle. Care was taken to insert the tonometer at the same depth in the patients and controls. The recordings were made four days after implantation. The tube was slowly perfused with hypoxic saline, and the PO_2 and PCO_2 of the efflux were continuously monitored both at rest and during a heel lifting exercise.[5]

As shown in the figure, the calf muscle PO_2 levels were consistently lower in the polymyositis patients than in the controls. The differences are statistically significant both at rest and during exercise ($p = 0{\cdot}004$, Mann-Whitney U test). The lowest values were found in the dermatomyositis patient. PCO_2, on the other hand, behaved similarly in the two groups. The response of PO_2 to exercise in the polymyositis patients was different from that in patients with occlusive arterial disease: in the latter condition the PO_2 is close to normal at rest but decreases during exercise.[5] It seems, therefore, that in polymyositis the larger vessels are patent and they are able to respond to the needs of autoregulation. The finding of low average PO_2 in polymyositis patients both at rest and during exercise suggests that the capillaries are unable to maintain normal tissue oxygenation. Thus, the behaviour of PO_2 in polymyositis is consistent with the hypothesis that circulation is hampered at the level of small peripheral vessels.

It may be asked whether the disability and inactivity of the polymyositis patients causes diminished blood flow and drop of temperature in the muscle tissue with resultant unspecific changes in tissue gas tensions. The patients, however, were ambulatory and had only minimal weakness at the time of the study. Furthermore, low tissue temperature would enhance the solubility of O_2 in the extracellular fluid, and higher (not lower) PO_2 would be found in measurements carried out at the standard temperature of 37°C.

The finding of marked relative muscle hypoxia in idiopathic inflammatory myopathy suggests that microvascular obstruction plays a role in its pathogenesis. The result may also have therapeutic implications.

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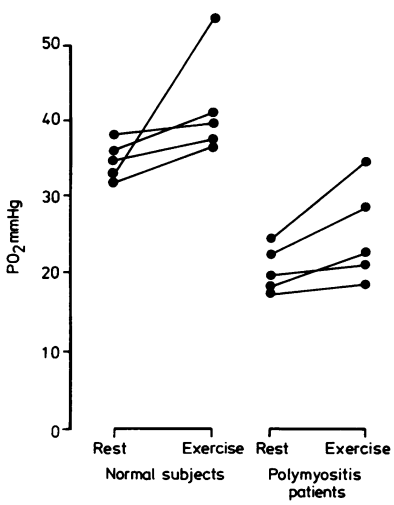

Fig *Calf muscle PO_2 in polymyositis patients and normal subjects during rest and heel lifting exercise.*

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Accepted 1 April 1986

Dilated tonic pupils in neurosyphilis

Sir: In patients with central nervous system syphilis, pupillary abnormalities are a relatively common finding, and the most important early ocular sign.[1] The following case is presented as an example of a previously noted[2] but not widely recognised association between bilaterally dilated tonic pupils, fixed to light and accommodation, and neurosyphilis.

A 48-year-old male presented with a 4 to 5 year history of mild photophobia. There was no history of diabetes, hypertension, trauma, headaches, fever, chills, nausea, vomiting, seizures, weakness, pain, or use of medications or illicit drugs. He claimed no alcohol use for at least four months but admitted to previous heavy ethanol abuse. He stated that he had occasional diplopia and intermittent urinary incontinence. At age 26 years, the patient presented with a penile chancre and positive serum VDRL, and was treated with a series of intra-

muscular penicillin injections for primary syphilis. He denied any homosexual activity. There was no family history of neurological disease.

Physical examination revealed a temperature of 36·8°C, blood pressure 132/76 mm Hg, pulse 84 per minute, and respirations 20 per minute. General examination was unremarkable, and the neck was supple. He was alert and fully oriented. Cognitive function was normal. Cranial nerve examination showed no visual field cuts, visual acuity 20/40 OU without correction, normal fundi, full extraocular movements, no nystagmus or facial weakness, and a normal gag reflex. The pupils, however, were approximately 7·0 mm in diameter bilaterally, clinically unreactive to light or accommodation, with no constriction even with prolonged near effort (fig 1). With the ophthalmic instillation of a 0·1% pilocarpine the pupils constricted to 4 mm (fig 2); with 1·0% pilocarpine instillation the pupils constricted to 3 mm. Slit lamp examination was normal except for a segmental pupillary response. The patient exhibited good strength in all extremities, with normal tone. Deep tendon reflexes were 2 to 3+ diffusely except for trace ankle jerks; plantar responses were flexor. There was a moderate stocking-glove distribution sensory loss to pin prick. Vibration and position sense were normal. An upper extremity postural tremor was present (by history since age 10 years), with slight bilateral finger-to-nose dysmetria. The gait was mildly wide-based and stiff.

Routine laboratory studies were normal. The serum VDRL was positive in a 1:64 titre and the serum FTA-ABS was 4+. Skull radiographs, electroencephalogram, and head CT scan with and without contrast showed no abnormalities. A lumbar puncture revealed clear, colourless fluid with 44 WBC/mm^3 (86% lymphocytes, 9% polymorphonuclear cells, 5% monocytes), 6 RBC/mm^3, protein 1·44 g/l, glucose 0·031 g/l, lactate 0·042 g/l, and a positive CSF VDRL in a 1:8 titre. CSF bacterial cultures and AFB smear were negative, and cytology showed inflammatory cells in excessive numbers with no malignant cells.

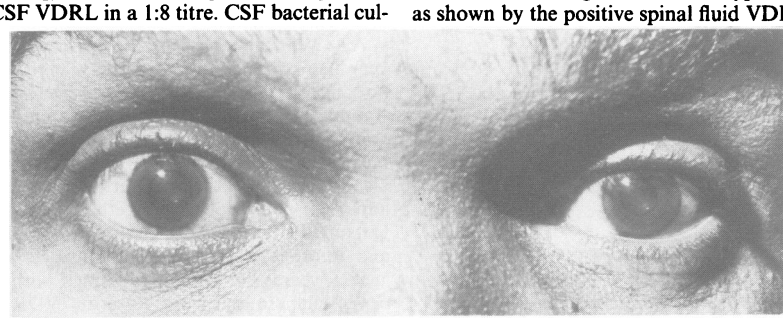

Fig 1 *Before instillation of pilocarpine.*

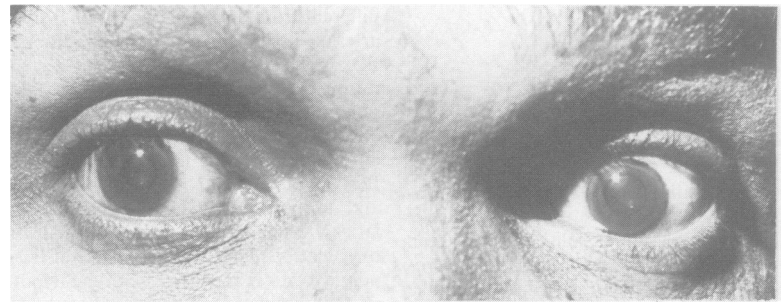

Fig 2 *Thirty minutes after instillation of 0·1% pilocarpine to right eye.*

The patient received 400 000 units of intravenous penicillin every six hours for 14 days and had an uneventful hospital course, with no changes in his clinical condition. A repeat spinal tap after completion of penicillin therapy showed a total of 4 WBC/mm^3 (90% lymphs, 10% polys), 5 RBC/mm^3, protein 1·22 g/l, glucose 0·045 g/l%, and lactate 0·16 g/l; cytology revealed only singular benign lymphocytes, with no malignant cells. Seven months later he was rehospitalised with mild dementia, right hemiparesis, and bilateral spasticity. There was no change in the pupillary signs and response to 0·1% pilocarpine. CSF examination showed clear colourless fluid, no cells, protein 0·051 g/l, glucose 0·056 g/l, VDRL negative. A left parietal lucency appeared on CT scan exhibiting slight enhancement with contrast at the periphery. Cerebral angiography revealed occlusion of both anterior cerebral arteries, constriction of proximal segments of the middle cerebral arteries and reconstitution of anterior cerebral artery circulation via collaterals. Subsequently, the patient's strength and mental status improved and he was discharged.

This patient easily fulfilled Hooshmand's criteria for the diagnosis of neurosyphilis,[3] as shown by the positive spinal fluid VDRL and CSF lymphocytic pleocytosis which resolved with penicillin alone. The percentage of neurosyphilis patients having pupillary abnormalities ranges in recently reported studies from 45 to 53%,[4 5] although during the course of tabes dorsalis in particular the percentage may be 90% or higher.[6 7] While the most widely known pupillary abnormality associated with neurosyphilis is the Argyll Robertson pupil,[8 9] various other pupillary findings are actually more common,[10] and larger dissociated pupils outnumber the miotic ones.[11]

In one group of 108 syphilitic patients, 33 were found to have absent pupillary light reactions, and 19 had no accommodation response,[12] but no mention was made of pupillary size. In children with juvenile paresis, and others with congenital neurosyphilis, the pupils are said to frequently be wide and fixed to both light and accommodation.[1 11 13] Dilated pupils, nonreactive to light and accommodation have occasionally been reported in patients with neurosyphilis.[1 2 14] One study, referred to in Loewenfeld's review article,[15] indicated that 3·3% of their neurosyphilis patients had pupils larger than 5 mm that were fixed to light, but reactive to accommodation. Adie's tonic pupil, which is large, nonreactive to light but slowly reactive to accommodation, usually unilateral, often associated with diminished deep tendon reflexes, and suggested by pupillary cholinergic hypersensitivity to dilute solutions of pilocarpine,[2 16–18] had been noted infrequently with neurosyphilis,[5 11] occurring in less than 5% of patients.[19] In one group of 21 patients with bilateral tonic pupils, some of whom had peripheral neuropathies, seven had positive serum VDRL and FTA, but CSF apparently was not examined.[18] A recent series of 29 patients with tonic pupils and light-near dissociation tested for syphilis revealed five with positive serology, of whom four had positive CSF serology.[20]

Although our patient did not have any response to accommodation, the pupillary denervation hypersensitivity demonstrated would appear to indicate that he did have tonic pupils.[13 17] This case further illustrates that patients treated with what had previously been thought to be adequate antibiotic therapy for primary or secondary syphilis may still develop chronic progressive CNS syphilis[14] (although in this and other reported cases of neurosyphilis a re-infection cannot be absolutely excluded). Patients with dilated tonic pupils (rather than only Argyll Robertson pupils) should have serological testing for syphilis and lumbar puncture when appropriate. This would help insure that a CNS syphilis infection that may effectively be treated does not remain undiagnosed.

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Accepted 24 February 1986

Parkinsonism due to corpus callosum astrocytoma: case report

Sir: Brain tumour is an uncommon cause of the Parkinson syndrome. In the most recent review by Polyzoidis *et al* in 1985,[1] 49 verified cases with supratentorial tumours were collected from the literature. The majority of the cases were of supratentorial extra-axial benign mass lesions. We report the case of a patient with Parkinsonism due to a corpus callosum astrocytoma.

The presenting illness of this 79 year old man began in November 1983 with difficulty in gait associated with progressive weakness of the entire right side. Tremor of the left hand appeared 2 months later.

At the time of examination on 8 February, 1984, he was unable to stand alone and he had urine incontinence. The findings were as follow: blood pressure was 150/70 mm Hg, pulse 88 beats/min and temperature 36·0°C with normal general examination. On neurological examination he showed no insight into the nature of his problems. His attention span was brief and he presented dressing apraxia. No papilloedema was present. A resting tremor (4–6 Hz) was found in the left arm. This was associated with marked cog-wheel rigidity. Tremor was present but less intense in the left leg. Voluntary movements were slow and his face was mask-like. Sucking and palmomental reflex were detected. He also had retropulsion. There was a mild right-sided hemiparesis with hyperreflexia and positive Babinski reflex. There were no sensory deficits or cerebellar signs. Routine tests results were normal. The computed tomography brain scan showed a large medially situated tumour (figure). This extended into both parietal lobes through the posterior portion of the corpus callosum. Stereotaxic biopsy revealed that the tumour was an astrocytoma.

Extrapyramidal symptoms characterised by rigidity, bradykinesia and tremor, have

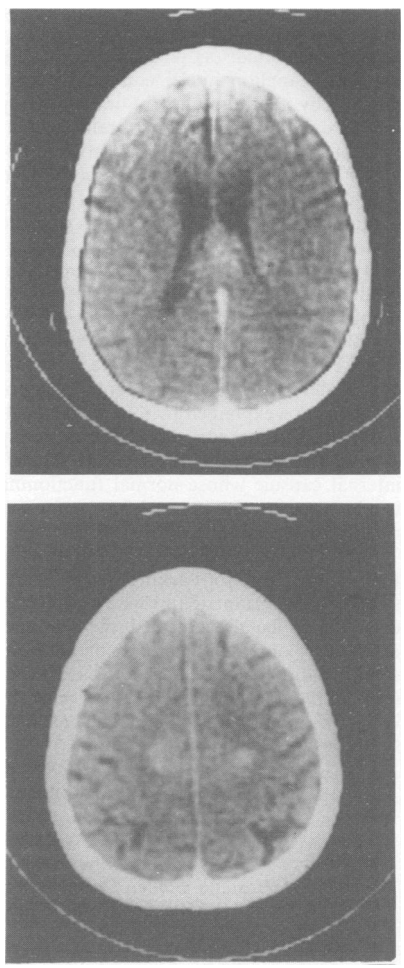

Fig *Contrast-enchanced computed tomography scan demonstrating a tumour in the posterior corpus callosum and extending bilaterally into the central white matter of both parietal lobes.*