Letters

of neuropsychiatric deterioration demonstrate focal or confluent plaques in the deep central white matter and the periventricular zones.⁷⁻¹¹ There is diffuse, spongy demyelination with almost complete sparing of the axons.

In 1965, Geschwind discussed the behaviour seen in rhesus monkeys with KBS in relation to visual-limbic disconnections resulting from fibre tract lesions in the temporal lobe.¹² Because temporal lobectomy demonstrated agnosias in modalities other than vision,⁴ a visual-limbic disconnection would not be expected to account for the entire Kluver-Bucy syndrome. However, experimental studies have shown that features of KBS can be produced in monkeys by removing the visual influences of the striate cortex from the temporal lobe.¹³⁻¹⁴ This was accomplished by destroying the temporal lobe on one side, and then destroying the contralateral visual cortex and the crossing visual fibres in the splenium of the corpus callosum.¹³ Features of KBS have also been produced both by interrupting the white matter tracts connecting the occipital and temporal lobes, and by severing the fibre tracts entering and exiting the temporal lobes medially.¹⁴

We have described the case of a 32 year old man who developed KBS as a delayed post-anoxic syndrome following carbon monoxide poisoning. That the onset of this syndrome occurred suddenly several hours following an ECT treatment suggests that ECT may have exacerbated or precipitated the appearance of delayed neuropsychiatric deterioration. The transient nature of the KBS seen in our patient is in keeping with

other descriptions of delayed post-anoxic leucoencephalopathy and suggests that the proposed disconnections are reversible.

Geschwind's hypothesis that disconnections resulting from fibre tract lesions may be associated with KBS in monkeys is supported by the experimental investigations discussed above.¹³⁻¹⁴ The neuropathological substrate of delayed neuropsychiatric deterioration following carbon monoxide poisoning is believed to be selective white matter injury.⁷⁻¹¹ This case of KBS as a delayed clinical syndrome following carbon monoxide poisoning suggests that disconnections resulting from white matter lesions may also underlie the human KBS.

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THOMAS A SANDSON

RALPH B LILLY*

MICHAEL SODKOL

From the Division of Neurology* Butler Hospital, Providence, RI, USA

*Address for correspondence: Dr TA Sandson, MD, 372 Longwood Avenue, Apt 41, Boston MA02215, USA

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Matters arising

Local autonomic failure affecting a limb

Sir: I refer to the article *Local autonomic failure affecting a limb*, by R H Johnson and B J Robinson.¹

The authors reported three cases where sudomotor and vasomotor dysfunction occurred together in one limb. They state that, while segmental anhidrosis has been reported in association with Holmes-Adie syndrome (HAS), their patients showed no features of that disorder. They did not mention an important difference between their cases and cases reported in association with HAS: viz that while their patients showed evidence of a preganglionic lesion (preservation of sweating around intradermal injection of acetyl choline), cases reported in association with HAS in the main have showed evidence of ganglionic or postganglionic lesions (lack of sweating in response to subcutaneous methacholine or pilocarpine).²⁻⁵

This notwithstanding, it seems premature to dismiss a connection with HAS completely. There has been one report of a case where a patient with normal pupils presented with segmental anhidrosis, but 6 years later developed a typical Adie pupil.⁶ Furthermore Johnson & Robinson do not mention the results of pupillometry or the results of instillation of methacholine in their cases.

The observation that one of Johnson & Robinson's patients had some ill-defined loss of sensation to pinprick in the area of sudomotor loss is of further interest in this connection. I have recently seen a case of HAS (confirmed by pupillometry and sensitivity to 2.5% methacholine) in whom, although there were no features of autonomic dysfunction, there was an area of sensory loss to pinprick on the medial border of the right forearm, from wrist to elbow. Extensive investigation, including electrophysiological studies, myelography and spinal MRI scan showed no lesion to account for this.

The possibility of a relationship between the cases of Johnson & Robinson and HAS (or the syndrome of HAS-plus, so to speak) remains to be discounted.

RODERICK DUNCAN
Division of Neurology,
Institute of Neurological Sciences,
Southern General Hospital,
Glasgow G51 4TF, UK

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Johnson replies:

The point of our article *Local Autonomic Failure in a Limb* was to draw attention to isolated local autonomic failure causing sweating loss and change of temperature in the affected limb. In company with the authors of two other reports of similar patients,^{1,2} we were unable to give an explanation for the disorder other than that it appeared to be due to a discrete lesion in the spinal cord without any clear evidence that this was related to syringomyelia. In that we did not provide an explanation for the problem, the possibility of subsequent development of a Holmes-Adie pupil, as suggested by Dr Duncan, cannot be discounted.

However, Holmes-Adie syndrome consists of a tonic pupil in association with absent stretch reflexes but this association is not absolute. This is discussed in a review of the clinical features which also considers the occurrence of the autonomic abnormalities which are occasionally found.³ On clinical examination our patients neither had absent stretch reflexes nor a tonic pupil and little can be made of a possible autonomic similarity with the Holmes-Adie syndrome, for although autonomic disorders seem to occur more commonly than would be expected by chance, the lesions may not only be post-ganglionic but in some patients afferent rather than efferent.⁴ Our patients therefore have no similarity with the Holmes-Adie syndrome at present in their clinical findings related to pupils, reflexes or to autonomic dysfunction.

It must further be questioned whether development in the future of only one feature of the Syndrome, that of tonic pupils, would really assist in understanding the disorder we have described. The Holmes-Adie

syndrome is purely a clinical description of associations rather than an aetiological explanation.

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Local autonomic failure affecting one limb

Sir: The cases of autonomic failure affecting one limb described by Drs Johnson and Robinson¹ are of particular interest because all the cases affected by failure of sweating in the left arm. In the third case there was associated normal vasomotor function in the affected limb implying single modality autonomic failure.

Recently a 54 year old woman was referred with a mixed history of hyperhidrosis of the right arm and face and pain in both arms but worse on the left side. These symptoms had started 4 years previously after a "whiplash" injury in a road traffic accident. The hyperhidrosis was aggravated by stress and was such that she misted her right spectacle lens. On examination there was in fact anhidrosis of the left arm and side of face. There was no Horner's syndrome on either side. Thermographic examination using liquid crystal contact thermography showed vasomotor failure in the right arm and normal vasomotor response to the proximal application of ice on the left side. There were no other abnormalities. In this case there was single function (sudomotor) failure again on the left side. This must raise the query why is the left arm involved in this loss in four cases? In addition single function (vasomotor) failure was present on the contralateral side to produce a picture of bilateral but discrete and different autonomic failure.

P A J HARDY

Pain Relief Foundation,

Rice Lane, Liverpool L9 1AE, UK

Reference

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Schistosoma in the spinal cord

Sir: We are at present undertaking a longitudinal study examining the clinical, serological and radiological findings in schistosomiasis of the nervous system and therefore read, with interest, the letter by Kerr *et al.*¹ Over a period of 19 months we collected 14 patients with cord and/or root involvement. Two of these cases had already been published² while details of the others will be submitted for publication shortly. Of these, six had expansion of the conus and irregularity and matting of roots. One further patient showed root involvement alone. Two of these seven patients were subjected to laminectomy but the rest were treated on the basis of clinical findings, CSF changes and systemic evidence of schistosomiasis infestation. These patients showed remarkable clinical improvement. Serial CT myelograms showed reduction in the size of the conus.

We therefore support the suggestion that patients with the appropriate clinical and investigative profile be given a therapeutic trial of praziquantel before being considered for laminectomy.

HC HARSHBARGER

AI BHECHTOLD

PLA

JE COSNET

Neurology Unit,

Department of Medicine,

University of Natal,

Wentworth Hospital,

P/Bag JACOBS 4008

Durban, Natal,

South Africa

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Aggravation of Parkinson's disease by cinnarizine

Sir: Marti Masso *et al.*¹ described an exacerbation of Parkinsonian symptoms after cinnarizine intake. Recently we reported movement disorders including Parkinsonism, induced by cinnarizine and flunarizine.² Both have similar chemical structures and pharmacological profiles.