Matters arising

Histocompatibility antigens and postencephalitic Parkinsonism

Sir: We have read with interest the article on histocompatibility antigens and postencephalitic Parkinsonism by Lees *et al*¹ recently published in your journal. The authors compared the phenotypic frequencies of selected histocompatibility antigens in 21 patients with postencephalitic Parkinsonism and controls, and did not find any differences between the groups. Missing, however, were the ethnic composition of both the patient and control populations, and the antigenic frequency data with resultant p values corrected statistically for total number of antigens tested in each HLA locus.

We had previously reported a highly significantly increased frequency of HLA-B14 antigen (corrected $p = 0.001$) in 18 unrelated American-Jewish patients of Eastern European extraction who had classical postencephalitic (von Economo's) Parkinson's disease, compared with 147 ethnically-matched controls;² the 17 patients with idiopathic Parkinson's disease concurrently studied did not show any evident differences in HLA frequencies compared to the same matched controls. In our paper, we emphasised that we were dealing with a specific, relatively homogeneous, ethnically-defined and matched study population, and that we were well aware of the known wide variations in prevalence of specific HLA antigens in various ethnic groups. The disease association we reported, while not necessarily reflecting genetic susceptibility to the disease, certainly suggests the possibility that a genetic factor may play a role in the pathogenesis of this particular variant of Parkinsonism in certain ethnic groups. The negative study of Lees *et al*, while of interest, is not comparable to ours, nor does it negate our contentions.

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References

- 1 Lees AJ, Stern GM, Compston D. Histocompatibility antigens and postencephalitic Parkinsonism. *J Neurol Neurosurg Psychiatry* 1982;45:1060-1.
- 2 Elizan TS, Terasaki PI, Yahr MD. HLA-B14

antigen and postencephalitic Parkinson's disease. Their association in an American-Jewish ethnic group. *Arch Neurol* 1980;37:542-4.

Lees *et al* reply:

Dr Elizan and Professor Yahr are correct in drawing attention to the lack of precision in detailing the ethnic composition of our patients and control populations in our report. All the patients with postencephalitic Parkinsonism were of North European Caucasian stock, had been born in the United Kingdom and their families have been settled here for at least two generations and matched controls were chosen. We in fact typed all 78 presently recognised HLA A,B,C and Dr antigens, but as no statistically significant results appeared we chose for brevity to tabulate only those antigens in which an increased frequency has been reported in either idiopathic Parkinson's disease or Von Economo's disease. We acknowledge in our discussion the fact that the two studies deal with different ethnic groups and are therefore not strictly comparable.

Stroke as an early manifestation of systemic lupus erythematosus

Sir: We have read with interest the article by Dr Haas entitled *Stroke as an early manifestation of systemic lupus erythematosus*.¹ We entirely agree that stroke can occur as an early manifestation of systemic lupus erythematosus. Unless the diagnosis is actively considered the true nature of the underlying disease may not be established. However, he did not stress that one of the difficulties in making a diagnosis is because of the lack of a specific diagnostic test for cerebral systemic lupus erythematosus. We would like to report a further case of systemic lupus erythematosus to illustrate the difficulties in arriving at the diagnosis and the particular use of skin biopsy in this case.

A 25-year-old aboriginal man was admitted four days after the sudden onset of right hemiplegia, mixed dysphasia and one grand mal seizure. Physical examination revealed no peripheral stigmata of vasculitis or infective endocarditis. Numerous pustular lesions were present over the buttocks and lower abdomen. His blood pressure was normal with regular pulses. No cardiac murmur or carotid bruit was detected. He had receptive and expressive

aphasia, right homonymous hemianopia, right facial weakness and right hemiplegia. The right deep tendon reflexes were brisker than the left, with right plantar response being extensor. Sensory testing was not possible because of dysphasia. Shortly after admission he developed transient episodes of pleuritic chest pain with radiologically proven bilateral basal pulmonary infiltrates and small pleural effusions. There was no evidence of infection to account for the pulmonary lesions.

The initial investigations showed a persistently elevated ESR, up to 122 mm/h with otherwise normal haematological and biochemical tests. CT head scan showed a large left cerebral infarction in the distribution of the middle cerebral artery territory. Antinuclear factor was strongly positive, with titre of 1/125 and a mixed staining. However, anti-DNA by membrane assay was not raised and serum C3 and C4 were normal. Although a vasculitic process like systemic lupus erythematosus was strongly suspected, normal anti-DNA activity and complement levels did not allow the diagnosis to be confirmed. However, antibodies to extractable nuclear antigen were positive (positive RNP and Sm antibodies). Muscle biopsy revealed only non-specific myositis with no arteritis. A skin biopsy showed granular IgM, IgA, IgG and C1q deposits at dermoepidermal junction and appendages. There was also *in vivo* speckled ANA. These findings helped to confirm the diagnosis of systemic lupus erythematosus. Prednisolone was then commenced about two months after the stroke. There was gradual fall in his ESR and some improvement in his neurological state. Before being discharged, he had a moderate degree of right hemiparesis, but was able to ambulate with a stick.

If one applies the newly revised systemic lupus erythematosus criteria of the American Rheumatic Association² the patient meets the criteria for systemic lupus erythematosus having four of the eleven criteria. He had seizure, pleurisy, fluorescent antinuclear antibody and anti Sm antibody. The new revision contains reference to anti Sm for the first time, and thus allows this case to meet the criteria. However, it is interesting to note that "stroke" is not included in the diagnostic criteria.

Hughes³ has commented that currently available routine laboratory tests are still not reliable enough for accurate assessment and management of cerebral systemic lupus erythematosus patients. DNA binding levels occasionally may be normal. In the most floridly psychotic systemic lupus