development of chorea was provoked by the administration of a well-known activator of mast cell secretion (mepivacaine 1%).

We cannot exclude the possibility that mast cell infiltration of the basal ganglia were responsible for chorea. However, we consider this possibility to be far less likely in view of the transient nature of the chorea and that no structural lesion could be demonstrated by the neuroradiological studies. In addition, the systemic mast cell disease has been reported to involve directly virtually all organs except the central nervous system.¹

Although no conclusion can be drawn from our report, we believe that the causation of the chorea in mastocytosis must be sought at a biochemical level rather than in a structural lesion.

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References

- Lewis R, Austen K. The mastocytosis syndrome. In: Fitzpatrick T, Eisen A, Wolf K, Freedberg I, Austen K, eds. *Dermatology in General Medicine*. New York: McGraw-Hill, 1987:1898-904.
- Soter N. Mastocytosis and urticaria pigmentosa. In: Provost T, Farmer E, eds. *Current Therapy in Dermatology*. St Louis: CV Mosby Co, 1985:243-5.
- Korenblat P, Wedner H, Whyte M, Frankel S, Avioli L. Systemic mastocytosis. *Arch Intern Med* 1984;144:2249-53.
- Ad Hoc Committee Research Group on Extraparapyramidal Disorders. World Federation Neurology: Classification of Extraparapyramidal Disorders. *J Neurol Sci* 1981;51:311-27.
- Di Bacco R, De Leo V. Mastocytosis and the mast cell. *J Am Acad Dermatol* 1982;7:709-22.
- Lewis R. Mastocytosis. *J Allergy Clin Immunol* 1984;74:755-64.
- Marsden CD. The basal ganglia. In: Swash M, Kannard C, eds. *Scientific Basis of Clinical Neurology*. London: Churchill Livingstone, 1985:56-73.

Absence of antibodies to cardiolipin in patients with Huntington's chorea, Sydenham's chorea and acute rheumatic fever

Sir: We have recently found an increased incidence of antiphospholipid antibodies (the lupus anticoagulant and antibodies to cardiolipin) in patients with chorea, associated with systemic lupus erythematosus and related diseases.^{1,2} Chorea also seems to be a not uncommon occurrence in patients presenting with a "primary" antiphospholipid syndrome.^{3,4} The administration of oral contraceptives to patients with antiphospholipid antibodies also appears to be associated with an increased risk of the development not only of thrombotic complications, but also of chorea.^{5,6} Chorea gravidarum seems particularly to be related to the presence of these antibodies.^{7,8} This emphasises the close relationship between both the external and the internal hormonal environment and the development of chorea.

In Sydenham's chorea, however, there is a different mechanism. Husby *et al* in 1976⁹ demonstrated that IgG antibodies in sera from children with rheumatic fever reacted with neuronal cytoplasmic antigens of the caudate and subthalamic nuclei of human brain, where they appeared to be preferentially increased. A higher proportion of sera (46.6%) were positive in children with chorea compared with those with carditis (14.0%). Absorption experiments indicated that the staining of caudate and subthalamic neurons represented cross-reactions between antigens present in Group A streptococcal membranes and neuronal cytoplasm.

Because of our interest in this subject we undertook a study of sera from three groups of patients; (A) Huntington's chorea, (20) (B) Patients with rheumatic fever, (20) (C) Patients with rheumatic fever and chorea, (6). Sera from Group A were obtained from patients attending the Institute of Neurology. Sera in groups B and C were obtained from the Rockefeller University, New York (Courtesy, Dr John Zabriskie) and the University of Pretoria, South Africa (Courtesy, Professor Richard Gledhill).

Antibodies to cardiolipin were estimated by a modification of the original ELISA by Gharavi *et al*.¹⁰ All specimens proved to be negative on this assay. We therefore conclude that the pathogenesis of the chorea occurring in SLE and with oral contraceptives in the presence of antiphospholipid antibodies is different from that seen in Sydenham's chorea or the hereditary Huntington's chorea.

Studies are presently underway to determine whether IgG and IgM cardiolipin

antibodies cross react with antigens present in caudate or subthalamic nuclei. The results of these studies are thus so far inconclusive.

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- Asherson RA, Derksen RHWM, Harris EN *et al*. Chorea in systemic lupus erythematosus and "lupus-like" disease: associated with antiphospholipid antibodies. *Semin Arthritis Rheum* 1987;16:253-9.
- Asherson RA, Hughes GRV. Antiphospholipid antibodies and chorea. *J Rheumatol* 1988;15:377-9.
- Asherson RA. The "Primary" antiphospholipid syndrome, Editorial. *J Rheumatol* (in press).
- Hodges JR. Chorea and the lupus anticoagulant. *J Neurol Neurosurg Psychiatry* 1987;50:368-9.
- Asherson RA, Harris EN, Gharavi AE *et al*. Antiphospholipid antibodies, chorea and oral contraceptives. *Arthritis Rheum* 1986;29:1535-6.
- Asherson RA, Farquharson RG, Harris EN, Hughes GRV. Complications of oral contraceptives and antiphospholipid antibodies: Reply to Letter of Bruneau *et al*. *Arthritis Rheum* 1988;31:575-6.
- Lubbe WF, Walker EB. Chorea gravidarum associated with circulating lupus anticoagulant in successful outcome of pregnancy with prednisone and aspirin therapy, case report. *Br J Obstet Gynaecol* 1983;90:487-490.
- Agrawal BL, Foa RP. Collagen vascular disease appearing as chorea gravidarum. *Arch Neurol* 1982;39:192-3.
- Husby G, Van de Rijn U, Zabriskie JB *et al*. Antibodies reacting with cytoplasm of subthalamic and caudate nuclei neurons in chorea and acute rheumatic fever. *J Exp Med* 1976;144:1094-1110.
- Gharavi AE, Harris EF, Asherson RA, Hughes GRV. Anticardiolipin antibodies: isotype distribution and phospholipid specificity. *Ann Rheum Dis* 1987;46:1-6.