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.7

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.

Accepted References

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

Syr. 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.12 Chorea also seems to be a not uncommon occurrence in patients presenting with a "primary" antiphospholipid syndrome.13 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.14 Chorea gravidarum seems particularly to be related to the presence of these antibodies.15 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 1976 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, (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.8 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 heredo-familial 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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