The World Federation of Neurosurgical Societies hope that the scale will prove acceptable to all those concerned with the management of patients with subarachnoid haemorrhage and will be adopted widely both in day to day clinical practice and in reports in scientific journals. Members of the Committee undertook to bring the scale to the attention of editor and readers of appropriate journals.

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References

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Chorea: a new manifestation of mastocytosis

Sir: Mastocytosis is an uncommon disease, characterised by the proliferation of mast cells in various organs of the body, which presents in a limited cutaneous or a systemic form. The central nervous system manifestations of mastocytosis include headache, dizziness, seizures and alterations in cognitive function. The occurrence of chorea, to the best of our knowledge, has never before been described in mastocytosis. We recently examined a patient with mastocytosis who developed choreic movements.

A 13 year old girl was admitted to hospital for study of pruritic skin lesions consisting of reddish-brown maculopapules. A skin biopsy was done under local anaesthetic (mepivacaine 1%). Approximately 4 hours later, she developed involuntary movements. Examination revealed choreatic movements specially involving the upper extremities and the orofaciolingual muscles, with the trunk and the lower extremities involved to a lesser degree. The hands showed choreothetoid movements. Neurological examination was otherwise normal. General physical examination was normal save for skin lesions. One day after initiation of the chorea, treatment with cyproheptadine (12 mg/d) and ketotifen (2 mg/d) was instituted. Resolution of the abnormal movements began gradually during the third day after their onset and completely returned to the premorbid state on the fifth day. One year later, the patient has remained well without further choreic episodes.

Pregnancy, birth and early development were normal. There was no history of chorea or rheumatic fever and the patient had not been treated with chorea-inducing drugs. Family history was not significant for any neurological disease.

The following investigations were all normal; haemoglobin determination, WBC and differential counts, ESR, urea and electrolytes, creatinine, blood glucose, liver function tests, calcium and phosphate, cholesterol, triglycerides, uric acid, serum protein electrophoresis, quantitative assays for immunoglobulin levels, C3, C4, autoantibody screening, syphilis tests, coagulation system tests, thyroid function tests, caeruloplasmin, serum and urinary copper, 24 hour 5-hydroxyindolacetic acid and urinary histamine levels, skull and chest radiographs, cerebrospinal fluid, electroencephalography, auditory and visual evoked potential and CT and MRI of the brain. The skin biopsy specimen showed mast cell infiltration of the dermis.

The most remarkable finding in our case is the presence of chorea that, to our knowledge, has never before been described in mastocytosis. Choreic movements were considered to be caused by mastocytosis, not just because no evidence of other aetiology was found, but also in view that the administration of a well-known activating of a mast cell secretion as topical anaesthetic provoked the choreic movements.

The role of the mast cell in production of disease appears to be twofold. As a consequence of a high tissue mast cell concentration, there are both local effects of mast cell infiltration, as it interferes with organ function, and systemic effects resulting from release of mast cell mediators such as histamine, heparin, prostaglandins and other peptides.

Chorea can be the result of structural or functional striatum damage. In our case, the possible explanation of these movements is that as an effect of a sudden and greatly elevated level of released mast cell mediators, mastocytosis may induce biochemical alteration in the basal ganglia. This hypothesis is supported by the fact that the
A universal subarachnoid hemorrhage scale: report of a committee of the World Federation of Neurosurgical Societies.

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