Letters

A universal subarachnoid haemorrhage scale: report of a committee of the World Federation of Neurosurgical Societies

Sir: At its meeting in Barcelona in September 1987, the Executive Committee of World Federation of Neurosurgical Societies received and unanimously approved a report from a committee that had been working for six years to devise a simple, reliable, clinically valid scale for grading patients with a subarachnoid haemorrhage. The committee's view was that the scale was needed for describing changes in an individual patient at different times, for estimating prognosis and for standardising assessment of management in different groups of patients; a requirement was that the scale would meld with currently used scaling systems.

The committee took into account an analysis of data from the international cooperative aneurysm study which contained 3521 patients from 68 countries; this showed that the two most important prognostic factors were the level of consciousness (important for the prediction of both death and disability) and the presence or absence of hemiparesis and/or aphasia (important only for disability in survivors). The analysis had shown that if consciousness was normal, headache and/or a stiff neck did not significantly affect outcome.

The committee resolved that five grades only should be used for patients with subarachnoid haemorrhage; patients with an unruptured aneurysm should be identified separately, or classified as 'zero'. It believed that the Glasgow Coma Scale should be used to assess the level of consciousness, because of its world wide acceptance in assessment of coma from head injury. The only additional factor should be the presence or absence of major focal deficit to differentiate between grades two and three. The committee also resolved that in assessing outcome from subarachnoid haemorrhage, categories of the Glasgow Outcome Scale should be used: Dead, Vegetative Survival, Severely disabled; Moderately disabled; Good recovery.

Table 1: WFNS SAH grading scale

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<thead>
<tr>
<th>Grade</th>
<th>GCS</th>
<th>Motor deficit</th>
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<tbody>
<tr>
<td>I</td>
<td>15</td>
<td>-</td>
</tr>
<tr>
<td>II</td>
<td>14–13</td>
<td>-</td>
</tr>
<tr>
<td>III</td>
<td>14–13</td>
<td>+</td>
</tr>
<tr>
<td>IV</td>
<td>12–7</td>
<td>±</td>
</tr>
<tr>
<td>V</td>
<td>6–3</td>
<td>±</td>
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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

Accepted 20 June 1988

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, have 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 activator 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
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.

Chorea also seems to be a not uncommon occurrence in patients presenting with a "primary" antiphospholipid syndrome. 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. Chorea gravidarum seems particularly to be related to the presence of these antibodies. 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, (C) Patients with rheumatic fever and chorea, (D) 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. 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 heredofamilial 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 far inconclusive.

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References


Chorea: a new manifestation of mastocytosis.

L M Iriarte, J Mateu, G Cruz and J Escudero

*J Neurol Neurosurg Psychiatry* 1988 51: 1457-1458
doi: 10.1136/jnnp.51.11.1457-a

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