Scoring Clinical Functions in Subacute Sclerosing Panencephalitis

We read with great interest the recent contribution by Aydin et al in the February issue of the American Journal of Neuroradiology. They demonstrated gray matter volume reduction in the frontotemporal cortex of patients with subacute sclerosing panencephalitis (SSPE) without any apparent lesions on MR imaging. The authors achieved excellent results from their imaging assessment and should be strongly commended. However, we would like to address an issue that revolves around possible improvements in the clinical scoring of patients.

The authors did not find any significant correlation between the neurologic disability index (NDI) scores of patients (7–14 years of age) and the distribution of cortical gray matter. They speculated that the lack of correlation might be because they studied only patients in the initial stages of the disease, so the NDI scores were distributed in a narrow range. As the authors described, the NDI scoring is a gross evaluation of motor and sensory functions and mental states based on an examiner rating (0–4 points) neurologic signs as mild/moderate/severe or frequent/occasional. The considerable subjectivity involved has been a concern; there is a suggestion that it is difficult to view a narrow score difference as significant. As well, the possible full cognitive scores of healthy children younger than 10 years of age can vary greatly. Using NDI for younger children could cause some variance in scores among examiners.

We often encounter the problem when evaluating clinical functions of patients with SSPE. Several new approaches, including a Functional Independence Measure (FIM), have recently been applied to various neurologic diseases. We have introduced a modified version of the FIM questionnaire by adding questions with examples that are appropriate for younger children (1–12 years of age). This was done to help us evaluate pediatric and adult patients, including those with SSPE. Medical staff or parents answer the questions related to actual movements, and scores are determined reproducibly in accord with an appropriate flow-chart example, which was standardized by using healthy control adults and children. For example, cognitive scoring has a maximum of 80 specific questions to pose, until finally the patient’s status can be precisely determined. However, the lack of SSPE-specific scores (ie, myoclonus or autonomic function) in FIM naturally means that we also use NDI.

Widening the narrow scoring ranges of NDI in combination with some newly established measures would be useful in eliciting correlations between clinical status and radiologic findings.

References

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