angiography. There are no specific guidelines with respect to duration of therapy but treatment is continued for a minimum of 6 to 12 months following remission. Atypical PACNS should be treated with 6 to 12 glucocorticoids plus additional immunosuppressive treatment if necessary to achieve remission. Treatment of BACNS is less aggressive, usually glucocorticoids for six months, often with adjunctive calcium channel blockers. The outcome in patients with PACNS treated with immunosuppressive treatment is less bleak than previously supposed with less than 10% mortality and approximately 20 to 30% developing significant disability. Patients diagnosed with BACNS usually do well, with 94% showing significant recovery and 71% showing no evidence of long term disability.

Conclusions

The central nervous vasculitides encompass a large number of primary and secondary disorders with a wide differential diagnosis. The presentation is variable and specific tests are lacking. Accurate diagnosis is important in order to exclude possible mimics which may require different therapeutic approaches and to avoid unnecessary immunosuppressive treatment with its attendant risks. There is increasing evidence that primary CNS vasculitis is composed of differing clinical subsets (on the basis of clinical, laboratory, angiographic and pathological findings) and these subsets vary in both their prognoses and treatment.

References


Carphologia, or Flocillation

Respecting the movement of the hands, I have these observations to make: when in acute fevers, pneumonia, phrenitis, or headache, the hands are waved before the face, hunting through empty space, as if gathering bits of straw, picking the nap from the coverlet, or tearing chaff from the wall—all such symptoms are bad and deadly.

Hippocrates Book of Prognostics

Apparently aimless plucking movements have been named either carphologia or flocillation, because of their fancied likeness to picking up pieces of straw or wool, respectively. The term carphology has recently been adopted by a columnist in a rival journal, but it is difficult to find any specific articles on the subject of these movements (see absence of references on Medline). Perhaps the account by Hippocrates (or his school), which I believe to be the original, said all that needs to be said. The movements have apparently been observed in dementing disorders such as Alzheimer’s disease or vascular dementia, delirium (phrenitis, literally “brain fever”), whence our word frenzy, may perhaps be equivalent to delirium), and some psychiatric disorders, and may possibly reflect frontal lobe dysfunction. Although their description is of great antiquity, these movements may still be misinterpreted.

A 58-year-old man presented with rest and action tremor of the right (dominant) arm, slow quiet speech, hypomimia, and with examination findings of mild rigidity, micrographia, and reduced right arm swing. Concurrently, he had developed progressive memory problems sufficient to prevent him from running his business; Mini-Mental State examination score was 19/30, with slowed responses. Movement disorders included mild rigidity, micrographia, and reduced right arm swing. The recognition, diagnosis and management of cerebral vasculitis: a European survey. Eur J Neurol 2002;9(4):343-7.

References