

angiography. There are no specific guidelines with respect to duration of therapy but treatment is usually continued for a minimum of 6 to 12 months following remission. Atypical PACNS should be treated with 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.¹¹ Patients diagnosed with BACNS usually do well, with 94% showing significant recovery and 71% showing no evidence of long term disability.¹²

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.

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Neurological Signs

Carphologia, or Floccillation

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* 4

Apparently aimless plucking movements have been named either carphologia or floccillation, 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,¹ 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.² 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. Neuropsychological assessment showed severe impairment on the Mattis Dementia Rating Scale, particularly on subtests of initiation/perseveration and attention, and also on the Delis-Kaplan Executive Function System, indicating a frontal-subcortical profile of dementia. Neurological signs were unresponsive to levodopa preparations. Over the subsequent two year period the patient developed progressive cognitive decline, slow saccadic eye movements, levator inhibition, retrocollis, the applause sign, and recurrent falls, on

one occasion causing a fracture of the proximal phalanx of the ring finger, all felt to be consistent with a clinical diagnosis of progressive supranuclear palsy (PSP).³

Shortly after admission to a nursing home because of ongoing falls, care staff reported the patient to be "self-harming". Specifically, he was reported to pinch repeatedly the skin on his left arm and chest with his right arm in a rough, jerky manner, sometimes sufficiently hard to cause bruising or even draw blood. These movements were observed in the clinic as intermittent picking or plucking movements on the clothing or skin with the tremulous right hand. Patient questioning revealed no suicidal ideation or desire to self-harm.

The superimposition of a jerky action tremor on carphologia may increase the amplitude and reduce the accuracy of these otherwise innocuous movements, such that they might pinch and even break the skin, and hence be misinterpreted as self-injurious behaviour. Clearly, the identification of individuals who self-harm is of fundamental importance because of the greatly increased risk of subsequent suicide.⁴ Passive self-harm in patients with dementia resident in nursing homes, such as refusal to eat, drink, or take medications, is of similar concern since this may be associated with increased mortality.⁵ Self-injurious behaviour is rare in movement disorders, although it may be a feature in some (e.g. neuroacanthocytosis, Lesch-Nyhan disease). Involuntary, tremulous carphologia should not be mistaken for self-harm.

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