Micrographia associated with a parietal lobe lesion in multiple sclerosis

N J Scolding, A J Lees

Abstract
The occurrence of micrographia in a 52 year old woman two years after an isolated episode of painful sensory disturbance led to the diagnosis of multiple sclerosis. Her handwriting returned to normal after a course of intravenous methylprednisolone. Previous reports of movement disorders occurring in the context of multiple sclerosis are briefly reviewed. The finding on MRI studies of an enhancing lesion in the dominant parietal white matter supports Kinnier Wilson’s suggestion that the anatomical origin of micrographia lies in the cerebral hemisphere rather than the corpus striatum.

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With the notable exception of motor symptoms reflecting involvement of the cerebellum and its connections, movement disorders are unusual in multiple sclerosis. Of those which are described, most are positive phenomena, and paroxysmal events, such as the classically described tonic spasms, are seen more commonly than sustained symptoms. We describe the case of a 52 year old woman, in whom the onset of micrographia two years after a spontaneously resolving painful sensory disturbance led to further investigations that confirmed the diagnosis of multiple sclerosis. Treatment with intravenous steroids was accompanied by a rapid resolution of her micrographia.

Case report
A 50 year old right handed woman developed a superficial burning sensation in her right anterior thigh spreading to the suprapubic area, together with hyperpathia, two months after an oophorectomy for benign ovarian cysts. This was unaccompanied by any motor or sphincter symptoms, and there were no objective abnormalities on examination. Investigations, including blood count, erythrocyte sedimentation rate, glucose, liver function, protein electrophoresis, ultrasound scan of pelvis, and MRI of the spinal cord were all normal, and her symptoms resolved spontaneously after three months.

Two years later, two months after minor surgery for a Meibomian cyst, she noticed an abrupt change in her handwriting. Her script had become smaller and less clear; the act of writing had become laborious. Other activities requiring fine finger control, including sewing, tying parcels and peeling vegetables were unaffected, but she had noticed a slight deterioration in the speed and accuracy of her right hand using a word processor keyboard.

At this time, general physical examination was unremarkable, and there were no neurological abnormalities in the cranial nerves or in the lower limbs. Power, tone and tendon reflexes, and sensation were all normal. Language function was entirely normal, as were other tests of higher mental function. Her handwriting was slow and small, however (fig 1), with a tendency to decrease further in size as she continued writing; she was unable to increase the size to command. Copying tests revealed no constructional apraxia and there were no other visuospatial abnormalities. There was very slight impairment of rapid alternating movement of the right hand and finger. Tests of mimicking, mime, and object use revealed no apraxia. Facial expression was normal, there was no glabellar tap response, no bradykinesia, and her gait was normal.

Routine blood tests were again normal or

Figure 1 Samples of the patient’s handwriting (top) before, (centre) during, and (bottom) three months after the episode of micrographia.
negative, including autoimmune and treponemal serological tests. Visual, auditory, and somatosensory evoked potentials (from both upper limbs) were also normal. An MRI scan of her head showed multiple T2 high, T1 low signal lesions in the white matter of both cerebral hemispheres, however, particularly in the periventricular areas. The largest lesion, in the left parietal white matter, showed rim enhancement (fig 2); it did not involve the basal ganglia. None of the lesions exhibited a mass effect, and the appearances were interpreted as being strongly suggestive of multiple sclerosis.

A course of intravenous methyprednisolone (1 g daily for three days) was associated temporally with a good improvement in the size and fluency of her handwriting (fig 1 (bottom)); this improvement has thus far (now six months after her original symptoms) been maintained. Furthermore, repeated examinations have consistently continued to fail to reveal the emergence of any parkinsonian signs.

Discussion

The clinical picture of an episode of painful sensory disturbance spontaneously resolving over two to three months, followed by an episode of motor disturbance that resolved after a course of high dose intravenous steroids, is highly suggestive of multiple sclerosis, a diagnosis strongly supported by the MRI abnormalities.

Extrapyramidal movement disorders are unusual in multiple sclerosis; of these, negative phenomena are rarer still, although paroxysmal akinnesia is reported.1 A small number of cases with pill rolling tremor have been described,14 but non-paroxysmal akinnesia and extrapyramidal rigidity are rare. Mao et al2 reported two cases with tremor, bradykinesia, hypomimia, and rigidity, one of whom was treated (unsuccessfully) with intravenous steroids, and Vierregge et al3 also reported two cases of parkinsonism associated with multiple sclerosis; both had hyperintense lesions on spin-echo MRI that involved the basal ganglia, and both made a significant response to intravenous steroid treatment.

Our case was notable for the occurrence of micrographia without other features of parkinsonism. Micrographia was first described by Pick in 1903; the association with parkinson's disease was soon recognised, and has since been accepted as a feature of diagnostic significance.8 Kinnier Wilson described cases of micrographia in cerebral syphilis and in arteriosclerosis, however, with subsequent postmortem studies revealing the absence of involvement of the corpus striatum.9 He also cited cases of severe postencephalitic parkinsonism with profound immobility and rigidity and yet normal handwriting, and of severe micrographia in a female with otherwise very minor parkinsonian symptoms.

McClennan et al have more recently confirmed that “micrographia is totally separable from tremor and rigidity”, basing their conclusion on a detailed study of 95 patients with micrographia associated with parkinsonism.10 They found a lack of correlation between micrographia and the side, severity, and duration of classical parkinsonian features; micrographia often preceded other manifestations by as much as three to four years, and exhibited a very variable response to L-dopa treatment, by contrast with its more predictable beneficial effect on other symptoms and signs.

Court described an association of depression with micrographia,11 although McClennan's group found that “elevation or depression of mood...did not...have much effect on micrographia” — which they also noted did not improve with antidepressant treatment.10

Although McClennan et al do not speculate on the anatomical substrate for micrographia, Kinnier Wilson concluded that abnormalities in the cerebral cortex were much more likely to underly micrographia than disorders in the corpus striatum. The matter remains unresolved: Martinez Vila et al recently reported a patient with isolated micrographia associated with a lenticular haematomata.12 Our case supports Kinnier Wilson's suggestion, as MRI scanning revealed no abnormality in the basal ganglia but a large lesion in the dominant parietal white matter. The lesion enhanced with gadolinium so that it is likely to reflect active disease13 and to be responsible for her symptoms.

Amnesia

Amnesia that includes the loss of memory for personal details seldom causes the neurologist diagnostic difficulty. The condition is not related to organic brain disease and it is perhaps surprising that novelists continue to regard it as such. Sometimes, for example with Saki, the problem is pursued for its comic possibilities. The whole of Rebecca West's novel, The return of the soldier, is constructed around the premise of memory loss for personal affairs. Dickens's characterisation is humorous in that Manette appears to retain some insight into his behaviour; one would almost consider the problem to be one of malingering, though clearly that was never Dickens's intention.

Charles Dickens, 1859, A tale of two cities
Miss Pross, with a terrified face, was at her ear. "O me, O me, all is lost!" cried she, wringing her hands. "What is to be told to Ladybird? He doesn't know me, and is making shoes!... "Doctor Manette. My dear friend, Doctor Manette!" The Doctor looked at him for a moment—half enquiringly, half as if he were angry at being spoken to—and bent over his work again.

So far it was possible to comprehend him without overstepping those delicate and gradual approaches which Mr. Lorry felt to be the only safe advance, he at first supposed that his daughter's marriage had taken place yesterday. An incidental allusion, purposely thrown out, to the day of the week, and the day of the month, set him thinking and countering, and evidently made him uneasy. In all other respects, however, he was so composedly himself, that Mr. Lorry determined to have the aid he sought. And that aid was his own.

"My dear Manette, it is the case of an old and prolonged shock, of great acuteness and severity to the affections, the feelings, the—the—as you express it—the mind. The mind. It is the case of a shock under which the sufferer was borne down, one cannot say for how long, because I believe he cannot calculate the time himself, and there are no other means for getting at it. In the case of a shock from which the sufferer recovered, by a process that he cannot trace himself—but, unfortunately, there has been"—he paused and took a deep breath—"a slight relapse."

George Eliot, 1860, The mill on the floss
"Dr. Turnbull thought him a deal better this morn-
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