added to the typical “alien hand” automatisms (fig 1C). The spreading of the ictal activity from rCMA to premotor and motor cortical areas resulting in the additional tonic phenomena is highly probable. The ictal EEG recordings from the date of the patient's admission with pure ictal “alien hand” signs revealed only very discrete non-specific changes in the central and right paracentral region (Cz and C4). In this case, an underlying ictal activity in the “hidden” lesional/peri-lesional region is strongly anticipated. But absolute certainty that the rCMA is solely responsible for the paroxysmal “alien hand” syndrome could only be drawn from invasive EEG investigation or electrical cortical stimulation. This treatment was not necessary for clinical reasons and thus was not undertaken in our patient.

In conclusion, rostral CMA very probably plays a crucial role in the production of ictal automatic motor behaviour of the contralateral hand. This finding may further implicate the participation of this part of the caudal anterior cingulate cortex in the genesis of ictal limb automatisms in epileptic patients.

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Atypical micrographia associated with corticostriatal white matter lesions in systemic lupus erythematosus

Micrographia is a heterogeneous condition in which various parts of the CNS may be involved. An anatomical substrate for micrographia, however, remains to be established. Here, we report on a patient with systemic lupus erythematosus (SLE), who presented with atypical micrographia, which was associated with bilateral lesions in the corticostriatal white matter.

Case report

A 30-year-old right-handed woman was diagnosed as having SLE in 2002. She had concomitant leukocytopenia, arthritis, nephritis and high titres of antinuclear antibodies, which satisfied the American Rheumatism Association criteria for SLE. Thereafter, maintenance treatment using corticosteroids was started. She was admitted to the Niigata University Medical and Dental Hospital, Niigata, Japan, in February 2005, because of a high fever and headache with affective incontinence.

Examination showed her muscle strength and the sensory function of her extremities to be normal. She did not have involuntary movements or akinetic-rigid symptoms, as her gait was normal and no rigidity was observed in the neck, body and extremities. No impairment was seen in the rapid alternative movements of her hands. She was well oriented and cooperative. Aphasia was absent; her speech was well articulated and grammatically correct, and she had no difficulty in naming objects. She had no abnormalities in praxis, showing an excellent capacity in imitating and pantomiming, and in using tools with either hand. She could perform a fist–palm–alternating task swiftly. Orofacial apraxia, visuospatial disturbance, unilateral spatial neglect or visual agnosia was not observed. She had no memory impairment. No general intellectual deterioration was seen; her score in the revised Hasegawa Dementia Scale, which is widely used for intellectual screening in Japan, was 28 of 30 (cut-off 20/30).

During the neuropsychological evaluation, the quality of the patient's spontaneous handwriting deteriorated—that is, the characters or drawings were always small. To evaluate this symptom in detail, we asked the patient to write a Japanese character and a Roman alphabet repetitively, and draw a triangle, a circle, a square and a star in a

manner identical to the samples given by the examiner. The initial characters of the patient's handwriting were smaller than those in the samples, and the small size of the characters was constant throughout the sequence of her handwriting (fig 1A). Furthermore, a marked initial reduction in size was observed in the symbols drawn compared with the corresponding samples (fig 1B). These copying tasks were carried out using her dominant (right) hand. Neither hesitation nor slowness in handwriting was observed. The patient could correctly evaluate the size of objects, as she could sort various objects on the table by size. She acknowledged the disorder, complaining that she could write only such small characters or symbols despite great efforts to write in the same size as that in the samples.

Examination of CSF showed a normal cell count, although a mild rise was observed in IgG level. The patient tested negative for serum antiphospholipid antibodies. MRI of the brain carried out on day 2 of admission showed hyperintensity signal lesions in the bilateral dorsal part of the striatum and adjacent white matter on T2-weighted and diffusion-weighted images (fig 2A). These lesions were contrast enhanced with gadolinium. The other parts of the CNS, however, were intact. Single-photon emission CT using ethyl cystine dimer labelled with technetium-99m carried out on day 12 of admission showed a decreased perfusion of the bilateral striatum and surrounding white matter.

After treatment with 1000 mg of methylprednisolone for 5 days, the initial reduction in the size of handwriting had improved (fig 1C). MRI of the brain carried out 3 months after the treatment showed that the white matter lesions had reduced in size (fig 2B).

Informed consent was obtained from the patient before carrying out the evaluations and giving treatment.

Figure 1 (A) Repetitive writing of a Japanese character and a Roman alphabet was requested. The sizes of the characters and letters in the patient's handwriting are smaller than those in the examiner's handwriting, and this small size is constant throughout the handwriting sequence. (B) Before the steroid pulse treatment, a marked initial reduction in size is observed on drawing a triangle, a circle, a square and a star. (C) The initial reduction in size is obviously improved after the steroid pulse treatment.

Figure 2 (A) Brain MRI carried out on day 2 of admission shows hyperintensity signal changes in the bilateral dorsal part of the striatum and adjacent corona radiata in T2-weighted and diffusion-weighted images. (B) Three months after the steroid pulse treatment, the area of hyperintensity signal in the corona radiata decreased in size as observed in T2-weighted images.

Discussion

Neurological evaluation showed that the symptoms of the patient were not accompanied by intellectual deterioration or neuropsychological abnormalities. Additionally, the patient was cooperative and well oriented to the copying tasks. These findings indicate that the smallness of the handwriting presented here corresponds to micrographia. The micrographia of the patient, however, is atypical compared with the common form of micrographia in Parkinson's disease, which is characterised by a spectacular shrinking in character size as the handwriting progresses.¹ This feature of Parkinson's disease is presumably attributed to the smallness of motor output and the use of visual feedback of a previous or an ongoing stroke recursively to programme the subsequent strokes during handwriting.² With regard to the micrographia presented here, the constant size of the character during the handwriting sequence suggests that the patient preserves the ability to use the feedback mechanism that provides the trial-by-trial adaptations of character size in handwriting.²

The micrographia in this patient was further characterised by an initial marked reduction in size. This feature is considered to be more prominent than that in patients with Parkinson's disease. Scolding and Lees³ showed that an isolated lesion in the parietal white matter caused micrographia, in which an initial reduction in the size of the handwriting was also noted. Strikingly, the MRI of this patient showed that the lesions affected mainly the corticostriatal white matter. Furthermore, the shrinkage of such lesions was consistent with the improvement in the symptom. Experimental data suggest that the functions of the corticostriatal pathway are associated with spatial attention⁴ and motor skill learning,⁵ which presumably maintain the quality of handwriting. Interestingly, the corticostriatal pathway is insidiously affected in patients with Parkinson's disease too.⁶ We therefore speculate that this disruption may contribute to aberrant kinesthetic processing in which the sensorimotor apparatus is "set"

small.⁷ This aberration may underlie the atypical micrographia of this patient. Taken together, these findings suggest that the disruption of the corticostriatal pathway is responsible for the initial reduction in the size of the handwriting.

Although the question of whether it is the dominant or non-dominant hemisphere that is principally implicated in this symptom remains elusive, our observations raise the possibility that the corticostriatal pathway contributes to the adjustment of the sensorimotor input in the control of elaborate movements such as handwriting.

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Myoclonus as an acute complication of low-dose hydromorphone in multiple system atrophy

Myoclonus has been described as a neuro-excitatory side effect of high doses of opioids. It has been seen almost exclusively in patients receiving palliative care after long-term administration of opioids. Gradual accumulation of opioid metabolites has been thought to cause opioid-induced myoclonus.¹ Naloxone has usually been ineffective in resolving this condition. In this study, we present a case of severe generalised myoclonus after a patient with known multiple system atrophy received a low dose of intravenous hydromorphone. The generalised myoclonus reversed completely immediately after he received naloxone.

Case report

A 75-year-old man with a history of multiple system atrophy with parkinsonism was admitted to the Montreal General Hospital, Montreal, Canada, with a hip fracture. Drugs administered on admission were fludrocortisone, glizide, intravenous immune globulin (for bullous pemphigoid) and midodrine. He was not taking levodopa because of severe orthostatic hypotension. To treat his pain, 1 mg of intravenous hydromorphone was given over 20 min. About 35 min after receiving the hydromorphone infusion, the patient had decreased consciousness, severe generalised myoclonus and rigidity. The neurology service was consulted.

On examination, the patient was difficult to rouse. He inconsistently opened his eyes with sternal rub, and would occasionally localise sound. No oral response was evident, and he did not obey motor commands. His pupils were pinpoint. Motor examination showed generalised rigidity. Throughout the examination, there were continuous, extremely severe multifocal myoclonic jerks. The myoclonus occurred in all four limbs, the axial musculature and the face. Movements were continual and asynchronous, about 4–6 jerks/s. Stimulus sensitivity could not be assessed because of the high baseline frequency. Plantar reflexes were downgoing bilaterally.

Laboratory values showed normoglycaemia and normal serum biochemistry, including calcium levels and renal function. Despite the low dose given, an adverse reaction to hydromorphone was suspected, and 0.4 mg of naloxone, an opioid antagonist, was given. Within seconds of administration, his condition improved dramatically, with a complete reversal of the myoclonus and a return to normal consciousness. As the effects of the naloxone dissipated, the patient had a gradual reappearance of the myoclonus, but never at the same intensity. The symptoms resolved completely over the next 2 h. Acetaminophen without opioids was used for subsequent analgesia. A resting