Hypothalamic amnesia and frontal lobe function disorders after Langerhans cell histiocytosis

Langerhans cell histiocytosis (LCH), a rare disease previously known as histiocytosis X, is characterised by abnormal cell proliferation. If the CNS is invaded, the hypothalamus is the typical site location. There are virtually no neuropsychological data on hypothalamic LCH sequelae. Memory disorders in the context of posterior but also anterior hypothalamic lesions, regardless of the aetiology, have in most cases been attributed to the involvement of the mammillary bodies (MB). However, Ptak et al reinterpreted a case, acknowledging that “Damage of the anterior hypothalamus, rather than the mammillary bodies, may [ . . .] have been responsible for [the observed] confabulatory amnesia” (page6).2 An interesting question is whether neuropsychological deficits are secondary to hypothalamic damage in itself or to a disconnection syndrome. The latter is based on the bilateral hypothalamic nucleus—widespread brain complex connections. Two subsystems are of particular interest in the present case, the hippocampus-fornix—hypothalamus-MB circuit, and the amygdala-stria terminalis plus caudate nucleus—hypothalamus circuit.

We report the case of a patient who presented with hypothalamic LCH and underwent thorough neuropsychological, radiological, and metabolic assessments.

Case report

A right handed woman (date of birth 1931), was admitted (March 2000) to the Hôpitaux Universitaires’ Neurology Unit (Strasbourg, France) complaining of memory deficits. In 1990, bronchopulmonary biopsies had led to the diagnosis of LCH. In 1992, she was presented with hypothalamic LCH and underwent thorough neuropsychological, radiological, and metabolic assessments.

Figure 1 T1 weighted coronal MR image showing a circumscribed hypothalamic tumour.

Secondary to hypothalamic LCH, our patient developed amnesia, dysexecutive syndrome, and personality alteration, in an otherwise normal context.

Ptak et al described a similar case (amnesia and frontal lobe dysfunction) in the context of circumscribed hypothalamic lesions.1 The postmortem diagnosis of sarcoidosis did not exclude the possibility of LCH. As the patient's lesion involved mostly the medial hypothalamus, the authors accounted for the symptoms in terms of the strong reciprocal connections with the anterior limbic structures. Further studies have demonstrated connections from the medial prefrontal cortex to the anterior and ventromedial hypothalami and those from the orbital region to the lateral hypothalamus.3

Comment

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In this case, the large lesion (optic chiasm, pituitary stalk, and MB), restricted to the hypothalamus, renders the findings particularly interesting in terms of hypothalamic connections. Given the evidence that LCH in this case is not an infiltrating but an encapsulated form of the disease, the small targeted zones of hypometabolism are not thought to be directly caused by the histiocytosis condition. They could be accounted for in terms of disconnections resulting from the hypothalamic lesion. The deficit in the memory consolidation process could reflect a dysfunction of the hippocampus-fornix-MB circuit. Failure of retrieval processes could reflect the prefrontal-hypothalamic connections. Moreover, the patient's false recognitions and confabulations showed the frontal involvement suggested by Ptak et al.4 However, for the time being, we are unable to account for the parietal and upper brain stem hypometabolism.

The hypothalamic hypermetabolism is interpreted in terms of an index of the active ongoing disease process.

In conclusion, we suggest that our patient's neuropsychological impairments, with their catastrophic consequences for daily life, require abnormality beyond the MB and might reflect the “superadditive” effect of damage at different, strategically interconnected sites. Theoretical considerations based on the mere “addition” of hypothalamic lesions plus a few very small hypometabolic zones at various sites in the brain would most probably not have predicted our patient's present cognitive status. Finally, given the paucity of hypothalamic LCH cases reported in the literature and the virtual absence of neuropsychological examination in those rare cases, the possibility of an as yet unknown specific effect of LCH on the CNS cannot be ruled out.

References

2 Orung D, Price JL. The organisation of networks within the orbito medial and frontal prefrontal cortex of rhesus monkeys and humans. Cerebral Cortex 2000;10:206–19

Meningioma of the optic nerve sheath: treatment with hydroxyurea

The best treatment of optic nerve sheath meningiomas remains controversial. Recent reports have emphasised the efficacy of fractionated stereotactic or conformal radiotherapy, and some clinicians favour this approach instead of surgery or observation.2 On the other hand, a beneficial effect of hydroxyurea on unresectable, residual, and recurrent meningiomas has been reported in...
A 46 year old woman presented with painless and progressive right sided visual failure for two years. On admittance, visual acuity of the right eye was 0.05. In addition, there was an afferent pupillary defect, and swelling of the right eye was 0.05. In addition, there was an afferent pupillary defect, and swelling of the right eye was increased to > 200 ms, and computed perimetric examination showed a significant recovery of visual field (fig 2B). However, cranial MRI showed no detectable change in tumour size (volume 0.15 cm3; fig 1B). At the time of writing the disease had remained stable for a further 18 months now.

Comment
This case shows the clinical value of hydroxyurea in the management of optic nerve sheath meningiomas, although there was no detectable decrease in tumour size. There is increasing evidence for the benefit of radiotherapy in optic nerve sheath meningiomas. Andrews et al reported an improvement in vision in 10 of 24 cases (42%) after treatment with fractionated stereotactic radiotherapy alone. A comparison of long term visual outcome by Turbin et al showed better results for patients treated by conventional multport or conformal planned delivery of radiotherapy than by surgery plus radiation, surgery alone, or observation during the follow up period. However, radiotherapy is associated with relevant treatment related morbidity (13% and 33.3% in two studies12). As follow up of the available case series is limited and these tumours may pursue a stable course for many years, the appropriate time for therapeutic intervention is unclear. In the present case, a profound deterioration of visual acuity led to the initiation of treatment. Hydroxyurea may be a reasonable therapeutic alternative to radiotherapy. Side effects of hydroxyurea such as myelosuppression, raised liver enzymes, and rashes are generally mild, easy to monitor, and reversible.

As the neuroradiological characteristics were unequivocal in our case, and as histological verification of optic nerve sheath meningiomas carries a high risk of irreversible damage to the optic nerve, the diagnosis was made purely by radiological means. This approach is in accordance with current standard of diagnostic measures of optic nerve sheath meningiomas based on clinical details and high quality neuroimaging without pathological confirmation.

References
1 Andrews DW, Faroozan R, Yang BP, et al. Fractionated stereotactic radiotherapy for the treatment of optic nerve sheath meningiomas: preliminary observations of 33 optic nerves in various series.13 We report a patient with a meningioma of the optic nerve sheath and nearly complete visual loss who was successfully treated with hydroxyurea alone.

Figure 1 Sagittal T1 weighted magnetic resonance imaging, angulated along the course of the right optic nerve (time of repetition 500 ms, time of echo 16 ms, slice thickness 4 mm), following Gd-DTPA administration. It shows a homogeneously enhancing, 5 × 10 mm fusiform tumour, originating from the upper part of the right optic nerve sheath and compressing the nerve below.

Figure 2 Computed perimetric examination of the right eye: (A) before treatment; (B) after 10 months of continuous treatment with hydroxyurea (20 mg/kg body weight/day). Black squares, absolute scotoma; crosses, relative scotoma.

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Recurrent anti-GQ1b IgG antibody syndrome showing different phenotypes in different periods

Anti-GQ1b IgG antibody is often found in the sera of patients with Miller Fisher syndrome, Bickerstaff's brain stem encephalitis, Guillain–Barré syndrome with ophthalmoplegia, acute ophthalmoplegia without ataxia, and occasionally, in isolated internal ophthalmoplegia and chronic ophthalmoplegia. These conditions may be designated as anti-GQ1b IgG antibody syndrome.1 We report a patient who showed three different phenotypes of the anti-GQ1b IgG antibody syndrome at different periods.

Case report

The patient was a 19 year old woman. At age 10, she visited a neurologist because of diplopia. There were clinical similarities between Miller Fisher syndrome and Bickerstaff's brain stem encephalitis, because she showed transient central nervous system involvement (drowsiness, respiratory disturbance, positive plantar responses, and EEG abnormalities) in addition to the triad of the Miller Fisher syndrome. High anti-GQ1b and anti-GT1a antibody titres decreased below the cut off level by day 55. The F wave on nerve conduction studies became normal by day 78. Fifteen weeks after the onset, she had almost recovered except for areflexia and slight restriction of ocular abduction of the both eyes.

Comment

Our patient showed three different conditions of the illness at three different periods between the ages 10 and 19: first, acute onset of ophthalmoplegia, ataxia, and areflexia at age 10, which is a typical presentation of the Miller Fisher syndrome; second, chronic progressive diplopia associated with internal ophthalmoplegia from age 17; and third, acute onset of complete ophthalmoplegia, ataxia, marked drowsiness, and respiratory paralysis with extensor plantar responses and EEG abnormalities at age 19.

We diagnosed the third episode as Bickerstaff's brain stem encephalitis, because she showed transient central nervous system involvement (drowsiness, respiratory disturbance, positive plantar responses, and EEG abnormalities) in addition to the triad of the Miller Fisher syndrome. High anti-GQ1b and anti-GT1a antibody titres at the time of the most recent illness and their decrease following recovery supported this diagnosis.

There are clinical similarities between Miller Fisher syndrome and Bickerstaff's brain stem encephalitis, because these conditions have been considered as consecutive spectra of the same disease. Miller Fisher syndrome is usually a monophasic illness, but, on rare occasions, it may occur after a long asymptomatic interval.1 It has been reported that clinical features of recurrent Miller Fisher syndrome are constant from episode to episode.2 This is in contrast with recurrent Guillain–Barré syndrome, which shows considerable variety in the distribution and severity of weakness between each episode.3 This is the first report of Bickerstaff's brain stem encephalitis as a recurrent episode of the Miller Fisher syndrome.

In the second phase of chronic progressive diplopia, our patient showed abnormalities of the pupils with slight ataxia and absence of the F wave in nerve conduction studies. As external oculo movement was not restricted, progressive diplopia might reflect pupillary abnormalities; diplopia has been discussed in isolated internal ophthalmoplegia without external ophthalmoplegia associated with anti-GQ1b IgG antibody.4 In addition, chronic external ophthalmoplegia has been found with raised serum anti-GQ1b IgG antibody.5 These findings suggest that chronic internal ophthalmoplegia may be associated with anti-GQ1b IgG antibody, although we could not examine this antibody during that period.

It is unique in our patient that three different phenotypes of the anti-GQ1b IgG antibody syndrome appeared at different times. There has up to now been no report in which different anti-GQ1b IgG antibody syndromes have recurred at different times in a single patient. Our case indicates that Miller Fisher syndrome, Bickerstaff's brain stem encephalitis, and chronic internal ophthalmoplegia form part of the spectrum of the anti-GQ1b IgG syndrome, although the mechanism of the variability in clinical phenotypes of the anti-GQ1b IgG syndrome remains unknown.

In conclusion, our case indicates that different phenotypes of the anti-GQ1b IgG antibody syndrome can occur at different times in the same patient, showing that this syndrome may be a distinct entity with a wide clinical spectrum on a unique immunological background.

References


Sydenham's chorea may be a risk factor for drug induced parkinsonism

Sydenham's chorea, the most common cause of acquired chorea in childhood, is a delayed complication of group A β-haemolytic streptococcal infection.7 It is thought to be caused by antibodies induced by streptococci which cross react with basal ganglia antigens.7 Despite the decrease in Sydenham's chorea in developed countries, there is a renewed interest in this condition because of the hypothesis that a similar mechanism may play a role in the pathogenesis of a subset of patients with tics and other neuropsychiatric disorders.7

The treatment of Sydenham's chorea is based on the combination of penicillin and anticholinergic drugs (valproic acid and/or dopamine antagonists). At the movement disorders clinic of the Federal University of Minas Gerais (MDC-UFGM), located in an area where Sydenham's chorea remains endemic, we have been struck by the occurrence of drug induced Parkinsonism among patients with Sydenham's chorea. We therefore
decided to investigate the incidence of this complication in a cohort of patients with the condition, as well as in a group of subjects with Tourette's syndrome. The latter is an interesting choice for a control group as Tourette's syndrome and Sydenham's chorea share clinical features—for example, childhood onset, the constellation of motor and behavioural disturbances, and the response to neuroleptic agents.

Methods
In the first part of the study we undertook a retrospective review of the case records of patients with Sydenham's chorea and Tourette's syndrome followed up at the MDC-UFMG from July 1993 to October 2002, looking for drug induced parkinsonism. We then compared the chlorpromazine equivalent dose used in patients with Tourette's syndrome, Sydenham's chorea, and drug induced parkinsonism. For each patient with Sydenham's chorea we randomly selected two age matched subjects with Tourette's syndrome. Sydenham's chorea was diagnosed according to a modified Jones criteria, and the Diagnostic and Statistical Manual of Mental Disorders, fourth edition (DSM-IV) criteria were used to identify patients with Tourette's syndrome. Drug induced parkinsonism was diagnosed when patients exposed to neuroleptics were found to have bradykinesia and at least one of the following: rigidity, tremor, postural instability. All patients were seen by one of us (FC). Differences were considered statistically significant at a probability (p) value of < 0.05.

Results
Sydenham's chorea was diagnosed in 91 patients and Tourette's syndrome in 97 during the study period. Five patients (5.5%) with Sydenham's chorea (mean (SD) age, 13.4 (2.1) years), of whom four were female, developed drug induced parkinsonism, whereas this complication was not identified in the Tourette's syndrome group (p = 0.03, Fisher's exact test). Parkinsonism was characterised by the presence of bradykinesia and rigidity. Of the subjects, terrors was identified in three, and postural instability was not observed. The mean cumulative chlorpromazine equivalent dose used in the patients with Sydenham's chorea when they developed parkinsonism was 16 518 (6254) mg. The onset of drug induced parkinsonism occurred after a mean exposure of 88.2 (65.8) days.

The cumulative chlorpromazine equivalent dose in the 10 Tourette's syndrome patients (two female; mean age 13.5 (1.1) years (p = 0.26 vs Sydenham's chorea)) during a three month period was 19 575 (6529) mg (p = 0.76 vs Sydenham's chorea). At the time of the onset of drug induced parkinsonism the mean chlorpromazine equivalent dose in the Sydenham's chorea patients was 176.6 (95.5) mg; in the Tourette's syndrome group the figure was 217.5 (220.0) mg (p = 0.05, paired t test).

Comment
We showed that 5.5% of our patients with Sydenham's chorea developed drug induced parkinsonism during treatment with neuroleptics, while this complication was not observed in a cohort of Tourette's syndrome patients of the same body weight. There are several possible explanations for this. First, the patients with Sydenham's chorea might have received a higher dose of neuroleptics. This hypothesis is ruled out by the finding that 10 randomly selected, age matched Tourette's syndrome patients and the five Sydenham's chorea patients with drug induced parkinsonism were treated with a similar cumulative chlorpromazine equivalent dose during the three month period when the latter developed parkinsonism. One may argue that although both groups received a similar cumulative dose of neuroleptics in the time between the start of treatment and the onset of drug induced parkinsonism, the patients with Sydenham's chorea could have been exposed to a higher dose of dopamine receptor blocking drugs at the time when they developed parkinsonism. However, this was not the case because if anything the mean dose of neuroleptic in the Sydenham's chorea subjects at the onset of drug induced parkinsonism was lower than in the Tourette's syndrome patients, though the difference failed to reach statistical significance. A second explanation for our findings is the concomitant use of valproic acid in three of our Sydenham's chorea patients, though the difference was not statistically significant. All patients were seen by one of us (FC). Differences were considered statistically significant at a probability (p) value of < 0.05.

References

Comparison of the tendon and plantar strike methods of eliciting the ankle reflex
Little work has evaluated the various ways of eliciting the ankle reflex. A previous study of elderly patients with normal/absent reflex found greater intraobserver and interobserver agreement with the plantar compared with the tendon strike method.1 Other studies showed that the reflex was best elicited in the knee position but moving comatose patients can be impossible or lengthy.1,2 We compared the reliability of the plantar and tendon strike methods of eliciting the ankle reflex in different disease states by examiners with different skill levels.

Four patients with pathologically brisk reflexes, five with reduced/absent reflexes, and the subjects with normal reflexes were screened from examiners so that only their legs were visible. None had identifying scars, wasting, or pes cavus. Subjects were examined by 15 third year medical students and five experienced neurologists. Initial training in both methods was given: in the tendon strike method the Achilles tendon of the supine patient was struck with the leg flexed at the knee and externally rotated at the hip, in the plantar technique the examiner's hand was struck while placed on the plantar aspect of a supine patient's foot. Reinforcement was permitted at examiners' discretion. Each examiner saw half of the subjects (that is, nine patients and controls) on four occasions. On each occasion they examined both ankles. Examiners evaluated the reflexes four times using each technique twice. The order of bed, subject, and method allocation to examiners was according to a randomised, partially balanced incomplete block design. Examiners rated the reflexes as normal, pathologically brisk, or reduced/absent and stated whether or not they were confident in their result. The study had local ethical approval.

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Table 1A shows the sensitivities for the first of examiners’ encounters with each subject/method. Sensitivity was high for the reduced/absent category for both experience levels, but low for normal or increased reflexes. The tendon method for students on brisk reflex patients was particularly inaccurate compared with plantar. This was not true for clinicians. Despite low sensitivity, examiners sometimes declared confidence in their incorrect classifications: 81% declared confidence when incorrectly classifying a brisk reflex using plantar compared with 63% using the tendon method. Intraobserver agreement for reduced reflexes was reasonable but lower for normal or brisk (table 1B). These were not always small misclassification errors: 23% of clinicians’ test pairs using tendon strike classified an increased reflex correctly once but as absent on the other test. In several instances the reflex was incorrectly classified on both tests. All 13 students who declared a preference preferred the plantar strike but no clinician stated a preference.

We found poor sensitivity and reproducibility for both techniques with normal and brisk reflexes for both examiner types. Results for reduced/absent reflexes may be inflated as most patients had absent reflexes which are easier to detect. Also they may have had other subtle lower motor neurone signs giving clues to examiners. The low sensitivities show that the ankle reflex should be interpreted in the light of other physical signs. Experienced clinicians had similar results with both techniques. The disparity preferred the plantar strike but no clinician stated a preference.

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Competing interests: none declared.

References

Mirror movements of the non-affected hand in hemiparkinsonian patients: a reflection of ipsilateral motor overactivity?

Mirror movements may result from a primary motor efferent system dysfunction with secondary motor reorganisation. A profound dysfunction of the motor pathways has been reported in Parkinson’s disease (PD) during execution of motor tasks.1–3 Recent PET studies have demonstrated overactivation of ipsilateral motor areas in hemiparkinsonian patients.1 However, the clinical expression of ipsilateral cortical activation was not specifically investigated in previous reports. In this study, we explored the presence of mirror movements (MM) during standardised unilateral hand tasks in a series of 21 hemiparkinsonian patients.

Patients were divided into two groups: de novo patients (n=11), age 53.2 (7.5) years (mean (SD)), duration of evolution 1.8 years (range: 1–5 years), UPDRS III motor score 12 (5.7), affected side: left n=5, right n=6; and treated patients (n=10), age 59.8 (7.6) years, duration of evolution 3.7 (1.8) years (range: 2–7 years), UPDRS III motor score 14 (7.5), affected side: left n=4, right n=6, mean daily dose of levodopa: 450 mg (range: 300–900 mg), improvement in motor disability >40% (range: 40%–80%). Evaluation was performed as follows: for de novo patients, before treatment; for the treated patients, in the “off” condition after at least 12 hours withdrawal of antiparkinsonian treatment (levodopa). Patients were not tested in the “on” condition, to avoid confusion between dyskinesia and MM. They were compared with 21 age matched normal subjects, age 56.4 (10.8) years.

Activation tasks

Subjects were told to hold their hands in the air with the elbows flexed and to perform a voluntary movement with one hand while the other hand was relaxed. Each hand was tested separately in the following four tasks performed 10 times as rapidly as possible with the widest amplitude: (1) wrist flexion/extension movements, (2) opening and closing of the hand, (3) finger tapping (thumb and index finger), (4) flexion-extension movements of the wrist. Tasks 1 to 3 correspond to items 23 to 25 of the UPDRS III, respectively.

Each task was scored as follows: 0=no MM, 1=MM (that is, the presence of repetitive unintentional contralateral movements that mimic totally or partially the intended movement. The “MM score” was the combined score for the task, for each side (maximum 4). Statistical analysis was performed using the non-parametric Spearman test, for a correlation analysis between the “MM score” and the UPDRS motor score.

In 80% of the de novo patients and 90% of the already treated and more severely affected patients, tested in the “off” condition, MM were observed in the relaxed hand while voluntary movements were being performed with the other hand. The most remarkable finding was that MM were never observed when voluntary movements were performed with the non-affected hand, whereas they were almost constant when voluntary movements were performed with the affected hand. They were observed both in the de novo group and the treated group. There were more often observed for alternate movements or repetitive flexion/extension movements of the wrist than for finger tapping. None of the control subjects displayed MM.

In the de novo patients, there was a significant correlation (r=0.60; p=0.0475) between the severity of motor impairment, as defined by the UPDRS III motor score, and the occurrence of MM as indicated by the “MM score”.

No such correlation existed in the treated group.

Mirror movements could reflect the higher than normal level of cerebral activation in response to complex movements reported both in normal subjects1,4 and in PD patients.1,6 However, none of the controls displayed MM and the four tasks were not complex as patients performed them without difficulty, albeit more slowly.

Table 1

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<tr>
<th>A</th>
<th>% Correct reduced/absent (95% CI)</th>
<th>% Correct normal (95% CI)</th>
<th>% Correct brisk (95% CI)</th>
<th>( \kappa ) (SE)</th>
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<tr>
<td>Students</td>
<td>Tendon strike</td>
<td>81.1 (64.8 to 92.0)</td>
<td>57.7 (45.4 to 69.4)</td>
<td>37.0 (19.4 to 57.6)</td>
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<td>89.2 (74.6 to 97.0)</td>
<td>54.9 (42.7 to 66.8)</td>
<td>70.4 (49.8 to 86.2)</td>
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<td>92.3 (64.0 to 99.8)</td>
<td>63.2 (38.4 to 83.7)</td>
<td>61.5 (31.6 to 86.1)</td>
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<td></td>
<td>Plantar strike</td>
<td>92.3 (64.0 to 99.8)</td>
<td>63.2 (38.4 to 83.7)</td>
<td>61.5 (31.6 to 86.1)</td>
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<table>
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<tr>
<th>B</th>
<th>Subjects reduced/absent</th>
<th>Subjects normal</th>
<th>Subjects brisk</th>
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<tr>
<td>Students</td>
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<td>86.5 (71.2 to 95.5)</td>
<td>67.6 (55.4 to 78.2)</td>
<td>66.7 (46.0 to 83.5)</td>
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<tr>
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<td>81.1 (64.8 to 92.0)</td>
<td>64.8 (52.5 to 75.8)</td>
<td>59.3 (38.8 to 77.6)</td>
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<td>Clinicians</td>
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<td>92.3 (64.0 to 99.8)</td>
<td>68.4 (43.4 to 87.4)</td>
<td>61.5 (31.6 to 86.1)</td>
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<td>Plantar 1st and 2nd</td>
<td>92.3 (64.0 to 99.8)</td>
<td>57.9 (33.5 to 79.7)</td>
<td>62.3 (40.6 to 89.8)</td>
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Alternatively, extended recruitment of cortical motor areas could reflect an overflow of commands into the contralateral hemisphere in unilateraly affected patients. In line with recent experimental results in a unilateral rodent model of Parkinson’s disease, we suggest that MM observed in the non-involved hand during movements of the akinetic hand reflect ipsilateral activation of the primary motor cortex. In the absence of sensorimotor activation contralateral to the affected akinetic (right) hand, the ipsilateral diffusion of activation may be considered as a compensatory mechanism. This ipsilateral activation could be explained in two different ways. Firstly, a corticocortical spread as the two hemispheres are connected via the corpus callosum and corticocortical pathways. Secondly, bilateral basal ganglia projections as several anatomical observations have shown that the basal ganglia are reciprocally and directly connected to the contralateral cortex. Thus, the activation of the ipsilateral motor cortex could result from the activation of one or both of these pathways. The precise role of the ipsilateral activation of the primary cortex in the pathophysiology of Parkinson’s disease is still unknown but it could be suggested that this phenomenon is a compensatory mechanism.

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