

Medial Frontal Cortex Infarction and the Alien Hand Sign

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• Two right-handed patients with infarction involving the left medial frontal cortex are described. Both patients exhibited a form of transcortical motor aphasia and a psychomotor disturbance involving the right arm characterized by forced grasping, motor perseveration, and the presence of apparently purposeful movements that appeared to be dissociated from conscious volition. The latter feature is noted to be identical to the alien hand sign as described in the left arm of commissurotomy patients and patients with callosal neoplasm or infarction. It is suggested that the observed dissociation of conscious intention from purposeful movement may be related to dysfunction of the medial frontal cortex contralateral to the impaired extremity as much as to hemispheric disconnection.

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The concept of the "alien hand" was introduced by Brion and Jedynak¹ based on their observations of three patients with tumors of the corpus callosum. In reviewing the syndrome following cerebral commissurotomy, Bogen described the alien hand sign as "a circumstance in which one of the patient's hands... behaves in a way which the patient finds 'foreign,' 'alien,' or at least uncooperative."²

Geschwind indicated that Kurt Goldstein "was perhaps the first to

stress the non-unity of the personality in patients with callosal section and its possible psychiatric effects."³ He then quoted Goldstein's description of a patient who complained "of a feeling of strangeness in relation to the movements of the left hand which she described with such curious expressions (she would say that someone was moving her hand and that she wasn't doing it herself) that she was regarded at first as a paranoiac."^{3,4}

In this article, we describe two patients who, in addition to other findings, exhibited the alien hand sign in the right arm in association with infarction involving the left medial frontal cortex as documented by radionuclide scan and computed tomography (CT).

REPORT OF CASES

CASE 1.—A 63-year-old, right-handed woman was first observed to have difficulty in speaking and weakness of the right leg. She appeared alert but did not initiate conversation. She spoke very haltingly in reply to questions. Her comprehension of speech was good, as was her ability to repeat complex phrases and sentences. She had a mild right facial weakness, a moderate paresis of the right arm, and a total paralysis of the right leg. A strong grasp reflex was noted in the right hand. Computed tomography demonstrated an area of decreased density in the left frontal medial cortex (Fig 1), and a subsequent isotope scan showed increased uptake in this same area (Fig 2).

Three weeks after onset, she still did not initiate conversation but rather remained silent until spoken to. Her verbal output was characterized by difficulty in initiating responses, struggle, and hesitation. In repetition of phrases, her speech was entirely intact. Her verbal comprehension was quite good. The movement in the arm

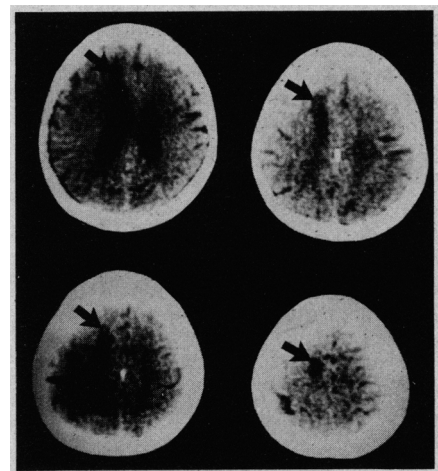
had recovered to the point where she had good strength and was not bound to synergies. The right leg remained impaired and she was not yet ambulatory.

The right arm was noted to display the following motor behavior:

1. There was a strong grasp reflex in the right hand.

2. The patient would reach out spontaneously with the right hand and then would not be able to release her grip voluntarily. She was unable consciously to inhibit this behavior although she was quite aware of it and was obviously frustrated by her inability to prevent it. She tended to restrain the movement of the right arm by holding it by her side with the more "obedient" left arm. At times, the right arm was noted to interfere with tasks being performed by the left arm. This "intermanual conflict"^{2,5} was observed at one point when it was noted that her right hand came up to keep her glasses on after she had begun to remove them with the left hand. She was aware of these conflicts but was, again,

Fig 1.—Case 1, computed tomograms demonstrate area of decreased density in left medial frontal cortex (arrows).



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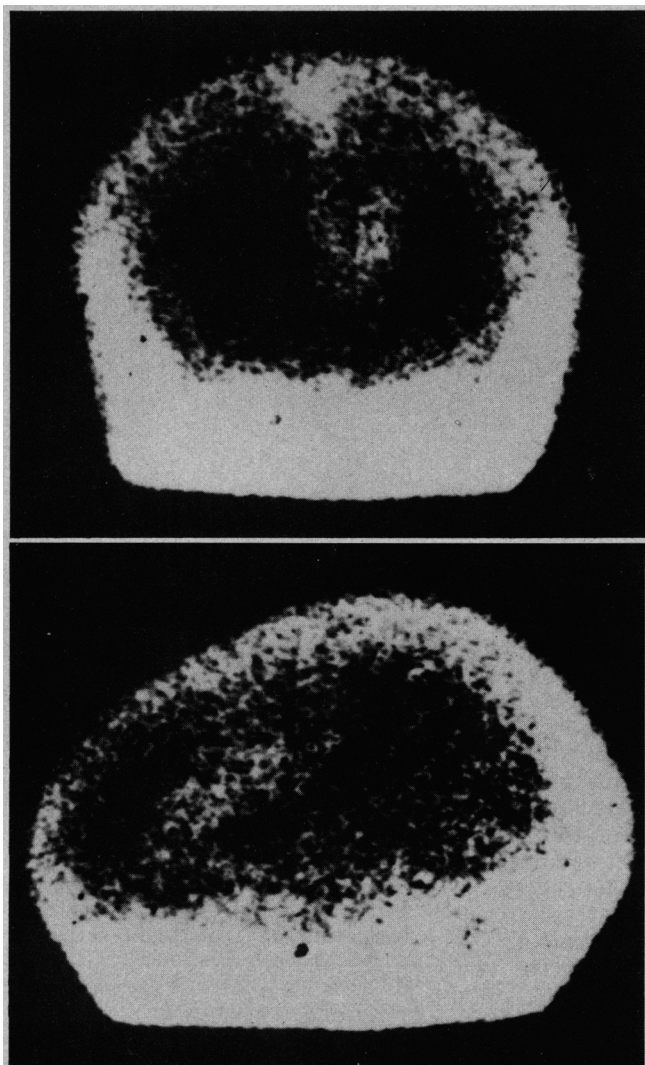


Fig 2.—Case 1, radionuclide brain scan showing area of increased uptake along posteromedial aspect of left frontal lobe. Above, anterior view; below, left lateral view.

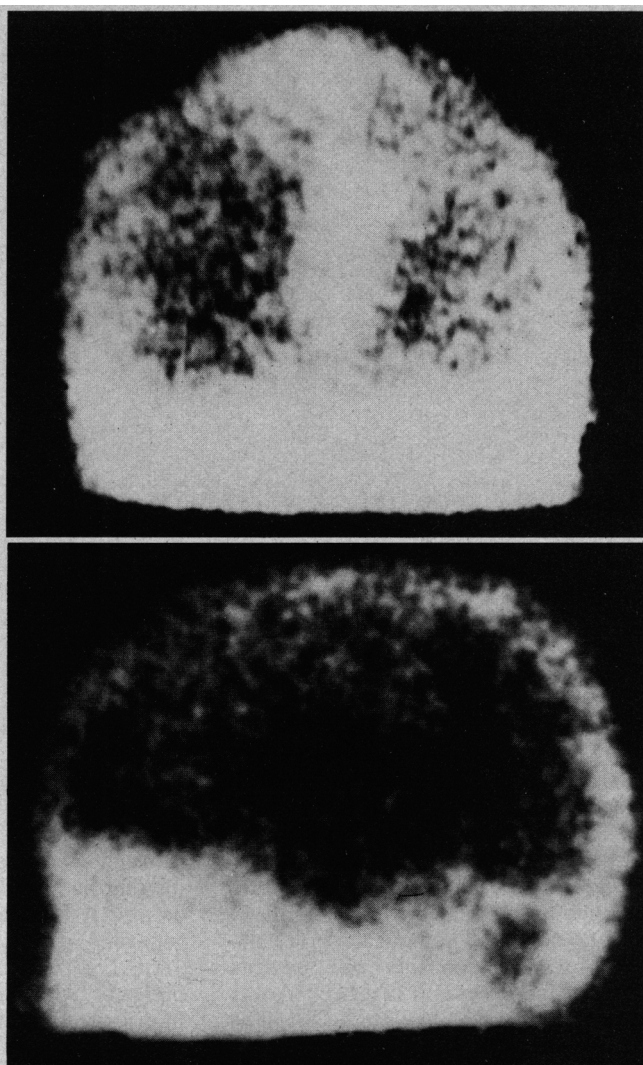


Fig 3.—Case 2, radionuclide brain scan demonstrates zone of increased uptake extending along medial aspect of left frontal lobe. Above, anterior view; below, left lateral view.

frustrated by being unable to prevent them consciously.

3. Motor perseveration was evident. The patient would begin to perform a task with her right arm but it would begin to perseverate uncontrollably. The left arm would then interrupt the perseveration by restraining the right arm and then complete the task. This was noted, for example, on several occasions when she was attempting to apply the brakes on her wheelchair.

Later, a slowly improving ability developed to inhibit to some extent the unwanted behavior of the right arm through concentrated effort. When these unwanted activities of the right arm began to interfere with performance of some task, they could be interrupted by the examiner through the issuance of a simple motor command, such as "place your right arm across your lap." The patient would then be able to obey this command and, in the process of doing so, override the aberrant motor behavior.

In the fifth week after onset, the patient

began to walk alone. The right arm would, however, tend spontaneously to reach out and grasp objects (eg, door knobs) that she passed, thus interfering greatly with her progression. This was minimized by having her repetitively tap on her thigh with the right hand as she walked or walk with a cane held in the right hand.

CASE 2.—A 76-year-old, right-handed woman was first observed after having collapsed with sudden muteness and right-sided weakness. A brain scan done one week following admission showed increased uptake in the left medial frontal cortex (Fig 3). A lesion in this area was confirmed by CT scan (Fig 4).

On examination three weeks after onset, she was fully alert and able to respond to questions with single-word replies although she did not initiate conversation spontaneously. Her responses were characterized by a great deal of difficulty in initiation. She could repeat complex phrases without difficulty. Her verbal comprehension was good. She tended compulsively

to repeat parts of questions asked of her before struggling to initiate a response and tended to whisper her reply. When she spoke on the telephone as opposed to face-to-face, her conversational speech was significantly improved.

A strong grasp reflex was noted in the right hand. Spontaneous goal-directed movements of the right arm were noted to occur that, judging by the patient's response to them, did not seem to be consciously intended. For example, at one point it was noted that the patient had picked up a pencil and had begun scribbling with the right hand. When her attention was directed to this activity, she reacted with dismay, immediately withdrew the pencil, and pulled the right hand to her side using the left hand. She then indicated that she had not herself initiated the original action of the right arm. She often reacted with dismay and frustration at her inability to prevent these unintended movements of the right arm. She experienced a feeling of dissociation from the actions of the right

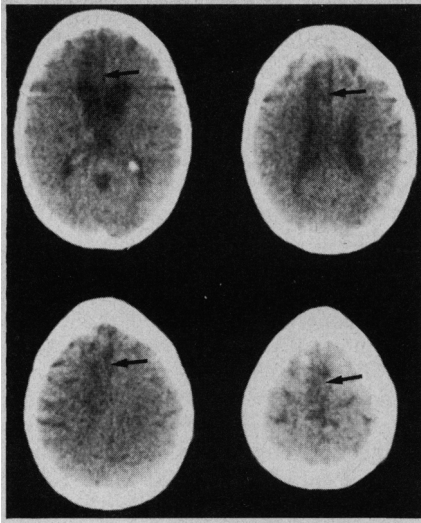


Fig 4.—Case 2, computed tomograms demonstrate area of decreased density involving left medial frontal cortex (arrows).

arm, stating on several occasions that "it will not do what I want it to do." She also displayed motor perseveration of the right arm when she attempted to use it. Often, she would attempt to restrain these unwanted actions of the right arm by holding it by her side with the left arm. The aberrant behavior of the right arm could be interrupted when the examiner issued a simple verbal command.

COMMENT

Our two patients both displayed a strong grasp reflex in the right hand. They were both noted to have motor perseveration, ie, inappropriate repetition of a simple movement of the right hand and arm usually triggered by an attempt at voluntary activity.

Both patients additionally exhibited a speech disorder consisting of a lack of spontaneous speech production with preserved ability to repeat. This pattern is often designated transcortical motor aphasia, although some authors have argued that it is not indeed an aphasia.⁶ Goldstein has attributed this form of speech impairment to either a "heightening of the speech threshold" or, more typically, an attenuation or loss of the "speech impulse."⁷ This type of speech disturbance has been described in patients who have sustained damage to the left medial frontal cortex.⁸⁻¹¹ Brown¹² has suggested that this form of "partial mutism" observed in such patients may be a disturbance intermediate between the total mutism associated with bilateral damage of the cingulate gyrus and the "selective speech loss" seen in left-sided neocortical damage of the classic motor areas for speech. He has further postulated that such a

speech disturbance would be expected to appear with damage to the "supplementary motor area and related paralimbic structures derived from, and possibly including, the cingulate portion of the limbic system."^{7,12}

Shahani et al presented the case of a right-handed patient who had suffered infarction of the medial frontal cortex due to occlusion of the left pericallosal artery.¹³ He had troublesome spontaneous grasping and motor perseveration in the right arm, of which he was fully aware but could not consciously inhibit. He tended to restrain these movements of the right arm using his left arm. The authors found, as we did in our patients, that the ability to execute a new movement with resulting inhibition of the aberrant movement was retained. They speculated that the association of the grasp reflex with motor perseveration was significant and that the problem common to both behaviors may be a deficiency in the mechanism that produces the voluntary inhibition of movement. The medial frontal cortex, they believed, was somehow involved with this mechanism.¹³

In our patients, apparently purposeful (ie, goal-directed) movements that appeared to be occurring independent of conscious volitional control were noted in the right arm. In patient 1, there was also some evidence of conflict between the motor behaviors of the two arms, with the right arm being the identified offender. Both patients exhibited the adaptive action of controlling the unwanted activity by restraining the right arm physically with the left arm.

Bogen has recently described several dissociative psychomotor phenomena observed in an "acute disconnection syndrome," a transient state seen in patients in the first few weeks following section of the corpus callosum for intractable epilepsy.² These include "intermanual conflict," the "alien hand sign" as defined previously, and "autocriticism," another phenomenon observed in both presented cases, in which the patient expresses astonishment and frustration frequently in reaction to "the capacity of the . . . hand to behave independently of conscious volition." Similar phenomena have been described by other investigators following commissurotomy⁵ and in association with midline tumors.¹ These dissociative phenomena can be distinguished from those classically associated with parietal lobe damage, such as autopagnosia, in that the latter seem to be related to impaired interpretation of body per-

ception, whereas the former would appear to be a misinterpretation of body action.

Are these behaviors due to damage to the corpus callosum alone and do they result from deficits in only inter-hemispheric transfer? Damage to the corpus callosum cannot be ruled out in the two cases reported. However, in the right-handed, split-brain patients, the alien hand sign was invariably observed with respect to the behavior of the *left* hand.^{2,5} Why, then, should the identical problem have been observed in the *right* hand of the right-handed patients with left medial frontal cortex infarctions? It is possible that these peculiar dissociative psychomotor behaviors result in part from damage to medial frontal cortex contralateral to the involved extremity. In the commissurotomy patients, their transient occurrence could have resulted from retraction of the medial surface of the nondominant *right* hemisphere intraoperatively in the course of exposing the corpus callosum for section.¹⁴

Two functional zones on the medial surface of the frontal lobes, damage to which may be implicated in the appearance of this psychomotor disturbance, as well as the speech disorder, are the supplementary motor area (SMA) and the cingulate cortex. These two areas are reciprocally interconnected.⁶ Electrical stimulation in conscious human subjects seems to elicit similar forms of behavior from both the SMA and anterior cingulate cortex.^{15,16} Penfield and Jasper reported that stimulation of the SMA produced vocalizations, speech arrest, complex coordinated or repetitive movements usually appearing in the contralateral arm, and arrest or slowing of voluntary action following which it was noted that the patient was "usually puzzled by his inability to execute" the action he had intended.¹⁵ Talairach et al found that continuous electrical stimulation of anterior cingulate cortex in epileptic patients produced complex, highly integrated forms of motor behavior that resulted from compounding of various simple "primitive" movements that appeared most often in the contralateral arm.¹⁶ Sanides¹⁷ has shown that, in the primate brain, the SMA is a paralimbic expansion zone of encephalized limbic (cingulate) cortex and that it is of cytoarchitectural form intermediate in type between limbic cortex and neocortex.

In primate studies, Mountcastle et al¹⁸ have attributed voluntary motor command functions to cells in posterior parietal cortex (area 7), but in a

more recent study, Robinson et al¹⁹ found that cells in this area were consistently activated by specific stimuli and that their activity was enhanced when a subsequent motor response occurred. Whether or not the enhancement of the neuronal activity and the associated overt motor response would appear following a sensory trigger was believed to depend on the coupling of a motivational-behavioral influence. The authors suggested that such an influence could arrive via afferents to posterior parietal cortex from the cingulate gyrus.^{20,21}

Damasio and Van Hoesen have described four patients with lesions involving the left SMA as localized by CT scan. These patients were noted to have initially exhibited a global akinesia that eventually became circumscribed to the right side of the body. The authors concluded that damage to the SMA decreased the "drive" to execute both spontaneous and volitional movement.⁶

Recently, regional cerebral blood

flow studies in man have demonstrated bilateral activation of the SMA in association with various complex movements of the extremities as well as with speech, and it has been hypothesized that the SMA functions in the "programming" of these motor acts, ie, the SMA was believed to be involved in the assembly and execution of time-ordered sequences of motor instructions in algorithmic form.²²⁻²⁴ On the basis of these regional cerebral blood flow studies as well as additional evidence in man and animals, Orgogozo and Larsen²⁵ have suggested that the SMA may act in a "supramotor" capacity in the cortical organization of voluntary movements. They postulated that

the SMA may contribute to the establishment of new motor programs and probably controls the execution of established subroutines according to external and internal inputs. Its anatomical connections and the available physiological data suggest that the SMA could be the place where external inputs and commands are matched with

internal needs and drives to enable the formulation of a strategy of voluntary movements.²⁴

The alien hand sign and the accompanying dissociative psychomotor disturbances, as well as the speech disorder associated with left medial frontal cortex damage, could be interpreted as faulty initiation, execution, and inhibition of preexisting essentially intact motor subroutines. These program controls may no longer be coupled normally to "external and internal inputs." The appearance of these dissociative phenomena may thus be related to damage to the SMA, its connecting fibers, or associated structures located on the medial frontal surface of the hemisphere contralateral to the involved extremity.

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