
MIRROR MOVEMENTS IN THE ALIEN HAND SYNDROME

Case Report¹

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The alien hand syndrome has been associated with different descriptions of abnormal motor behavior. We report on two patients with transient left hemiparesis who remained with limb apraxia and were particularly impaired by a variety of involuntary skilled movements of their apractic limb. After the report and a review of the literature, we discuss the presence of one type of abnormal movement observed in this syndrome. These movements resemble the mirror movements seen in the normal development of motor control and in some pathologic conditions later. Their appearance in the alien hand syndrome seems to demand both intrahemispheric and interhemispheric motor pathway damage.

KEY WORDS: Alien Hand, Mirror Movements

The alien hand syndrome has been described in different conditions, the most common of which has been callosal disconnection because of cerebrovascular accidents, tumors or surgical procedures. Initially described as an inability to recognize one's own hand,¹ it was later associated with the presence of abnormal movements of a complex and semipurposive nature. This abnormal motor behavior has been called diagnostic dyspraxia^{2,3} or intermanual conflict.⁴ It has been suggested as pathognomonic of callosal disconnection but its mechanism is unknown.

The two patients described here suffered a vascular disconnection syndrome. They presented with a variety of abnormal movements in the left hand, over which they expressed a complete lack of control. One type of abnormal movement that was noted had many characteristics of the mirror movements present during normal early development of motor control and in abnormal developmental conditions. A review of the literature suggests that this type of abnormal movement is not infrequent in callosal disconnection.^{2,3,5-15} The investigation of such movements in the alien hand syndrome may allow some insight into the nature of normal motor control and recovery from brain damage.

CASE REPORTS

Patient 1

A 50-yr-old, right-handed, female laboratory technician developed increasing difficulty in using

her left hand over a period of 24 h. Despite adequate strength, she could not perform requested movements with her left hand. Computerized brain tomography showed an infarct along the medial aspects of the right frontoparietal lobes corresponding to the border zone between the right anterior and middle cerebral arteries. Noninvasive carotid tests revealed a complete occlusion of the right internal carotid artery. Ten days later, on admission to a rehabilitation department, her main impairment was left-sided apraxia of both hand and leg. She could not name an object placed in her left hand or recognize with the left hand an object similar to one held with the right hand. This tactile anomia and lack of intermanual transfer of form discrimination suggested an interhemispheric disconnection syndrome as the underlying mechanism for the unilateral apraxia. The apractic behavior was worse on verbal command, but she did poorly also in the actual use of tools. She did much better in imitating gestures and pantomimes. Bimanual activities on command produced an involuntary immobilization of the left hand. Cutting food with a knife and a fork or tying shoelaces was impossible. She would try to guide her left hand with the right. During the following weeks she regained ability to perform daily bimanual activities but developed involuntary movements in her left hand. Three main types of involuntary movements of her left hand were observed. The first was interference with activities initiated by the right hand, which the left hand imitated in a mirror like pattern. For example, when turning the pages of a book in one direction with the right hand, she would turn them back with the left hand; opening a drawer with the right hand she would immediately close it with the left hand. The second type was interfering with willed bimanual activities by anticipating the role of the left hand. For example, while eating, the left hand with the

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fork, would "think ahead" and try to pick up a piece of meat before the right hand with the knife finished cutting it; before the right hand would finish taking a cigarette out of a box the left hand would close the box lid. The third type was independent activities of the left hand, unconnected to initiation of either right or bimanual activities. In this case when her left hand would "do what it wants," it would usually reach for nearby objects involuntarily; e.g., a pencil or a cigarette and sometimes make use of them. For example, unnecessarily manipulating a handle lever while driving; lifting a hot pot from the stove. These movements would occur more frequently when she was engaged in another task like reading or talking. Despite a general improvement in her apraxia tests, higher level limitations persisted. She was unable to follow three-step commands with her left hand, committed errors in pointing to body parts or spatial directions and was dysgraphic in her left hand. One year after her stroke her condition has not changed significantly. The involuntary movements of her left hand remain a major source of distress, disabling her from returning to work.

Patient 2

A 62-yr-old, right-handed retired policeman developed acute left hemiparesis. Computerized brain tomography showed multiple lacunas in both cerebral hemispheres including one in the body of the corpus callosum. On admission to a rehabilitation department 20 days after the stroke, the hemiparesis resolved except for mild weakness in the left leg and he could walk independently. Still, the function of the left hand was limited because of the limb apraxia extending also to the actual use of tools. The left hand also showed tactile anomia and loss of the ability to verbalize finger position without real loss of proprioception suggesting the presence of an interhemispheric disconnection syndrome. During his rehabilitation and later weeks at home, he developed involuntary movements of the left hand over which he lacked control. These consisted of the left hand "doing the opposite" of what the right hand would attempt to do. For example, if buttoning a shirt with the right hand the left hand would proceed to unbutton it and if pulling a cup of coffee toward him with the right hand the left hand would push it away. These behavioral abnormalities did not prevent him from finally getting the task done and they subsided gradually during the next few months.

DISCUSSION

The two patients described showed signs of unilateral left tactile anomia, agraphia, apraxia and lack of somesthetic transfer suggesting callosal disconnection.⁴ In addition they complained of uncontrollable abnormal motor behavior of the left hand. The combination of ego-alien motor behavior, in the

presence of callosal disconnection signs, characterizes our patients as suffering from the alien hand syndrome. The computerized brain tomographies did not localize the exact site of the responsible lesions. One case showed an extensive borderzone infarct between the right anterior and middle cerebral arteries territories and the other multiple lacunar lesions. However, based on previously reported cases^{5,6} with a similar clinical presentation, it seems that the critical lesions are situated along the medial border of the frontal lobe or on the anterior half of the corpus callosum. Thus, causing a disconnection of the neural pathways coming from the dominant hemisphere into the motor areas of the contralateral hemisphere. The first patient presented three types of abnormal movement of the left upper extremity: 1, imitative, mirror like, movements associated with purposeful activities initiated by the other hand; 2, anticipatory movements associated with intentional bimanual tasks; 3, prehensile or reaching movements, independent of the contralateral limbs, but sometimes associated with ongoing unrelated activities. The second patient showed mainly the first type of movements and to a milder degree also the third type.

The terms diagnostic dyspraxia^{2,3} and intermanual conflict⁴ have been applied nonspecifically to cover a variety of abnormal motor behavior present in the alien hand syndrome. The clinical presentation of our patients and a review of the case descriptions in the literature suggests that these heterogeneous movements can be classified into several categories. We would like to limit our discussion to those abnormal movements in which the affected hand imitated movements previously initiated by the contralateral limb. The imitation movements observed in our and other reported cases were at times symmetric and simultaneous causing competition between the hands, e.g., one hand reaching toward an object and the other reaching toward it or toward an identical nearby object.^{3,5,7-14} At other times the imitation was more complex causing intermanual conflict, e.g., one hand pulling an object closer, the other pushing it away,^{2,7,14,16} as for example, when our first patient would turn the pages of a book with her right hand and unturn them with the left hand or when our second patient would button his shirt with the right hand and unbutton it with the left hand. We suggest that there is an intriguing similarity between these movements and the category of movements known as identical associated movements (IAM), also called corresponding, homologous, imitative or mirror-associated movements.¹⁷ IAM were described over a hundred year ago.¹⁸ They appear in normal motor development during infancy and disappear toward puberty.¹⁹ They have been described in pathologic conditions, mostly unilateral cerebral damage and delayed motor development, but also in hereditary conditions and spinal disease.²⁰⁻²³ They occur only after a purposeful willed movement in the contralateral hand. After the observation that at the onset of motor development

motor control is bilateral, Westphal postulated that voluntary impulses from each hemisphere reach subcortical structures of both the same and through the corpus callosum, the opposite hemisphere. Later on, impulses to contralateral ganglia are inhibited so that ipsilateral hemispheric motor control is abolished.¹⁸ Modern studies reinforce the theory that a potential capacity for bilateral control exists in each hemisphere, but is normally suppressed in the adult. Cerebral blood flow studies have shown an increased blood flow in both supplementary motor areas in response to a movement intended only in one extremity.^{24,25} In animals, single cell recordings from the same areas, during a voluntary unilateral limb movement, have shown increased firing bilaterally.²⁶ In humans, electrophysiologic recordings show a readiness potential appearing bilaterally, 800 ms before an intentional unilateral act is performed.²⁷ Action potential recordings from muscles of hemiplegics and normal limbs reveal activity when the contralateral homologous muscles contract voluntarily.²⁸⁻³⁰ The supplementary motor area (SMA) has been proposed as a supramotor area^{24,31,32} in which motor programming takes place before its final execution by the ipsilateral primary motor area (PMA). Each SMA has anatomic connections to the contralateral SMA and to both ipsilateral and contralateral PMA and basal ganglia.³³ It could be argued that this supramotor area is also important in the primitive organization of bilateral movement and that the development of mutual inhibition between the secondary motor areas enabled the emergence of singular contralateral motor control. Damage to either side of this mutually inhibitory system would allow the reappearance of primitive bilateral control. In fact, ablation studies of the SMA in animals have produced mirror movements in the contralateral hand.³⁴

The imitative movements present in patients with the alien hand syndrome differ from IAM in few aspects. First, the alien hand syndrome is always associated with callosal disconnection signs. Second, the more severe interhemispheric disconnection occurring in the alien hand syndrome probably causes a wider release of abnormal motor control with more complex presentations (e.g., imitative reaching for a glass) than the IAM (e.g., imitative flexion of a finger). Third, in contrast to IAM^{17,18,23,29,30,35} the imitative movements in the alien hand syndrome seem to appear solely in the hand under the control of the damaged hemisphere, when it shows apraxia or weakness.^{1,5,7-9,13,14,36,37} This would suggest two necessary conditions for the occurrence of these movements: impaired voluntary motor control of one hemisphere over the contralateral hand, because of the intrinsic damage to its motor areas or disconnection from the motor-dominant hemisphere, presenting as paresis or apraxia and reduced inhibition of the primitive bimanual control of the dominant hemisphere.

These speculations about the mechanism subserving mirror movements in the alien hand syndrome

do not serve to explain all the abnormal movements observed in this condition. Some patients show a marked tendency to reach and manipulate objects that is not related to intentional movements in the contralateral hand. It presented as a separate type of abnormality also in our patients. The imitative tendency of these movements, when present, is guided by externally originating stimuli. Its mechanism is probably based on intrahemispheric pathophysiology³⁸ because it can be observed also in patients without signs of callosal disconnection.³⁹ Still, recovery from these abnormal movements, as well as results of cerebral blood flow studies after unilateral hemispheric stroke,⁴⁰ also supports the role of the contralateral hemisphere in regaining volitional control over the ipsilateral hand. As yet the treatment of this handicap is deficient. Medications do not affect the different types of movements. Holding a cane or some object in the problematic hand diminishes its tendency to reach involuntarily for other objects.

It seems appropriate to suggest that each type of abnormal movement be investigated separately, rather than the whole syndrome as one entity. Establishing the reproducibility of each class of abnormal movement and investigating its characteristics may provide valuable insight into the motor control reorganization after unilateral cerebral damage.

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