Diagnostic Apraxia and Ictal Alien Hand

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ABSTRACT

Alien hand is a strangely behaving hand and occurs in disease of medial frontal lobe and also corpus callosum. When it is with frontal lobe disease, it manifests as groping and hooking with either hand, but callosal lesions produce left-hand abnormalities of varying degrees. Occasionally this phenomenon can be transient as an ictal manifestation. Case 1 is a 20-year-old female who was on treatment for panic and anxiety for 4 years and later noticed strange action by the left hand pushing food plate and notebook when she is using them with the right hand. Case 2 is a 14-year-old boy who presented with episodes of strange situation where his left hand was pushing his right hand into his mouth, and he had no control over it. He bit his right-hand fingers, and on one episode, he was bleeding from his injured fingers, yet he felt someone is making his left hand do this. Both patients had a demonstrable structural lesion. Patients, when they have stereotyped strange behavior with limbs, need to be investigated for organic disease and initiate treatment accordingly.

Key words: Callosal lesion, diagnostic apraxia, ictal alien hand, parietotemporal region, psychosis

INTRODUCTION

How movements are controlled and coordinated is hypothesized based on functional magnetic resonance imaging (fMRI) based information as follows. Normal voluntary movements follow a sequence. Each hemisphere controls the opposite side limbs. Supplementary motor area and anterior medial frontal cortex starts the schema and transmits to motor area which transmits the information to the corticospinal tracts. Simultaneously, an afferent copy of the activity generated is received by the posterior parietal lobe for constant self-awareness and interact with the efferent copy transmitted by motor cortex. Failure of this mechanism will cause misinterpretation of self-generated movements as externally guided.[1] This defective matching of the efferent copy signal will cause loss of agency for the movement created.[2] Direct activation of motor cortex is seen in alien movements and the sequential activation is seen in nonalien movements. When the efferent copy is not produced by supplementary motor area, it is mismatched with the reafference and hence is misinterpreted as produced by external force. In disconnections of the movement coordinating circuit, different parts of the body produce movements which are not consciously recognized as produced by one’s own brain. Thus, there is dissociation between...
the movement generated and the awareness of the movement generated.[5] Therefore, for normal control, continuous congruence between the efferent copy and afferent input is needed. Passive limb movements which are not internally generated produce a parallel copy which if not matched with the efferent copy signal and is interpreted as external – exafference. The sensory motor links between the self-generated movements and the passive movements if not properly understood, the sense of agency for movement is lost. The possibility of a premotor center converting intentions to action is also being postulated based on fMRI studies.[6] Damage to this mechanism can cause disinhibition of exploratory mechanisms and autonomy. There is also inhibitory control, by this fronto-parieto-occipital pathway over actions which withdraw from external stimuli, and possibly there is balance between the “intent to capture” (grasp) and “intent to escape” (avoidance) systems.[5] This mechanism is called as positive cortical tropism. When parietal lobe is dysfunctional, negative cortical tropism sets in where a stimulus on the limb inhibits movement toward the stimuli causing avoidance and levitation. However, each interhemispheric control units can activate activities by unitary control mechanisms when the language-based control of the dominant hemisphere over the actions of both hemispheres is lost. This can result in each hemisphere activating its corresponding limb with opposing purposes.

Clinical syndromes
Frontal alien hand occurs both in dominant and nondominant hand with frontal release signs such as grasping, groping, and compulsive manipulation of environment. They are unable to suppress this, though the body part is recognized well. There are generally no neglects or apraxias. This can be seen with lesions of anterior corpus callosum, supplementary motor area, medial prefrontal cortex, and anterior cingulum.[6] Callosal alien hand affects the nondominant limb only with features suggestive of intermanual conflict with no frontal release signs but shows features of apraxia, agraphia, neglects to varying degree.[7-9]

Diagnostic apraxia
This syndrome is defined as an abnormal motor activity of the left hand activated by voluntary movement of the right hand. Commissural fibers from superior parietal lobule pass through part of the body of corpus callosum. Superior parietal lobule is concerned with selection of movement based on integration of visual and somatosensory information. Disconnection of right superior parietal lobule from left results in problems with volitional control of movements of the left hand.[10] Left hand can show unilateral ideomotor apraxia, agraphia, tactile dysnomia, left ear extinction on dichotic testing, etc.[11] It might do antagonistic movements like when the right hand wants to turn to next page of a book left might close it. It can also cause nonantagonistic irrelevant movements. Occasionally, inability to move during bimanual task can be seen. Some interesting report of both hand cooperating in bimanual task but the left starting to undo once it is over and going on and on that way.[11] Agnostic dyspraxia is the term used for compulsive automatic execution of movement by one hand when the patient is asked to perform with other hand. Whereas the conflict is between the desired act in which the unaffected hand is engaged and affected hand opposes it.[12] The intermittent alien hand is described with both frontal, callosal and parietal lesions as a ictal phenomenon.[13,14]

Treatment
Patients with demyelinating or vascular lesion improve over time. Meanwhile engaging the aberrant limb in activity will reduce the aberrant behavior. Patient can be told to carry a cane even if he does not need it. The limb can be mechanically restrained keeping it between the legs, or cover it so that afferent inputs are blocked.

CASE REPORTS

Case report 1
Our patient is a 20-year-old college student who experienced panic for nearly 4 years with no obvious reason. She had a feeling of losing control over her activities but could not explain what exactly was happening. Her memory, language and other activities of daily living were normal. She was on regular treatment from psychiatrist but continued to be symptomatic. Six months before, she visited our institution she often dropped her book, food plate, etc. She was not found to have any involuntary movements, incoordination, or weakness of limbs. However, she expressed panic that something is happening with in her. Her sleep was not disturbed and did not have weight loss. Her performance at college was preserved but for reported jitteriness and dropping. Her handwriting remained normal. Then, which was surprised to find that she was dropping objects not due to her jitters, but the left hand was playing mischief slowly pushing her book held in the right hand, and though she could observe that, she could not stop that hand. There was no weakness or apraxias found during the examination. However, the patient had difficulty in writing with the left hand when told to attempt and had clumsiness in bimanual acts. She also noticed that if she is not concentrating, her left hand slowly pushes her food plate out of the table [Videos 1-2]. The family strongly concluded the role of evil spirit and no response was seen to treatment based on that. She did not have any problem in using her left hand for routine activities, and she was unable to understand why her left hand was behaving like this. In the test situation, it was observed that when
she was asked to keep her both hands outstretched her left hand was slowly moving to words the right, and she had no explanation why this was occurring. Her hand also showed some noncompetitive meaningless movements with both hands, left more than right. Then, patient learned a self-restraining technique of sitting on her left hand while eating or reading. Investigations at our institute revealed slightly anxious person. However, neuropsychological assessment was normal. Her magnetic resonance imaging (MRI) showed a circumscribed lesion in the right posterior temporoparietal region. There was no edema or contrast enhancement. The possibility of a benign lesion like old healed granuloma was considered [Figure 1]. Biopsy and excision were advised, but patient’s relatives were unwilling. Patient uses the self-restraint techniques.

Case report 2
A 14-year-old boy studying in 10th standard woke from sleep screaming that his right hand was being pushed into his mouth by someone. He appeared confused and parents concluded it as dream. He was otherwise normal. He slept and ate more than his siblings and was obese. Few days later he again woke up from sleep screaming and his right hand was being pushed into his mouth bitten and bleeding and strangely his family noticed that his left hand was pushing his right hand into his mouth though it was injured and bleeding. The patient was unable to answer why his left hand is behaving so and he could not withdraw that. These episodes recurred in varying intervals with varying severity but he did not show any features of alien hand otherwise. Electroencephalogram showed a right parietal epileptic focus [Figure 2] and MRI showed an area of gliosis in the right posterior parietooccipital region and minimal gliosis on the left side [Figure 3]. The patients attack stopped with antiepileptic medication suggesting an ictal alien hand syndrome. Ictal Video electroencephalography (VEEG) could not be done in this patient.

DISCUSSION
Control of hand movements is a well-coordinated and planned activity of the brain in the distributed networks of the brain which are well monitored constantly. When these links break each part of brain becomes automatic and loses agency making the person feel as if another person is operating with in himself. These phenomenons have to be recognized as mostly they are misdiagnosed as mental illness.

CONCLUSION
The motor control and locus of its control remains a function of the fronto-parieto-occipital circuit. When broken it results in bizarre movements by the left hand, which for an uninformed person cannot be differentiated as organic. Therefore, these cases are
being recorded to bring about awareness into these phenomena.

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Conflicts of interest
There are no conflicts of interest.

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