Paroxysmal alien hand syndrome

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Abstract

Four patients are described who presented with a paroxysmal form of the alien hand syndrome. Two patients with damage to one frontomedial cortex had brief episodes of abnormal motor behaviour of the contralateral arm that featured groping, grasping, and apparently purposeful but perseverative movements, which both patients interpreted as alien or foreign. The other two patients, with posterior parietal damage, reported a paroxysmal feeling of unawareness of the location of the contralateral arm, lack of recognition of the arm as their own, purposeless movements, and personification of the arm. These cases represent a new form of the alien hand syndrome manifested by brief, paroxysmal episodes, which may be due to ictal mechanisms.

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The alien hand sign or syndrome is a psychomotor disorder characterised by a dissociation between intention and action. One of the patient's hands performs apparently purposeful movements, which are independent of volitional control, as if the hand is driven by an external agent.¹⁻⁹ The symptoms vary according to location of the lesion. In patients with either surgical or spontaneous lesions restricted to the corpus callosum, the hand ipsilateral to the language and motor dominant hemisphere acts in a manner opposite to what the patient voluntarily does with the dominant hand, a phenomenon called diagnostic dyspraxia by Akelaitis¹ and intermanual conflict by Bogen.³ This abnormal motor behaviour is always triggered by volitional movements of the normal hand.^{5,8,10} When a medial callosal lesion affects adjacent parietotemporal regions, the non-dominant arm may not be recognised as the patient's own when held by the contralateral arm behind the back in the absence of sensory loss⁷ or, alternatively, the hand "behaves in a way which the patient finds 'foreign', 'alien' or at least uncooperative".³ Patients with medial frontal lobe damage affecting the supplementary motor area and the cingulate gyrus may show a seemingly purposeful and uncontrollable tendency of the hand contralateral to the lesion to grope and grasp objects, sometimes accompanied by motor perseveration or compulsive manipulation of tools placed before them.^{4,6,9} Among patients

with an infarction in the territory of the right posterior cerebral artery spontaneous arm movements such as those related to gestures or postural adjustment, are felt as out of control due to opticosensory ataxia and as alien due to the severe sensory loss.¹¹ The alien hand syndrome in patients with unilateral hemispheric lesions, unlike those with callosal damage, is always present in the hand contralateral to the lesion, regardless of the pattern of cerebral speech dominance and not necessarily triggered by volitional movements of the normal hand.^{4,6-9}

The alien hand syndrome may last for days or weeks but has not been described as a paroxysmal manifestation. We report four patients who showed a paroxysmal alien hand syndrome probably caused by seizures originating in the contralateral frontomedial cortex (two patients) and in the contralateral posterior parietal region (two patients).

Case reports

Case 1

A 65 year old right handed man reported the following: "While I was travelling on a bus I noticed that a hand was approaching me on the right from behind, trying to catch me. After grasping my trouser leg, the hand did not release it. First, I thought somebody was assaulting me, but then I realised that it was my own right hand, although I did not feel it belonged to me. Thereafter, the fingers developed creeping movements, and repetitive jerks involving the whole arm soon followed. I was unable to control my right hand and I had to grasp and hold it with my left hand. My right arm felt heavy and awkward. I was very anguished, anxious and frightened, and had palpitations. . . ." The episode lasted for a few minutes, and recurred briefly that evening at the patient's home: "I couldn't sleep at all that night because I was terrified my right hand would assault me while sleeping." Neurological, neuropsychiatric, and general examinations yielded normal results. Laboratory analyses (haemoglobin concentration, white 7 blood cell count, erythrocyte sedimentation rate, blood glucose, blood urea, plasma electrolytes, calcium ions, Venereal Disease Research Laboratory test, lipids, liver function test, protein electrophoresis, coagulation studies, T3 and T4, urinalysis, electrocardiography, doppler ultrasound of neck and intracranial vessels, echocardiogram, and interictal electroencephalography) also yielded normal results.

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Figure 1 CT scan showing focal atrophy of left medial frontal cortex.

Computed tomography showed focal atrophy restricted to the left medial frontal cortex (fig 1). The patient was prescribed carbamazepine (800 mg a day), and the episodes did not recur during the next two years.

Case 2

This 54 year old right handed man experienced two transient episodes of seemingly purposeful movements of his left arm which he interpreted as foreign ("the arm acted independently of my own will, certainly it was not mine, the arm was driven by somebody else, it felt quite superfluous"). The first episode took place while he was driving. The patient was forced to stop the car because his left arm had an uncontrollable tendency to grasp and pull the steering wheel in a chaotic way. The second episode took place a week later while he was undressing. The left limb grasped and pulled up his trousers in a perseverative way, and he had to sit down until this involuntary behaviour stopped. He was very distressed and frightened and had profuse sweating. On examination, the patient was

alert, oriented, and cooperative. Cranial nerves and motor, sensory, and cerebellar functions were all normal. Interictal electroencephalography yielded normal results. CT showed a right parasagittal intra-axial mass just in front of the motor strip (fig 2). Surgery was performed, and a diffuse lymphocytic lymphoma was removed. The post operative period was uneventful with a mild residual weakness in the arms that disappeared after a few weeks. He was treated with diphenylhydantoin and had no symptoms during the next three years.

Case 3

This 34 year old right handed man had a left parietal arteriovenous malformation surgically removed in March 1990. He was left with a right hemiplegia and cortical sensory loss from which he gradually recovered. One year later he had a focal seizure with secondary generalisation. He said: "I suddenly lost all notion of where my right arm was and did not know whether it was in front or behind my body. The limb did not seem to be my own even after looking at it. I had the feeling my right arm was moving spontaneously in a purposeless way but I was not certain if it was so." This abnormal feeling lasted for only a few seconds and was followed by jerking movements of the arm and a generalised tonic-clonic seizure. Although seizures recurred twice, he experienced the feeling his right arm not belonging to him several times, and on one occasion he shouted at the affected limb: "What are you doing there, are you crazy?" On examination the patient was alert and oriented and had normal language, praxis and gnosis. Cranial nerves were normal. Reflexes were brisk on the right side with an equivocal extensor plantar response. Tone and strength were normal. Two point discrimination and graphesthesia were mildly impaired on the right hand, but the remaining sensory modalities were normal. There was neither neglect nor optical ataxia. Identification of body parts, tactile localisation and oral calculation were normal. Interictal electroencephalography yielded normal results. CT showed a post surgical lesion in the left posterior parietal region without evidence of arteriovenous malformation (fig 3). The patient was started on diphenylhydantoin with good control of the seizures, although he still experienced isolated and brief episodes of the alien feeling affecting the right hand.

Case 4

A 50 year old right handed woman had a sudden onset of headache and left hemiparesis in November 1989. CT and angiography disclosed an intracerebral right parietal haematoma secondary to a ruptured arteriovenous malformation. The haematoma was evacuated and the malformation surgically removed. She had a gradual and full recovery. Eighteen months later she experienced two transient episodes lasting a few minutes each. She said: "Suddenly I had a strange feeling

Figure 2 CT scan showing enhanced right frontal parasagittal lesion mainly affecting supplementary motor area.

Figure 3 CT scan showing postsurgical lesion in left posterior parietal area.

on my left side; later I could not recognise the left arm as my own; I felt it belonged to someone else and wanted to hurt me because it moved towards me; I saw it quite big and distorted like a monster; I was terrified." During the third episode which was witnessed by one of the authors she said: "Look, it's coming; please help me." The left arm elevated in extension from the horizontal plane, and the forearm flexed on the arm, followed by distal arrhythmic isolated clonic jerks while approaching the patient's face. During the episode the patient repeated in a panic-like state: "Please stop this monster; it will kill me; I also see your face is distorted and big; what is going on?" While the episode lasted for less than 3 minutes, it soon recurred with the same features but was promptly stopped with 10 mg intravenous diazepam. Since then, the patient has received phenytoin 300 mg daily with no further episodes. Neurological examination yielded normal results. Interictal electroencephalography showed focal slowing over the right parieto-

Figure 4 CT scan showing resection of an AVM which included part of superior temporal and inferior parietal gyri.

temporal region. CT showed the resection of an arteriovenous malformation which included part of the superior temporal and inferior parietal gyri (fig 4).

Discussion

We examined four patients who experienced paroxysmal episodes of the alien hand phenomenon. Both patients with lesions in the medial frontal cortex (mainly affecting the supplementary motor area and the cingulate gyrus) had brief episodes of abnormal motor behaviour of the contralateral arm characterised by groping, grasping, and apparently purposeful but perseverative movements, accompanied by the feeling of estrangement of the affected arm and hand, and a panic-like sensation. Those patients with posterior parietal lesions had shorter lasting episodes which consisted of unawareness of arm location, lack of recognition of the affected arm as their own, feeling of purposeless movements (doubtfully accompanied by actual motion), or true simple movements and personification of the arm.

In the first two patients the seemingly purposeful paroxysmal movement originating in the medial frontal region would seem to have been misinterpreted by the patient, hence its alien character,⁴ while the parietal lesion in the other two patients may have directly caused a disturbed body perception with the feeling of estrangement or non belonging with the illusion of movement in one and actual movements in the other. Possibly this is not an alien hand syndrome as it is not a true dissociated psychomotor phenomenon. The clinical manifestations in both patients, however, fit the concept of the alien hand syndrome as originally proposed by Brion and Jedynak.² On the other hand, the clinical description of the paroxysmal phenomenon provided by both patients with frontal lesions are compatible with the alien hand syndrome definition proposed by Bogen.³

Before further discussion, some limitations of our study should be pointed out. We could not obtain electroencephalograms during the seemingly ictal phenomena of the alien hand syndrome, and procedures performed interictally failed to show seizure foci. Several facts, however, support our hypothesis of ictal alien hand syndrome. Firstly, all the episodes were very brief (up to several minutes) and were not present on a daily basis as in the classic alien hand syndrome. Secondly, the paroxysmal alien hand syndrome in patients 1 and 3 was immediately followed by focal motor and generalised tonic-clonic seizures respectively. Thirdly, in patient 4 one of the episodes was arrested by intravenous diazepam; and lastly, the episodes completely resolved in three patients and decreased in frequency in the other after they were started on antiepileptic drugs. Other causes of paroxysmal phenomena such as transient ischaemic attacks have not been reported to produce an alien hand syndrome, and none of our patients had risk factors or clinical or laboratory evidence of

ischaemic cerebrovascular disease.

When the alien hand syndrome is the result of a callosal lesion the abnormal motor behaviour is dissociated from conscious volition due to interhemispheric disconnection. The fact that our two patients with medial frontal lobe damage described their paroxysmal motor behaviour as alien or foreign is attributable to dysfunction of the supplementary motor area. This area is involved in the control and execution of internally generated motor sequences, and an epileptogenic focus may release such learned motor engrams from both volitional control and intention.¹²

Electrical stimulation of the supplementary motor area and cingulate gyrus in humans may elicit motor behaviours that resemble the alien hand syndrome, such as simple, repetitive, or complex coordinated movements of the contralateral arm (in the case of supplementary motor area stimulations)¹³ and contralateral continuous, stereotyped, single, and primitive finger and hand movements (for example, touching, rubbing, pinching) that are often part of more complex movements adapted to the situation (in the case of cingulate gyrus stimulations).¹⁴ During stimulation of the supplementary motor area subjects may feel puzzled by their inability to execute the action they had intended to perform. After cingulate gyrus stimulations patients may experience the behaviour as somewhat foreign to themselves or attempt to explain it away as a reaction to the stimulus. In these subjects as well as in patients with seizures arising from the medial frontal region¹⁵ the elicited or ictal motor behaviour may be modified by external stimuli (for example, if a lit cigarette is offered during stimulation the patient may smoke it). Patients with an alien hand syndrome after medial frontal cortex damage may show similar symptoms such as a compulsive manipulation of tools placed before them.^{4,9}

Seizures originating in the medial frontal lobe (including the supplementary motor area and the cingulate gyrus) may feature tonic or clonic movements of one or more limbs, writhing and twisting body movements, contraversive movements, autonomic manifestations, disturbances of normal motor behaviour, and simple or complex motor automatism without loss of consciousness. The motor automatism may include vocal-respiratory or axial movements and manual or pedal activities such as rubbing and clapping hands, hugging oneself with both hands, palpating objects or striking the leg of an observer.¹⁵⁻¹⁸ Although we could not find previous reports of paroxysmal alien hand syndrome, Geir *et al*, described a patient (case 1) with automatisms of the right hand that were adapted to the situation (during a meal the patient stuck the fork repetitively into the bread)¹⁶ and case 3 of Waterman *et al* showed repetitive screaming and both hands going around the neck with tearing of the shirt as the clinical manifestation of a forced ictal frontal lobe automatism.¹⁵ Similarly, the patient reported by Banks *et al* with frontal

and callosal damage grasped his throat with his alien left hand.¹⁹

Patients with posterior parietal damage may experience a feeling of absence or strangeness of the contralateral arm, may personify the affected arm,²⁰ and may show the levitation phenomenon (that is an abnormal posture assumed by the arm when the attention is distracted from it).²¹ Epileptogenic lesions in the posterior parietal region and interparietal sulcus may produce a variety of paroxysmal asomatognosic disorders such as feeling of absence of a body part, a sense of foreignness and illusions of corporeal displacement, and a sense of movement without actual motion.²² Limb, axial, and eye movements may be evoked by epileptic activity or electrical stimulation within areas 5 and 7.^{13, 23-26} Motor responses produced by stimulation within area 5 seem to be components of complex and coordinated movements such as turning of the head and shoulder towards the contralateral side, part of reaching or withdrawal movements of the contralateral arm accompanied by finger flexion or extension, or transient assumption of a particular limb posture. Similar movement patterns have been observed in patients with epileptic foci within the parietal lobe.^{13, 25} Stimulation within area 7, near the intraparietal sulcus, has been reported to evoke eye movements and head turning to the contralateral side, as well as movements of the contralateral arm.^{23, 26}

Underlying these phenomena is the fact that neurons in both the superior and inferior parietal lobule are related to active movements of the hand and arm as they discharge during the active reaching of objects of motivational interest and during active manipulation of these objects.²⁷ Furthermore, neurons in the inferior parietal lobule mediate the control of eye movements, visual space perception, and visual attention and play an important part in both the generation and initiation of neural commands for movement directed towards the extrapersonal space. A complex type of joint and skin neurons in the superior parietal lobule may be related to the integration of body image.²⁷⁻³¹

The inferior parietal lobule receives cortical afferents from primary and secondary sensory motor areas and has reciprocal cortical connections with premotor and prefrontal cortex, area 5, superior temporal sulcus, cingulate gyrus, area 46, and visual association areas. Moreover, it receives relatively direct somatosensory, visual, and oculomotor inputs from subcortical structures and projects subcortically to motor pathways through the basal ganglia, superior colliculus, and pretectum, the pontine nuclei, and the vestibular nuclei.^{28, 32, 33} Thus, the inferior parietal lobule may be considered a multimodal association area contributing to the origin of a secondary, parallel motor pathway. According to Jeannerod the "inferior parietal lobule may belong to a widely distributed system including other subsystems such as area 46, the superior temporal sulcus and the cingulate gyrus, characterized by a supramodal status, a

high degree of associativeness and a common converging output for the control of motor behaviour."³⁴ Disruption of this system produced by seizures arising in the posterior parietal cortex may underlie the alien hand like syndrome of our cases 3 and 4.

In conclusion, we report four patients who showed an alien hand syndrome most probably as an ictal manifestation of seizures originating in either the frontomedial or the posterior parietal cortex. The ictal alien hand syndrome should be considered as another behavioural manifestation of epileptogenic activity in cortical association areas involved in the formulation of motor programmes or the integration of perceptual schemas into goal-directed motor plans.

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