

NEUROLOGICAL SIGN

The alien limb

Keith A. Josephs* and Martin N. Rossor†

*Divisions of Behavioural Neurology & Movement Disorders, Mayo Clinic, Rochester, MN, USA;

†Dementia Research Group, Department of Clinical Neurology, Institute of Neurology, Queen Square, London, UK, E-mail: M.Rossor@dementia.ion.ucl.ac.uk

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The term alien hand syndrome appeared in the literature over three decades ago, although the first description was probably recorded almost a century ago (Fisher 2000). The term was originally used to describe the patient's inability to differentiate their own left hand from the examiner's left hand when placed in their right hand, behind their back. However the term has evolved, and the terminology is confusing. Alien hand syndrome, alien limb syndrome and alien limb phenomenon are synonymous and can occur in either the arms or legs. The syndrome can be broadly divided into two types, an anterior syndrome and a posterior syndrome.

THE ANTERIOR ALIEN HAND SYNDROME

This is the classic syndrome. It is a motor disorder defined by involuntary hand or arm movements that occur either in addition to or instead of a planned or willed movement. The movements are complex and do not look like a recognizable movement disorder such as chorea, ballismus, dystonia, ataxia or myoclonus. The patient is usually aware of the arm movements but is unable to control them. They are described as feeling foreign or alien and on occasions the feeling is so profound that the arm is ascribed its own identity. The patient may even refer to the arm as 'my little friend' or 'it'. The left arm is more commonly the 'alien' one, but the right arm or rarely both arms may be affected depending on the site of the lesion. The complex movements consist of a combination of dyspraxic motor and frontal lobe reflex phenomena (see below).

The anterior alien hand syndrome has been further divided into a callosal type and a frontal-callosal type (Feinberg *et al.* 1992). The callosal type is characterized mainly by intermanual conflict (see below) while the frontal-callosal type is dominated by the presence of frontal lobe reflexes.

Examination begins with observation during history taking when direct questions must be asked about any involuntary movements of the arms. While the patient is seated, the alien arm may reach out and grasp objects and have difficulty in releasing them (tonic grasping). The hand may involuntarily grasp and grope other body parts or pieces of clothing, or items within reach (instinctive grasp reaction). Sometimes the good hand

may reach over and try to prevent these movements by holding the alien arm, or may perform a movement that is antagonistic to the movement of the alien arm (intermanual conflict).

Manifestations of the alien limb syndrome may be brought out by a variety of procedures, such as testing for object use and ideomotor dyspraxia, i.e. pantomiming for transitive (e.g. using a screwdriver) and intransitive (e.g. waving goodbye) actions; tactile naming (asking the patient to name an object by touch alone); somaesthetic transfer (with the patient's eyes closed, the examiner touches the tip of one finger on the left hand and asks the patient to identify the same finger on the right hand); and cross-replication of hand posture (with the patient's eyes closed the examiner places the patient's hand in a specific posture and asks him or her to replicate the posture with the other hand). It can also be revealing to ask the patient to differentiate between his or her arm and the examiner's arm without visual input. The patient should be tested for their ability to perform tasks requiring bimanual interaction, e.g. tying up a shoelace. Utilization behaviour can be elicited by strategically placing objects within the patient's reach. A thorough general neurological examination is necessary to delineate any weakness, sensory impairment or involuntary movements.

During the examination particular attention should be paid to other dyspraxic features:

- Intermanual conflict (Fig. 1) – antagonistic actions between the two hands, e.g. on asking the patient to answer the telephone by picking up the receiver with his or her right hand, the left hand may interfere with the action of the right by simultaneously lifting up and putting down the receiver.
- Mirror movements – while performing a movement with one hand the other hand involuntarily performs a similar or mirror image movement, even when the involuntary movement is brought to the attention of the patient, and the patient is asked to restrain the mirrored movement.
- Diagonistic dyspraxia – these are movements that are in opposition to or irrelevant to the movement that is planned or willed, e.g. when asked to pull the chair forward, the arm may instead push the chair backwards to the surprise, frustration, anger, or even amusement of the patient.



Figure 1 Intermanual conflict. On asking the patient to answer the telephone by picking up the receiver with his (normal) right hand, the left hand (the alien one) interferes by quickly lifting up and putting down the receiver.

And to frontal lobe reflex phenomena:

- Utilization behaviour – inability to stop using or manipulating objects that are either in the patient's visual field or are brought into their visual field.
- Perseveration – repetitive performance of a motor task.
- Tonic grasping – difficulty in releasing grip.

THE POSTERIOR ALIEN HAND SYNDROME

The posterior alien hand syndrome is less well characterized. The features are somewhat similar to those described in the anterior alien hand syndrome and it also most commonly affects the left side. The patient complains of involuntary movements that are unwilling, and a sense of 'foreignness' to the arm. The movements however, are less complex, more episodic and slow in onset becoming more explosive and are significantly ataxic. Personification of the arm is very common and patients may report self-injurious behaviours, e.g. choking or strangling, pinching or repetitive facial trauma from the alien hand. Unlike the anterior alien hand syndrome, the neurological examination reveals dense hemianaesthesia, a homonymous hemianopia, neglect, moderate-severe ataxia, as well as occasional visuomotor dissociation, visuospatial deficit and hemiasomatognosia (body schema disturbances) ipsilateral to the alien hand. Notably, there is no dyspraxia nor any frontal reflex phenomena.

ANATOMICAL AND PATHOLOGICAL CORRELATES

The anterior alien hand syndrome is usually due to damage to the anterior-mid corpus callosum \pm mesial frontal cortex, including the supplementary motor area, while the posterior alien hand syndrome most commonly results from damage to contralateral occipito-temporal cortex, splenium of the corpus callosum and thalamus (Levine & Rinn 1986; Feinberg *et al.* 1992). The most common pathologies are vascular lesions, tumours, and surgical lesions, especially callosotomy.

The alien hand syndrome has also been described in various neurodegenerative disorders most notably corticobasal degeneration (Fisher 2000). However, in addition to the features described in anterior alien hand syndrome, the examiner may notice arm movements that the patient is unaware of. These include wandering and/or levitation of the arm (involuntary upward deviation of the limb when both limbs are placed in an outstretched horizontal position) with or without writhing finger movements. There may also be cortical sensory loss, dystonia and myoclonus of the affected arm. The pathology underlying corticobasal degeneration mainly affects posterior frontal and superior parietal cortices but also involves subcortical grey and white matter structures.

SUMMARY

The alien hand syndrome is a constellation of signs and symptoms. No one sign or symptom, for example arm levitation, is sufficient to make the diagnosis. A thorough neurological history and examination are necessary to exclude disorders that may mimic the alien hand syndrome. The most common neurodegenerative disorder associated with the alien hand syndrome is corticobasal degeneration.

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