Arq Neuropsiquiatr 2001;59(3-A):521-525

INVOLUNTARY HAND LEVITATION ASSOCIATED WITH PARIETAL DAMAGE

Another alien hand syndrome

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ABSTRACT – The alien hand syndrome (AHS) usually consists of an autonomous motor activity perceived as an involuntary and purposeful movement, with a feeling of foreignness of the involved limb, commonly associated with a failure to recognise ownership of the limb in the absence of visual clues. It has been described in association to lesions of the frontal lobes and corpus callosum. However, parietal damage can promote an involuntary, but purposeless, hand levitation, which, sometimes, resembles AHS. In the present study, four patients (cortico-basal ganglionic degeneration – n=2; Alzheimer's disease – n=1 and parietal stroke – n=1) who developed alien hand motor behaviour and whose CT, MRI and/or SPECT have disclosed a major contralateral parietal damage or dysfunction are described. These results reinforce the idea that parietal lobe lesions may also play a role in some patients with purposeless involuntary limb levitation, which is different from the classic forms of AHS.

KEY WORDS: alien hand syndrome, parietal lobe, Alzheimer's disease, cortico-basal ganglionic degeneration.

Movimento involuntário de levitação da mão associado a lesão parietal: uma variante da síndrome da mão alienígena

RESUMO – A síndrome da mão alienígena (SMA) geralmente consiste de uma atividade motora autônoma involuntária e aparentemente proposital, acompanhada de uma sensação de estranheza em relação ao membro afetado, muitas vezes associada a uma dificuldade em reconhecê-lo na ausência de pistas visuais. Tal síndrome vem sendo descrita em associação a lesões dos lobos frontais e corpo caloso. No entanto, lesões parietais podem promover um quadro de movimento involuntário de levitação da mão, aparentemente desproposital, mas que por vezes lembra uma SMA. No presente estudo, quatro pacientes (degeneração ganglionar córtico-basal – n=2; doença de Alzheimer – n=1 e infarto parietal – n=1) que desenvolveram tal quadro clínico e cujos estudos de TC, RM e SPECT revelaram um maior dano parietal contralateral, são descritos. Os dados encontrados reforçam a teoria que lesões parietais podem representar algum papel na gênese da levitação involuntária e desproposital da mão, a qual deve ser diferenciada das formas clássicas da SMA.

PALAVRAS-CHAVE: síndrome da mão alienígena, lobo parietal, doença de Alzheimer, degeneração ganglionar córtico-basal.

The motor phenomenon of the alien hand syndrome (AHS) was probably first reported by Goldstein in 1908[1], although Brion and Jedynak were those who clearly defined this neurological entity in 1972[2]. It has been commonly associated with frontal lobe damage, lesions of the anterior corpus callosum, or a combination of both. Two forms of AHS may exist according to Feinberg[3], a frontal and a callosal one. Frontal AHS is characterised by a compulsive manipulation of tools by the dominant hand, and is usually associated with frontal release signs (grasping and groping). The supplementary motor area (SMA), anterior cingulate gyrus, medial prefrontal cortex are implicated in this type of AHS. The second form, callosal AHS, is characterised by the presence of intermanual conflict and absence of frontal release signs. This type of AHS occurs in the non-dominant limb and a single lesion in the corpus callosum is the main finding.

According to Doody and Jankovic[4], the classical syndrome of AHS comprises three or four hallmarks: a) A feeling of foreignness of the limb; b) Failure to

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Received 29 October 2000, received in final form 1 March 2001. Accepted 10 March 2001.

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recognise ownership of it when visual clues are removed; c) Autonomous motor activities that are perceived as involuntary and are different from other identifiable movement disorders; d) Personification of the affected body part.

Furthermore, some patients with parietal damage may also feel a strangeness of the contralateral limb in association with an involuntary abnormal posture when the attention is distracted from it, resembling a levitation phenomenon[5]. Such clinical condition has been already reported and discussed by others[6-9].

The present study enrolled four patients who had a syndrome of purposeless involuntary hand levitation and, at least, three of the four hallmarks listed above, whose neuroimaging findings are suggestive of parietal lobe involvement.

CASES

Patient 1 – A 70 year-old woman was seen with a three-year history of gait disorder and parkinsonism, mainly affecting her left side, which was unresponsive to high-doses of L-DOPA. After one year, she started to rise involuntarily her left hand, when she was distracted or when she moved the right side. She also complained that the left hand did not belong to her, when visual clues were removed. Gradually, all the left side of the body became more rigid and dystonic. Rare myoclonic jerks appeared in this limb associated with a grasp reflex. Vertical upside gaze palsy and an important postural instability were also noticed. She obtained 17 out of 30 points at the Mini-Mental State Examination (MMSE)[10], with a poor performance on verbal memory tasks. Estereoagnosia with morphoagnosia and bilateral apraxia were also noticed. The EEG disclosed slow waves in right temporal lobe, and the MRI showed a right temporo-parietal atrophy (Fig 1). SPECT revealed a marked hypoperfusion in the same region (Fig 2). Clinical diagnosis was cortico-basal ganglionic degeneration (CBGD).

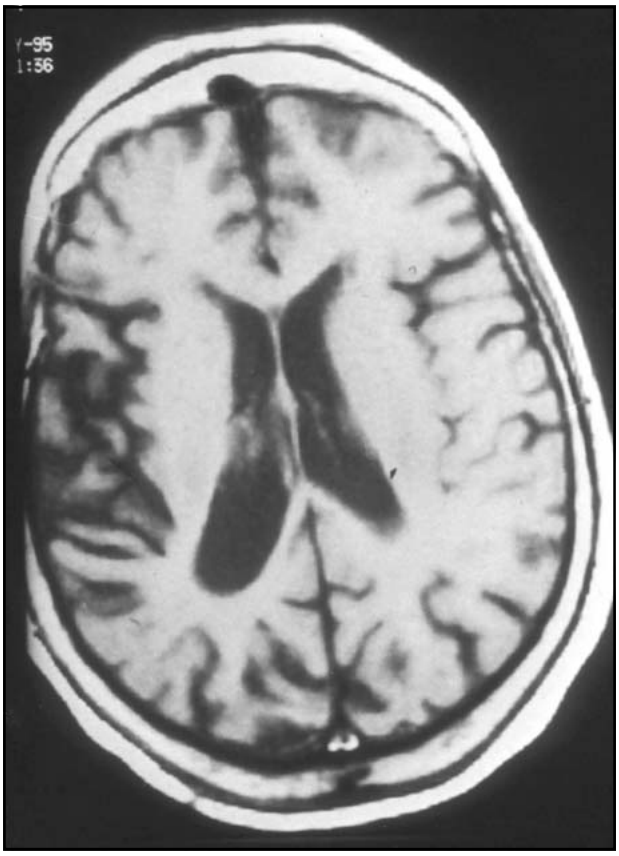

Fig 1 (patient 1). Right temporo-parietal atrophy disclosed by MRI.

Patient 2 – A 75 year-old woman was admitted with a three-year history of progressive left hand incoordination and difficulty in dressing. No memory complaints were

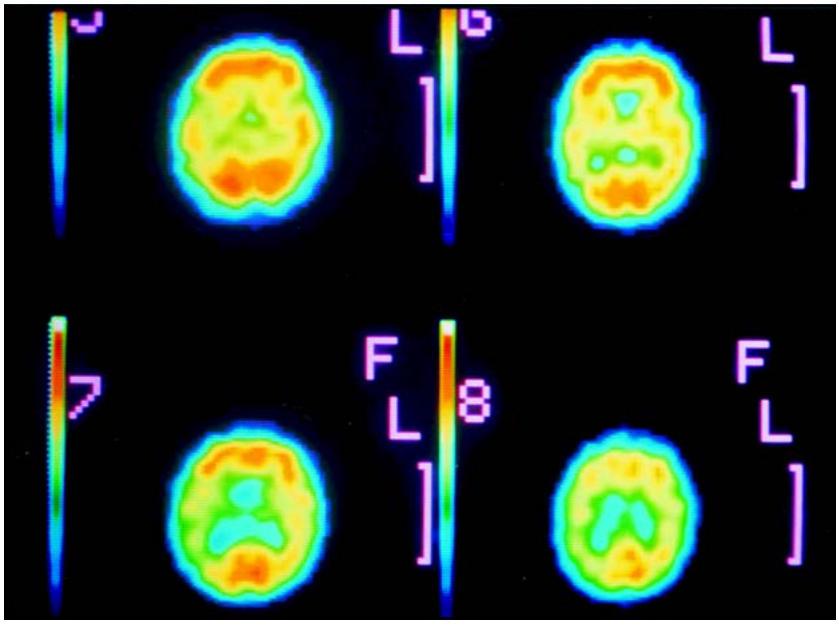

Fig 2 (patient 1). Right temporo-parietal hypoperfusion disclosed by SPECT.

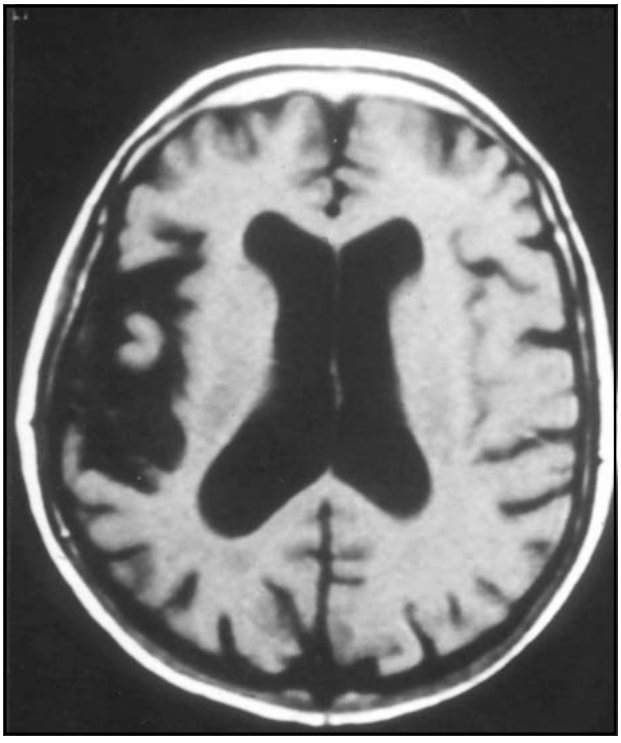

Fig 3 (patient 2). Right temporo-parietal atrophy disclosed by MRI.

recorded and she scored 26 points at the MMSE, with a poor performance on motor tasks because of ideomotor and constructive apraxias. Left side parkinsonism was present and the AHS appeared one year later. It was characterised by a purposeless left hand levitation, with this hand being considered "uncooperative" by the patient, who denied its ownership. MRI disclosed a right temporo-parietal atrophy (Fig 3). Clinical diagnosis was CBGD.

Patient 3 – A 63 year-old man was seen with a three-year history of progressive memory loss. By the time of the first evaluation, he scored 14 points at the MMSE. Right hemi-neglect, estereoagnosia, simultanagnosia and optic ataxia were observed. The right side was also mildly rigid with sporadic myoclonic jerks. Erratically, the right hand was raised without purpose, mainly when his eyes were closed. He also did not recognise it as his own limb. EEG showed slow irregular waves in left temporal region. MRI disclosed a global atrophy, mainly in the left parietal lobe (Figs 4 and 5). SPECT showed bilateral posterior hypoperfusion, most evident in left temporo-parietal region (Fig 6). Laboratory tests were unremarkable. Clinical diagnosis was AD.

Patient 4 – A 60 year-old woman suddenly developed numbness in the left hand. Neurological examination disclosed a mild left hemiparesis with Babinski sign and left hand dystonia. Severe impairment of graphestesia and stereognosia in the left limb were seen. The AHS was characterised by erratic, slow and irregular levitation of the left hand, increased by closure of the eyes. Frequently she complained that her limb did not belong to her and it had its "own life". MRI showed a hypointense area in the right

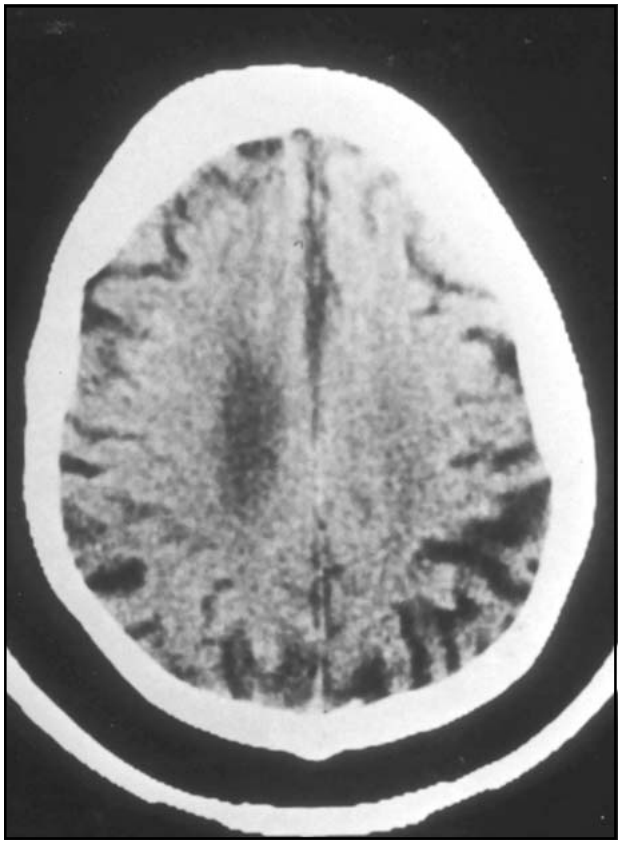

Fig 4 (patient 3). Left parietal atrophy.

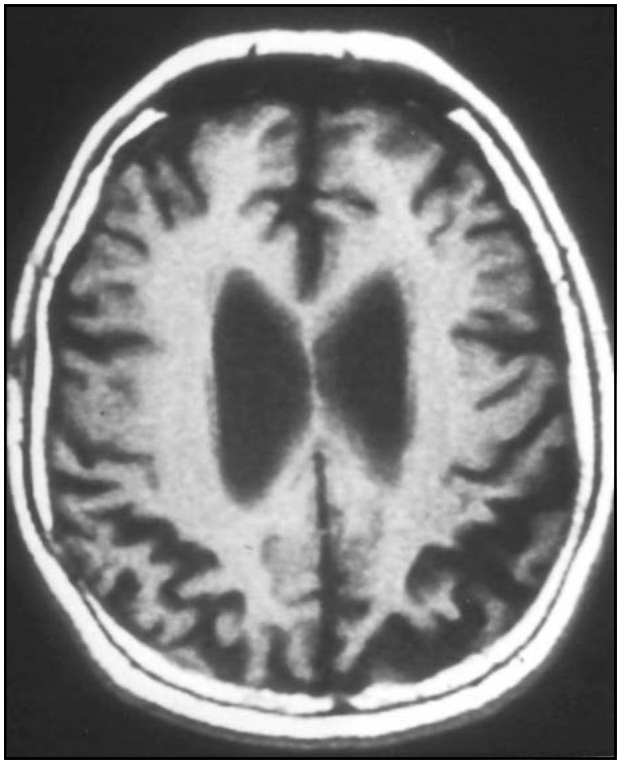

Fig 5 (patient 3). Left parietal atrophy.

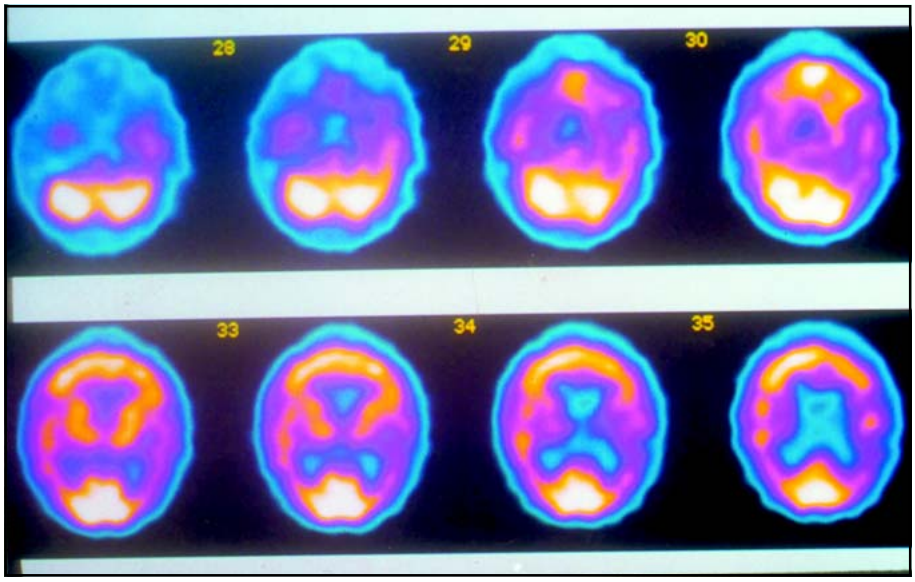

Fig 6 (patient 3). Left temporo-parietal hypoperfusion disclosed by SPECT.

Table. Summary of clinical data from the four patients with AHS

Patient	Main clinical feature	CT / MRI	SPECT	AHS	Probable Diagnosis
1	Parkinsonism	RTP atrophy	LP in RTP	Left hand levitation	CBGD
2	Parkinsonism	RTP atrophy	-	Left hand levitation	CBGD
3	Dementia	Global atrophy (+ left parietal lobe)	LP in RTP/LTP, but marked in RTP	Right hand levitation	AD
4	Mild left hemiparesis	Right parietal T1 hypointensity	-	Left hand levitation	Stroke

RTP, right temporo-parietal area; LTP, left temporo-parietal area; LP, low perfusion; AD, Alzheimer's disease; CBGD, cortico-basal ganglionic degeneration.

superior parietal lobule on T1-weighted scans, consistent with an old "watershed" infarct in the area between the posterior and middle cerebral arteries territories.

The Table summarises the clinical and neuroimaging features of patients reported herein.

DISCUSSION

The motor behaviour of AHS varies according to the topography of the responsible lesion. Fronto-medial and callosal damage are the most frequent findings in the majority of the reported patients[2-4,8,11]. This feature is easy to explain since these areas are clearly well related to motor planning and to its final pathways. Besides, electrical stimulation of frontal SMA is able to generate coordinated motor activity of the contralateral limb[12,13].

In the present study, however, an evidence of contralateral parietal damage or dysfunction, as disclosed by MRI and SPECT, was observed in four patients who experienced a feeling of absence or foreignness of the contralateral limb with hand levitation when attention was distracted from it and they denied the ownership of the hand. In spite of being a purposeless movement, all the other manifestations are essentially clinical hallmarks of a true, but not classic, AHS.

Three of our patients were affected by degenerative diseases, what does not allow us to rule out possible associated involvement of the frontal lobes in these cases. Notwithstanding, our patient 4 had an ischaemic parietal damage, with no evidence of a frontal lobe lesion, what gives a strong support to the hypothesis of the participation of the parietal lobe lesion in the pathophysiology of such kind of AHS.

The unusual association of involuntary movements with parietal dysfunction has been already reported before[6-9] and Ventura et al.[15] reported a 58 year-old right-handed woman who developed a left AHS after a right capsulothalamic haemorrhage. No

lesion was detected in the corpus callosum and a PET scan showed a diffuse right cortical hypometabolism, mainly in the sensorimotor area. Two other groups[9,14] have recently described isolated cases of AHS associated with posterior cerebral artery occlusion, which promoted a parietal lesion. In agreement to our hypothesis, Ay et al.[9] claimed that a "sensory" or "posterior" form of AHS has to be distinguished from the "motor" or "anterior" form described more commonly in current clinical practice.

The pathophysiology of AHS associated with parietal lobe lesion is not well understood. Dolado et al.[7] described a right-handed patient, with a right parietal infarct, who developed a left AHS when visual clues were removed. The authors hypothesised that dorsal parieto-occipital lesions may interfere with peristriate outflow pathways toward parietal zones, where visual somato-sensory interactions are likely to occur, and this interference may explain the emergence of the alien hand behaviour. Leiguarda et al.[8] also described two groups of patients with a paroxysmal form of the AHS. One group had frontomedial and the other, parietal damage. The former group had brief episodes of abnormal motor behaviour of the contralateral arm. In the other group, a paroxysmal feeling of unawareness of the location of the contralateral arm and purposeless hand movements, generally limb levitation, were observed. These last features resemble all cases of our study. These authors have proposed an epileptogenic basis to explain the paroxysmal nature of the AHS: an epileptic focus may release learned motor engrams from SMA and cingulate gyrus. Based on this hypothesis, these authors[8] argued that limb, axial and eye movements may also be evoked by epileptic activity in the parietal lobe. This occurs especially if the epileptic focus is within Brodmann areas 5 and 7.

This epileptogenic theory is very interesting, but it has to be confirmed[8]. Recently, however, Feinberg et al.[16] described a 61 year-old woman with a right hemisphere glioblastoma who developed AHS in the left hand. Simultaneous EEG documented continuous spikes in the right fronto-temporal region and diazepam IV reversed the movements and EEG. Our cases did not have any evidence of epilepsy nor of epileptogenic activity on EEG.

In conclusion, the present study reinforces the idea that the clinical spectrum of AHS should include not only the classical frontomedial or callosal AHS, but also a parietal form, characterised by stereotyped simple hand levitation. This form of AHS probably arises as a consequence of a major parietal lesion or dysfunction secondary to different diseases.

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