



Alien limb phenomenon following posterior cerebral artery stroke: a distinct clinical entity

Dennis A. Nowak¹ · Anna Engel¹ · Marlene Leutbecher¹ · Christiane Zeller¹

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Abstract

Alien limb syndrome following stroke within the territory of the posterior cerebral artery is exceedingly rare. A right-handed female experienced left homonymous hemianopia, visuospatial neglect, and proprioceptive loss of her left hemi-body. She experienced unintended, involuntary movements of her left arm and hand, which interfered with and disturbed motor actions of daily life performed with her right upper limb. There was no denial of ownership, but she interpreted movements of her left upper limb to be annoying, out of her will and unwanted. The alien limb phenomenon improved in parallel with improvement of proprioceptive loss over a 12-week of in-patient rehabilitation. A recently proposed theoretical concept of the alien limb phenomenon after posterior artery stroke is discussed.

Keywords Alien hand · Posterior cerebral artery · Stroke · Theory

Introduction

The alien limb syndrome is uncommon and defined by unwanted and uncontrollable movements of one limb in association with the subjective feeling that the limb is foreign, not belonging to one's own body and having a will of its own [3, 4, 6]. The alien limb phenomenon was most commonly described for the upper limb, but also the lower limb may be affected [6]. Today, several phenotypes of the alien limb syndrome can be discerned based on clinical presentation and lesion location [6]. Frontal alien limb features reflexive grasping, compulsive manipulation of objects and occurs primarily at the dominant hand [4, 11, 13]. Lesions in the dominant medial frontal lobe and anterior corpus callosum may cause this clinical entity. The callosal alien hand syndrome is characterised by inter-manual intention conflict, also termed callosal dyspraxia [6] and usually affects the non-dominant hand [4, 6, 8, 9]. Within recent years, a posterior alien limb phenotype has been described [6] that is most commonly observed in corticobasal degeneration [3, 6]. This clinical entity comprises rigidity, apraxia, cortical

sensory deficits and neglect, all in association with the alien limb phenomenon.

Only more recently, a clinical phenotype of alien limb has been documented in association with hemianopia, visual neglect and loss of proprioception after posterior cerebral artery stroke [1, 5, 10]. This entity appears to be exceedingly rare and its pathophysiology is still not completely understood [1, 5]. Here, we present a case of alien limb after stroke within the territory of the non-dominant posterior cerebral artery. The course of recovery and outcome after 12 weeks of in-patient neurological rehabilitation are illustrated. We discuss a recently proposed theory of alien limb after posterior cerebral artery stroke [1].

Case report

A 78-year-old right-handed Caucasian woman suffered sudden-onset somatosensory loss of her left hemi-body and homonymous left visual field impairment. In addition, she complained problems to control her left arm and hand. The left arm and hand were “moving unintended”, “having a will of their own”. When she was washing or dressing, her left hand was interfering with and disturbed activities of her right hand. She denied loss of possession, but described her left arm to be “annoying”, “performing out of her will” and “not obeying to her commands”. She suppressed the

✉ Dennis A. Nowak
dennis.nowak@vamed-gesundheit.de

¹ VAMED Klinik Kipfenberg, Neurologische Fachklinik,
Konrad-Regler-Straße 1, 85110 Kipfenberg, Germany

unwanted movements by placing the arm and hand under the blanket in bed, under a wheelchair table when sitting or wedging it between her legs. In the unwanted movements were no levitations. She was able to stop the movements, when guided to focus her attention to the left visual field, which was, however, possible for only a few seconds. The unintended movements of her left upper limb did not stop when she was distracted, e.g. by talking to the clinical examiner, a care giver or by performing wilful actions with her right upper or lower limbs. Computed tomography revealed an ischemic lesion within the territory of the right posterior cerebral artery (Fig. 1).

Nine days following acute symptom onset, she was transferred to our hospital. At this time, her neurological examination showed a left homonymous hemianopia and loss of proprioception of her left hemi-body. Proprioception was particularly impaired at her left upper limb with severe impairment of position sense, kinaesthesia and vibration sensation. Pinprick and light touch sensation of her left hand and arm were also, but less severely deteriorated. In addition, there were tactile extinction of her left upper limb and left visuospatial neglect. There were no ideomotor apraxia, no aphasia or writing impairment. No brainstem, cerebellar or frontal release signs or symptoms were evident. She still suffered from involuntary motor activity of her left hand, interfering with and disturbing daily life activities of her right hand, e.g. giving support when transferring from bed to wheelchair, brushing teeth or hair, buttoning a shirt or using cutlery. At this time, she interpreted the unwanted movements of her left hand to be “involuntary and alien” in nature. She tried to suppress unwanted motor activity by holding the left hand firmly with her right hand. The unintended movements of her left upper limb were not distractible and did not represent clinical levitations. She was aware that her left arm and hand belonged to her body. When not performing motor tasks with her right hand, the left hand

was frequently neglected, lying unattended under the blanket or wheelchair table or dangling along the wheelchair side.

A 12-week period of in-patient rehabilitation commenced. She received daily sessions of physiotherapy, occupational therapy and neuropsychological training over the entire rehabilitation program. Her progress in recovery of daily life cognition and motor activities was assessed once a week by the Functional Independence Measure [7], Early Rehabilitation Barthel Index [14] and Barthel Index scales [12]. The weekly scores of the Functional Independence Measure scale are illustrated in Fig. 2.

Over the first 5 weeks of rehabilitation, sensation to pinprick and light touch of the left hand and arm recovered slightly. Position sense, kinaesthesia and vibration sensation remained severely impaired. During this time, she constantly complained about unwanted behaviour of her left arm and hand and was hardly able to use it in bimanual motor

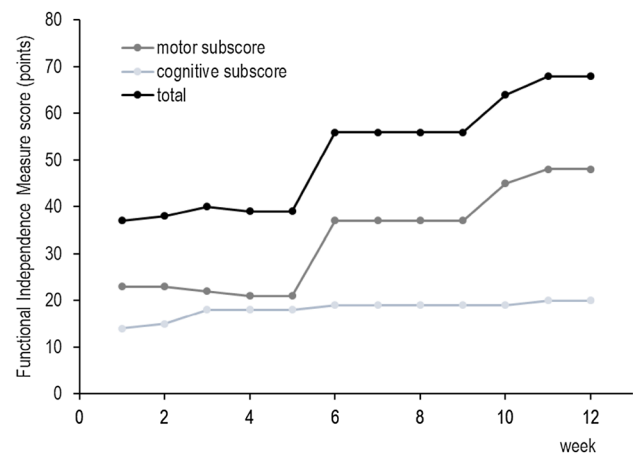


Fig. 2 Changes in cognitive and motor subscores as well as total score of the functional independence measure scale over the course of 12-week in-patient rehabilitation

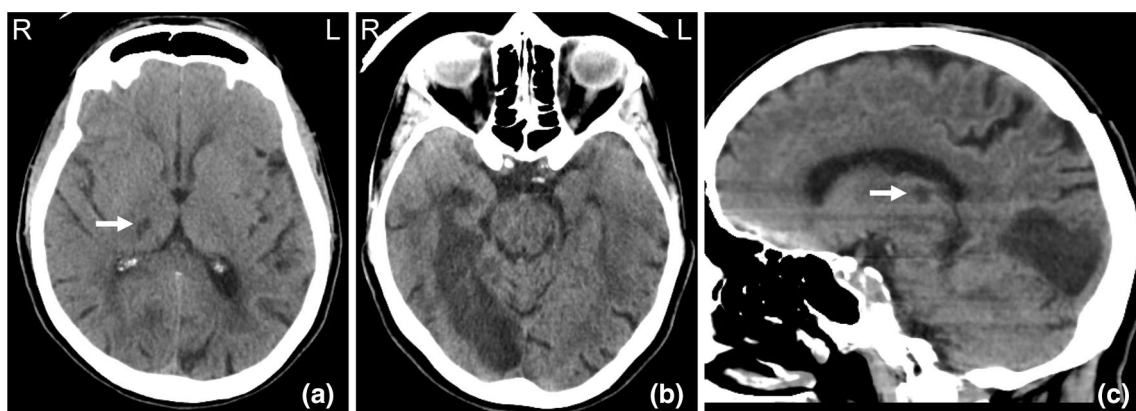


Fig. 1 Axial (a, b) and sagittal (c) computed tomography scans 2 weeks after acute symptom onset, revealing hypodense lesions within the right posterior thalamus (arrows), right occipital, medial temporal and inferior parietal lobes

activities of daily life such as squeezing out toothpaste, opening a bottle, washing and clothing. Visuospatial neglect of her left hemi-body remained unchanged. Between weeks 5 and 6, position sense, kinaesthesia and vibration sensation of her left arm and hand improved significantly. Visuospatial neglect as well as hemianopia remained stable. The alien limb phenomenon of her left arm and hand improved in parallel with proprioception. Involuntary movements of her left hand and arm occurred less frequently. She was less annoyed of her left upper limb and able to use the left hand constantly for bimanual motor actions such as dressing, buttoning or using cutlery. For the following 3 weeks, her neurological status remained mainly unchanged. From weeks 9 to 10, recovery of sensation to pinprick and light touch made significant progress. Position sense, kinaesthesia and vibration sensation of the left hand and arm also recovered significantly. Unintended movements of her left upper limb occurred only occasionally with less negative impact on voluntary motor action of her right upper limb. Within the commencing 2 weeks her neurological status made additional, but mild progress.

Thirteen weeks post-stroke, the alien limb symptoms had markedly improved. Only occasionally, the left hand interfered with voluntary motor behaviour of the right hand. She was able to perform bimanual activities such as sharpening a pencil, folding a sheet of paper and placing it into an envelope. Proprioception of the left upper limb also had significantly improved with only minor persisting impairment of position sense, kinaesthesia and vibration sensation at the metacarpophalangeal and interphalangeal joints. She was able to identify forms (triangle, circle, square) placed on her right palm without visual feedback and show them on a figure board with her left hand. However, she was unable to identify these forms when placed on her left palm without visual input (tactile anomia). Homonymous hemianopia was unchanged. Visuospatial neglect had mildly improved.

Discussion

Alien limb after posterior cerebral artery stroke is scarce with only a few cases available in the current literature. In our patient, left-sided alien limb was associated with left hemianopia, left visuospatial neglect and loss of proprioception of the left hemi-body. Alien limb symptoms recovered during a 12-week course of in-patient rehabilitation, as proprioception subsided. These clinical observations are in accordance with earlier reports, suggesting that loss of both proprioception and visual input from the affected upper limb causes the alien limb syndrome to manifest after posterior cerebral artery stroke [1, 5]. In addition to these key features, visual or tactile extinction had occasionally been described [1].

Coulthard et al. compared patients with posterior cerebral artery stroke who did or did not exhibit the alien limb syndrome using anatomical lesion overlay and subtraction [1]. Areas affected only in patients with alien limb syndrome, but not in those without the syndrome, were within the medial parieto-occipital lobe, thalamus and deep parietal white matter [1]. Also in our and earlier cases [3, 10] of alien limb syndrome after posterior cerebral artery stroke, these structures were affected (see Fig. 1). Damage to the occipital lobe is responsible for hemianopia. Damage to thalamic structures causes somatosensory impairment of the contralateral hemi-body. A theoretical framework of alien limb after posterior cerebral artery stroke proposed by Coulthard et al. [1] implies that loss of proprioception and visual feedback hampers the representation of an internal model about the current state of the affected upper limb [16]. This model, despite speculative, is illustrated in Fig. 3. Lack of information regarding the current state of the left hand and arm (e.g. muscle activity, joint position, limb orientation in relation to environmental objects, etc.) in conjunction with a deficit of movement intention may have resulted in a perception that the left hand and arm are moving unintended, e.g., unwanted and under “a will of their own.”

Sense of intention of motor action has been allocated to regions of the parietal lobe [2], whereas motor execution is well represented within frontal brain areas [11]. Awareness of voluntary movement of the upper limb is disturbed after lesions in the parietal cortex [15]. In our patient, a stroke-afflicted disconnection of the right dorsal parietal cortex from other relevant brain regions may have caused a lack of motor intention from the left arm and hand [6]. The question arises of how visuospatial neglect, frequently encountered in patients with alien limb, after posterior cerebral artery stroke of the non-dominant, e.g. right hemisphere, may have contributed to or reinforced the alien limb phenomenon. Our case clearly shows that improvement of both unintended motor activity and intention conflict occurred in parallel with improvement of proprioception, but not visuospatial neglect. This seems to suggest that the occurrence of alien limb after posterior cerebral artery stroke is closely linked to a deficit of proprioception.

Interestingly, a distinct phenotype of alien limb characterised by hemianopia and “purposeless wandering” and/or “instances of levitation” of the affected dominant hand had been described after posterior cerebral artery stroke of the dominant, e.g. left hemisphere [5]. In these cases, neglect, severe loss of proprioception, denial of ownership or interference with motor activity of the unaffected (non-dominant) upper limb were typically absent. Taken together, these and our clinical observations imply that the manifestation of an alien limb after posterior cerebral artery stroke requires incongruence between sensory feedback (visual and somatosensory input), internal representation of one’s

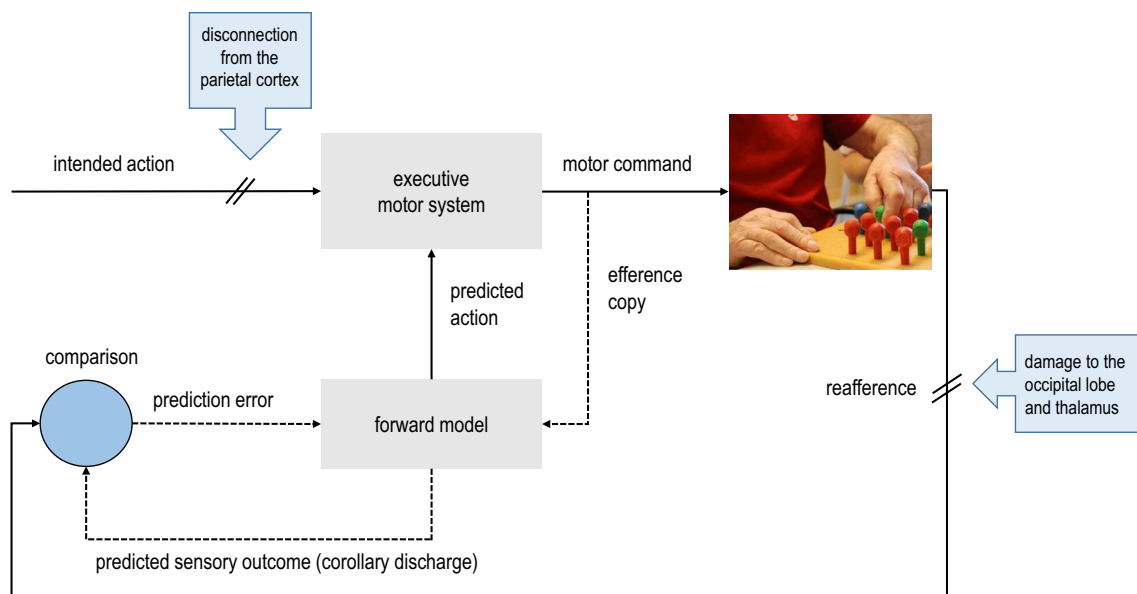


Fig. 3 An internal model (forward model) updates the brain about the current state of the arm and hand regarding muscle activity, joint position, limb position in three-dimensional space, etc. Given an intended action, the motor system generates both a motor command to be processed by the spinal motor neuron pool and a copy of the descending motor command (efference copy). The motor action generates sensory feedback (reafference). The internal model of the arm and hand uses a copy of the motor command (efference copy) and generates an estimate of the motor action (predicted action) in con-

junction with an estimate of the sensory outcome of the motor action (corollary discharge). A mismatch between the predicted and the performed motor action causes corrections. A mismatch between the predicted and actual sensory outcome of the action (prediction error) triggers an update of the internal representation of the arm and hand. Line crossings indicate the critical moments in motor intention and reafference to be disturbed in the alien limb phenotype to be observed after posterior cerebral artery stroke

own body scheme and motor intention (Fig. 3) [1, 5]. As body scheme appears to be represented within areas of the posterior parietal cortex of the non-dominant hemisphere [2, 15], a lack of motor intention after posterior cerebral artery stroke of the dominant hemisphere requires trans-callosal disconnection from the non-dominant hemisphere [5]. A limitation of the present report is that no functional neuroimaging or neurophysiological examinations were performed.

Alien limb syndrome after posterior cerebral artery stroke has a quite favourable outcome. In the case under discussion here, 13 weeks from symptom onset, involuntary movements of the left upper limb had markedly improved as had motor intention conflict. This is well in accordance to the course of recovery in cases of the frontal alien limb phenotype [13].

Compliance with ethical standards

Conflicts of interest On behalf of all the authors, the corresponding author states that there is no conflict of interest.

Ethical standards All procedures performed were in accordance with the 1964 Helsinki declaration and its later amendments. No ethics approval was obtained for this case report.

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