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Posterior alien hand syndrome: case report

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Abstract The alien hand syndrome (AHS) is involuntary uncontrolled movement of an arm with a sense of estrangement from the limb itself. AHS was initially used to describe interhemispheric disconnection phenomena in patients with lesions in the anterior corpus callosum, but it has been found in patients with posterior cerebral lesions without involvement of the corpus callosum, for example parietal infarcts or corticobasal degeneration. The posterior alien hand syndrome is less

frequent and presents with nonpurposive behaviour like lifting the arm or writhing fingers. We report an 80-year-old woman with a posterior AHS of the dominant right hand. MRI showed atrophy of the pre- and postcentral gyri without involvement of the corpus callosum. We discuss the aetiology of the posterior AHS and the differences from the anterior varieties.

Keywords Posterior alien hand syndrome · Cerebral infarct

Introduction

The alien hand syndrome (AHS) is defined as involuntary uncontrolled but partly purposive movement of an arm or hand with a feeling of foreignness or personification concerning the affected limb. The complex autonomous motor activity can be differentiated from other movement disorders [1]. The main features are persistent groping of the affected hand, compulsive manipulation of tools, intermanual conflict and aggressive actions, such as hitting or touching the face and body. Patients perceive these movements as aggressive and hostile; they often feel threatened and terrified by the affected limb [2].

Goldstein [3] first described the syndrome in a discussion of apraxias in 1908. The term AHS was introduced in 1972 by Brion and Jedynak [4] to describe a feeling of alienness of an arm in patients with lesions of the corpus callosum. Initially the term was restricted to those with disturbed interhemispheric communication due to lesions of the anterior corpus callosum alone. Since 1982 acceptable usage has broadened considerably, defining AHS more generally as a cluster of clinical

features without specific neuroanatomical correlation [5,6]. One reason for was due the finding, that AHS can result from lesions in different sites [7]. At present, three subtypes are recognised: two frontal and one parietal, with different underlying lesions and clinical presentations. In the frontal varieties, AHS with associated lesions in the language-dominant medial frontal cortex and anterior corpus callosum is differentiated from AHS caused by a lesion in the anterior corpus callosum alone. The term posterior AHS is used for patients with lesions of the parietal and occipital cortex and subcortical areas, excluding the corpus callosum [1, 8, 9]. While frontal and frontomedial AHS is more likely to present with exploratory grasping, compulsive manipulation and intermanual conflict, these specific and seemingly purposive movements are less distinct in posterior AHS [6].

Case report

An 80-year-old woman complained of slowly progressive immobility of her right arm over 3 years. Apart from arterial hypertension she was well until the age of 77 years when she first noticed impairment of controlled movement of her right hand, that became

progressive. She felt as if her right hand did not belong to her and had a “mind of its own”, complaining that her right arm “doesn’t do what it’s supposed to”. The involuntary movement consisted primarily of uncontrolled elevation of the right arm, sometimes provoked by a sudden noise or cough.

On examination, the patient was alert and orientated. Cranial nerves were intact. Extension of the right hand and fingers was markedly reduced, but flexion was nearly normal. Muscular tone of the right arm and right-handed reflexes were slightly increased, but no pyramidal signs were observed. Although not paretic, movements of her right shoulder and upper arm were markedly delayed. She was unable to imitate with her right hand postures adopted by the left and showed her left hand when asked to show the right.

MRI showed marked atrophy of the left pre- and postcentral gyri (Fig. 1) and white-matter lesions of their subcortical areas without involvement of the corpus callosum (Fig. 2). Cervical MRI was normal.

Discussion

Isolated cases of AHS without involvement of the corpus callosum have been reported [8, 9, 10, 11,12]. The patients had lesions in the parietal cortex and subcortical areas, mainly caused by parietal infarcts or corticobasal degeneration. AHS-like symptoms have been described in one patient with epileptic seizures [13].

In the posterior AHS movement disorders are less specific. The affected arm often shows undirected non-purposive movements and there is no intermanual conflict. In contrast to their classical frontal counterparts, sensory phenomena are described [1]. Disrupted cortico-striato-thalamic function which leads to poor

Fig. 1a, b. Axial MRI. **a** T1-weighted image showing marked atrophy of the left pre- and postcentral gyri with enlargement of the central sulcus. **b** A T2-weighted image reveals high signal in the subcortical white matter

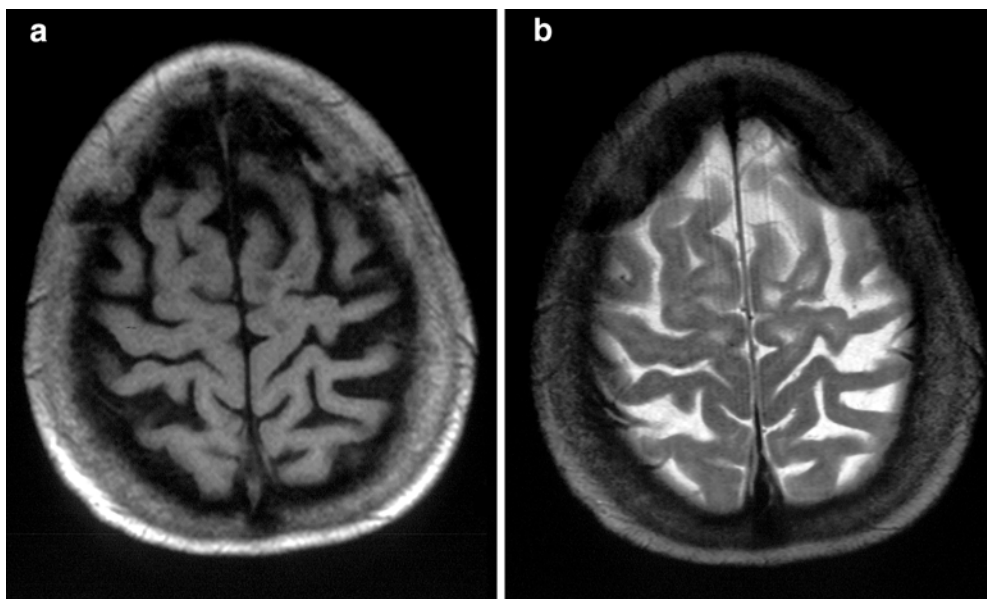
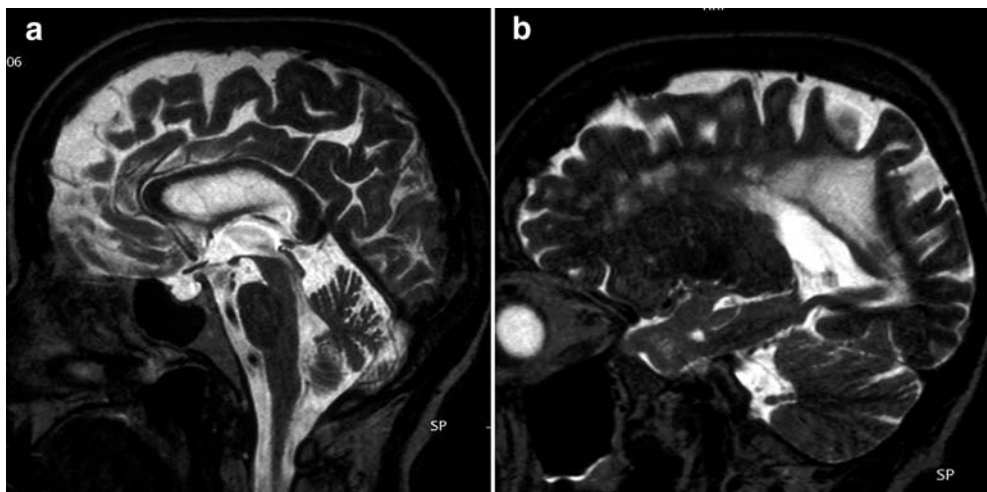


Fig. 2a, b. Sagittal T2-weighted images. **a** A midline image showing no circumscribed lesion in the corpus callosum. **b** A left paramedian image shows extensive high signal in subcortical and deep white matter, especially parietal



proprioception and sensory ataxia is a possible underlying pathology AHS [6]. In one report there was abnormal basal ganglia blood flow in a patient with hemidystonia while CT, angiography and electroencephalography (EEG) were normal [14].

Our patient fulfils the criteria for posterior AHS. She described strong feeling of estrangement from the affected limb. The distinct feeling of foreignness goes beyond the common apraxic disorder or absent perception of a limb in the sense of anosognosia or asomatognosia. Early on, a sudden noise or cough provoked uncontrolled lifting of the arm. To our knowledge, this has not previously been described the AHS.

MRI showed left central/parietal cortical and subcortical lesions and extensive atrophy of the pre- and postcentral gyri. We think these changes to be most likely of vascular origin, because of the subacute onset of the symptoms. Pathognomonic symptoms for corticobasal degeneration, such as extrapyramidal symptoms or dementia were absent. There was no signal change in the corpus callosum, thalamus or basal ganglia.

We think the sensation of estrangement is due to a distortion of the body image secondary to the postcentral cortical lesions, even though this phenomenon is usually observed in the nondominant hemisphere. The progressive involvement of the left pyramidal tract by a vascular process could have hindered the development of further involuntary movements. Bundick and Spinella [6] reported a posterior AHS caused by an infarct in the middle cerebral artery territory with motor symptoms disappearing while the patient was in hospital, while the proprioceptive disorder persisted.

The intensity of symptoms and subjective perception of the movements may differ between individuals because of the multifocal underlying pathology in posterior AHS. Moreover, they may follow a more dynamic course than their frontal counterparts. Some symptoms of posterior AHS may appear only for a limited time while others persist or progress with the underlying process.

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