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Unusual association of diseases/symptoms

Posterior alien hand in a left-handed person

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Summary

Posterior alien hand syndrome usually involves the non-dominant hand with lesions usually in the right hemisphere. This is the first case in a left-handed person, involving the dominant hand.

BACKGROUND

Alien hand syndrome (AHS) is a rare clinical condition, characterised by a perception of foreignness of the limbs, especially the upper limbs, and movements displaying a typical dissociation between an action and its voluntary/intentional control. A disconnection syndrome caused by neurodegenerative diseases, stroke, surgery, tumours, seizures or hemicranial aura[1–3] is observed in most cases.

There are three clinical variants. The first (motor type) is associated with lesions in the supplementary motor area, mostly on the left, and takes the form of grasping and groping. The second is associated with lesions of the anterior corpus callosum, and presents as complex intermanual conflict movements. The third, known as 'posterior or sensitive alien hand', is associated with lesions in the right parietal or occipital lobe, or the right thalamus. It is characterised by a sensation of extraneousness of the limb coupled with a more elementary motor activity in the form of frequent 'levitation' movements.

CASE PRESENTATION

An 89-year-old left-handed farmer, awoke with involuntary movements of the left upper limb and fewer of the left lower limb. His family history was positive for left handedness (two nephews). His medical history included hypertension and chronic obstructive bronchopneumopathy. On neurological examination at admission, he was alert and oriented to person, space and time. The Oldfield test was positive for left-handedness. He had normal comprehension, mild paresis of both left limbs with mild hypotonia and hyperreflexia, and slight hemiataxia. He presented a left hemisensory defect of both superficial and deep modalities, left spatial neglect and left homonymous hemianopia. His left limb displayed levitation episodes that could not be controlled voluntarily, neither spontaneously nor when trying to execute the finger-to-nose manoeuvre (see video 1). Grasping objects wit the left hand was ataxic and clumsy, although successful, both spontaneously and after a command. He regarded his left upper limb as his own, but complained that it 'had a will of its own', and his left hand 'did whatever it wanted'. He felt incapable of opposing this 'will'. He was unable, with eyes closed, to recognise his right hand with his left hand and often attributed it to the examiner. His involuntary movements were sometimes more jerky, especially if out of visual control. When his hand slapped his face and neck, he thought that the examiner or the nurses were hitting him. These motor abnormalities were distinct from both ataxia and paresis. His involuntary movements were more frequent when his attention was lower: during twilight, when he was more tired, and while falling asleep, when they aroused him.

Video 1 Involuntary movements and uncontrolled left arm levitation in a patient with posterior alien hand syndrome. 10.1136/bcr.10.2010.3401v1

INVESTIGATIONS

The first CT scan in the Emergency Room was normal. The second CT scan and MRI revealed ischaemic lesions at the right thalamus and calcarine cortex (positive on diffusion-weighted MRI). No other cerebral lesions were detected. MR angiography was normal. A video-EEG with recording of involuntary hand movements did not disclose any epileptic abnormalities.

TREATMENT

Three drugs commonly employed to restore nocturnal sleep (trazodone, mirtazapine, clonazepam) did not alter the clinical picture.

OUTCOME AND FOLLOW-UP

After 2 weeks the symptoms began to improve. After 2 months, AHS completely regressed, leaving a minor left sensorimotor hemisyndrome and left homonymous hemianopia.

DISCUSSION

Our patient recognised his hand as his own with open eyes; there was no intermanual conflict, grasping or groping, and self-restriction of movements was rare. Levitation was frequent, and personification of the hand was present. His awareness of the alien hand was less when its movements were not visually controlled. All these features are characteristics of the posterior AHS and differentiate it from the frontal and callosal types.[1–3] Involuntary movements are simpler, but associated with aberration of the sensory experience that often leads the patient to regard his arm as

endowed with its own intentionality. One peculiar aspect of our case was the nocturnal versus diurnal fluctuation. Movements were more jerky and frequent when attention was reduced, when the patient was tired, and during twilight and early sleep. Fluctuations of symptoms with anxiety and fatigue have been reported,[3 4] but no relation with sleep has been previously described. These fluctuations may be interpreted as the result of a reduced intentional executive control.

Classically, frontal AHS is reported only in dominant, and posterior AHS only in non-dominant hands, whereas the callosal form is reported in both.[5] However, this has been recently questioned and posterior AHS in dominant hands has been described.[5 6] Relationship with handedness is much more complex. This, in fact, is the first report of posterior AHS in a left-handed patient. Whether this is only due to chance or the brain organisation of left-handed people prevents or mitigates the development of AHS[1] is a open question.

Learning points

- Posterior AHS is less frequent than the other types and displays peculiar features (see video 1).
- This is the first case in a left-handed person.
- Posterior AHS usually involves the non-dominant hand with lesions usually in the right hemisphere. Only one case of AHS in the dominant hand has been reported so far (left lesion in a right-handed patient). In our case too, the dominant left hand was affected, and the lesion was in the right hemisphere.

Competing interests None.

Patient consent Obtained.

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