Supernumerary phantom limb after stroke

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The perception of a phantom limb is commonly reported after amputations. However, only a few cases have been described after a stroke. This article presents a patient who reported a supernumerary phantom limb (pseudopolydymelia) after spontaneous intracerebral haemorrhage and discusses the possible underlying mechanisms for this rare phenomenon.

The patient was a 71 year old, right handed retired secondary school teacher. He presented with severe weakness and sensory loss in the left arm and leg of sudden onset. General physical examination was unremarkable except for a blood pressure of 220/80. Neurological examination confirmed the presence of a dense flaccid left hemiplegia with facial involvement. There was also spinothalamic and posterior column sensory loss in the left half of the body. Assessment of the visual fields was not possible because the patient had severe visual impairment (see below). Similarly, formal assessment for hemispatial neglect (with pen and paper tests) was not possible. However, behavioural observation suggested the presence of left hemineglect. For example, the patient was not concerned when his left arm was caught in the wheelchair or when it was trapped under him whenever he rolled over in bed. The patient had full insight into his neurological impairment and answered correctly all items of the anosognosia for hemiplegia questionnaire. He was able to name his body parts correctly. Computed tomography of the brain confirmed the presence of an intracerebral haematoma 4 cm in diameter in the right lentiform nucleus and thalamus with extension into the lateral, third, and fourth ventricles. There was no hydrocephalus.

The patient suffered from longstanding hypertension and type 2 diabetes. For several years he was completely blind in the right eye due to a vitreous haemorrhage and his vision in the left eye was reduced to light perception because of macular degeneration. The medical history also included a brain stem ischaemic stroke from which he had made a complete neurological and functional recovery seven years earlier. There was no history of epilepsy, psychiatric illness, or drug or alcohol misuse. His regular medication was nifedipine, enalapril, and bendrofluazide. It was not clear why he was not taking antiplatelet drugs despite his vascular risks and previous ischaemic stroke.

Eight weeks after the onset of stroke the patient reported to the ward staff for the first time that he had a third leg. The patient’s belief of having a third leg persisted throughout his stay in hospital that lasted five months.

DESCRIPTION OF THE SUPERNUMERARY PHANTOM LIMB

The patient repeatedly reported that he had a third leg protruding from his left knee. He consistently maintained that the phantom leg was attached to his knee with a “bone plate” that “had no flesh on it”. However, he reported that the phantom limb itself looked normal and had a shinbone and a foot. It usually “appeared” in the morning when he was helped to put on his trousers. The patient stated that the phantom limb prevented him from turning over in bed, but did not adversely affect him otherwise. When asked about how he knew about this leg he said that he could see it (despite his severe visual impairment) and feel it with his hand. He believed that the phantom limb belonged to him, although he readily accepted that it was not “normal” to have three legs. Initially he reported that the “leg” was growing from his own knee, but then reasoned that (given its size) he would have noticed it before the stroke. At other times he believed the leg was attached to him by the nursing staff, but could not explain why. The patient was aware of phantom limb phenomena as his wife was an amputee. He was also aware that a stroke may affect perception and cognition. He did not believe either issue applied in his case.

ASSESSMENT OF COGNITIVE FUNCTION

The patient presented appropriately on the unit, being well orientated, alert, and with good communication and social skills. He was able to give a full history and showed good insight regarding the stroke and resultant hemiplegia. At no point did he seem confused or delusional. During the assessment and in more general conversation his speech was slightly slow, but this seemed to be attributable to a methodical verbal style rather than as a result of effortful processing.

The patient’s cognitive function was assessed formally, although because of his visual difficulties only material in the verbal modality was used. Receptive and expressive language were intact. Attention and concentration were good, with strong working memory (digit span forward and back both scoring 9 leading to a scaled score of 12). Verbal memory was assessed with the adult memory and information processing battery. On a list learning task initial recall was at the 25th centile, but there was a strong learning curve with final recall being beyond the 90th centile. On free recall task he had remembered the 15 items in their order of presentation, again showing a methodical approach supported by good attention and working memory. In a story recall task he performed in the 75–90th centile range in both immediate and delayed conditions.

Executive function was found to be similarly robust. The patient scored 14 on the “similarities” subtest of the Wechsler abbreviated scale of intelligence (WASI), which points to above average ability at verbal reasoning and conceptual thinking. Similarly he performed well on an assessment of verbal abstraction. His scaled score on the WASI vocabulary sub-test was 16. This points to performance in the superior range. A test of verbal fluency (that also gives information on the ability to inhibit responses and shift from one criterion to another) showed functioning in the borderline impaired range, although it is notable that the patient had a tendency to generate rather low frequency words (for example, “archipelago” and “syncline”).
In summary, the neuropsychological assessment confirmed that the patient’s cognitive abilities were largely intact. There was no evidence of other delusional beliefs or hallucinations, and in general his insight was good.

DISCUSSION

Phantom phenomena commonly occur after limb amputations and are sometimes reported by patients who have had a mastectomy, amputation of the penis, tooth extraction, or gum resection. However, to date very few cases have been reported after stroke. An electronic search of Medline database found fewer than 20 published cases in the past 50 years.

The patient reported here had the classic features of the syndrome of supernumerary phantom limb. The phantom perception was vivid, recurrent, and stable (that is, the description did not change over time). The patient was mentally alert and with normal cognition. There was no lack of insight for the neurological deficit (anosognosia for hemiplegia) or its consequences (anosadiaphoria). A psychological origin for the perception of phantom supernumerary limbs was proposed by Rogers and Franzen. However, our findings do not support this hypothesis. Our patient’s belief of having a third leg occurred on the background of normal cognitive function and in the absence of clinically detectable psychiatric illness or epilepsy. His behaviour throughout the observation period was completely rational and there were no delusions or hallucinations. The patient had no history of drug or alcohol misuse. Other authors, for example, Halligan et al and Halligan and Marshall have also reported normal cognition and psychological profile of their patients. This suggests an organic cause for the reported supernumerary phantom limbs.

The anatomical and functional integrity of the non-dominant parietal lobe and other brain structures and sensory feedback from the peripheral nervous system seem essential for bodily awareness (body schema). Disruption of any of these structures may result in the perception of phantom sensations. Supernumerary phantom limbs were first reported in patients with parietal lobe lesions. However, subsequently it became clear that other brain areas, including the thalamus, supplementary motor area, and the motor cortex contribute to the conscious perception of body schema. Sensory deafferentation seems to be the only mechanism in some patients. For example, Jacome described a patient who reported having two phantom supernumerary canine teeth that pressed on her tongue after resection of hypertrophic gums. The patient had no neurological disease. Her brain computed tomogram and MRI scans, as well as EEG studies were normal.

The reported supernumerary phantom leg in our patient occurred after a lesion involving the thalamus. It is possible that dysfunction of the non-dominant parietal lobe occurred as a result of diaschisis leading to disruption of the body schema. The symptoms developed in a patient with intact cognitive and psychological function. This combination of structural brain disease and the absence of mental illness seems to refute a psychological basis for post-stroke pseudopolymelia. This is consistent with previous findings of phantom phenomena occurring in patients with lesions of the neuro-axis, but no evidence of psychiatric or psychological abnormalities.

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