Neurocase (1999) Vol. 5, pp. 293–296

Acquired Synesthesia in Retinitis Pigmentosa

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Abstract

Patient PH developed retinitis pigmentosa in childhood and progressively lost his vision until he became completely blind at 40 years old. At age 42, he started experiencing vivid 'synesthesia'; tactile stimuli on the hand evoked a vivid visual sensation of 'movement', 'expansion' or 'jumping'. Intriguingly, the synesthesia was much more vivid when the hand was in front of the face rather than behind. The effect is unlikely to be confabulatory since touch thresholds (Semmes Monofilaments) were normal and identical for hand in front versus hand behind the head, while 'thresholds' for evoked visual sensations were significantly higher for the 'behind' condition. Also, the critical fusion frequency for the tactile sensation was much higher than the visually evoked ones. We propose three explanations. (i) 'Remapping' or 'cross-talk'. As a result of de-afferentation, sensory input to the somatosensory pathways (e.g. insular cortex) also innervates extrastriate visual areas. (ii) When a person is touched, there may be spontaneously evoked tactile associative 'memories' that would not normally evoke actual visual qualia because of competing 'spontaneous activity' from the visual pathways. However, upon de-afferentation, the associations may be experienced as synesthesia. (iii) After de-afferentation, the 'back projections' to somatosensory areas from visual areas may be strengthened.

Introduction

There is now a wealth of empirical evidence suggesting that connections in the adult brain can be modified by de-afferentation. For example, after amputation of a finger, the sensory input from adjacent fingers 'takes over' the cortex corresponding to the amputated finger. Also, after dorsal rhizotomy of one upper limb in monkeys, tactile stimuli applied to the ipsilateral side of the face activate the 'hand' area of area 3b in the cortex (Pons *et al*., 1991).

Consistent with these findings, we found that after arm amputation, in human patients, there is a similar reorganization of cortical maps; using magnetoencephalography (MEG), we showed that the input from the face and from regions proximal to the amputation begin to activate cortical areas corresponding to the hand (Ramachandran, 1993; Yang *et al*., 1994). Intriguingly, some of these patients also experience tactile sensations in their phantom hand if their face is touched, and we suggested that these referred sensations were a direct perceptual consequence of the reorganization of sensory pathways that has been demonstrated in both humans and lower primates (Ramachandran *et al*., 1992; Ramachandran, 1993). We refer to this idea as the 'remapping hypothesis' of referred sensations. These perceptual phenomena cannot be due to non-specific effects such as 'arousal' since they are often modality specific and topographically organized. For instance, a drop of water trickling down the face is felt as 'water trickling down the phantom', a vibrator on the face as 'vibration of the phantom hand' or a drop of hot water as 'warmth in the phantom', etc. The findings imply, instead, that a massive functional reorganization has occurred in the sensory pathways.

These findings also raise the interesting question of whether such remapping can occur across modalities, e.g. between vision and touch. There are no long-range connections that cross traditional cytoarchitectonic boundaries, so if such effects did occur, they would have to be mediated by sprouting of new pathways or through activation of indirect polysynaptic routes (e.g. there might be convergence of multiple sensory modalities in parietal cortex and de-afferentation of one modality might lead to a dominance of another—in a manner loosely analogous to shifts in ocular dominance after monocular deprivation). Could such spurious inter-modal connectivity be the basis of at least some kinds of synesthesia? For example, there are cells in both parietal and frontal cortex with dual receptive fields (both tactile and visual), and one wonders whether these cells might be involved in mediating such phenomena (Graziano and Gross, 1995).

For instance, if a patient became blind in early childhood, would his/her visual areas become activated by tactile or auditory inputs? If so, would such a person 'see' sounds or 'see' tactile stimuli? Obviously, one could not ask this of someone who was blind from birth because he/she would

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not know what ‘vision’ was. However, we can ask this of someone who lost vision late in life.

Case report

Patient PH came to see us because his vision had progressively deteriorated from the age of 5 years until he became completely blind at 40 and now, at age 42, he was experiencing odd ‘intrusive’ visual sensations when he tried to read Braille. We conducted several experiments to determine whether this was a true case of ‘acquired synesthesia’.

PH could not read the blackboard at age 5 years and was declared legally blind. His vision at that time was 20/100 and he recalls seeing colors, shapes and objects. At age 13, he was diagnosed as having retinitis pigmentosa, a hereditary disease that causes degeneration of the retina. In this disease, rod degeneration decreases night vision initially, and progressive tunnel vision may eventually result in complete blindness. PH’s vision deteriorated progressively until the age of 40, when he lost all vision including that for light and dark. At that time, there was no reaction to intense light in the right pupil, and a questionable reaction on the left. He also did not exhibit a consensual response or experience the entopic effect, indicating complete blindness.

PH started learning Braille in high school and found this helpful, but he had not noticed any unusual sensations at the time. He was a very avid and rapid reader of Grade 2 Braille (standard literary): PH read for 1–1.5 h per day for 7 years of college, and then for 3 h per day at his job for 18 years. During most of this period, he read 70 words per minute. Recently, his reading speed has decreased and he reads for a cumulative total of about 1 h per day, mostly addresses and agendas.

About 3 years ago, he started ‘hallucinating’ a small cluster of small red dots in his central vision. They were present continuously and eventually coalesced to form a reddish cartoon face that looked a bit ‘demonic’. It would sometimes change color slightly, especially the eyes, and its shape would also distort from time to time. He could not recall anything that enhanced or diminished this hallucination. The image seemed to move with eye movements. The face was initially flat, like a cartoon, but about 1.5 years ago it acquired solidity and depth.

His present symptoms began about 9 months prior to our testing him. He told us that whenever he touched things that had an edge, such as the arm of a chair or the corner of a room: ‘The edge would flash in my head and I would see a whole wall at an angle—it was exaggerated (compared) to what I knew it was’. The effect occurred with either hand. It did not always occur, but has become more consistent lately, he added. Eight months prior to testing, he started noticing a very vivid touch-to-vision correspondence while reading Braille. At first, he noticed vividly colored dots that would ‘shift visually’.

We conducted several experiments with PH. Our goal was to establish that this was a true case of ‘acquired synesthesia’ and that the patient was not merely using a figure of speech when he claimed to ‘see’ the Braille letters.

First, we simply tapped his finger and asked him if he saw anything (the tapping motion was up and down rather than from side to side). He said this caused the whole visual field to ‘jump’ up and down. The effect occurred with either hand. With repeated tapping and careful questioning, it became apparent that when his arm was placed in one visual hemifield and tapped, the visual sensation of ‘jumping’ occurred in the corresponding hemifield (left or right). This was repeated several times on two consecutive testing sessions. Additionally, it was determined that if he held his hand out in front when we tapped it (so that it occupied what used to be his normal visual field), he saw much more vivid distortions and ‘swirling’ in the field than if his fingers were tapped with his hand placed behind his head.

We were concerned that the visual experiences might have been confabulatory in origin, even though he said that he had not heard of the phenomenon of synesthesia. A more formal experiment was carried out to minimize this possibility and to explore his synesthetic experiences in greater detail.

Methods

We measured tactile sensory thresholds using Semmes Monofilaments. Our question was whether the threshold for touch would be identical to or different from the threshold for a change in visual sensations evoked by touch. Also, we wanted to find out whether the threshold for ‘visual distortions’ would change noticeably when his hand was in his ‘visual field’ (i.e. in front of his face) versus behind his head, whereas his touch thresholds should remain constant.

We measured these thresholds with a staircase procedure using Semmes Monofilaments (Lafayette instruments). Both touch and ‘visual distortion’ thresholds were measured by touching his hands when they were in front of the body versus behind the body. When his thresholds were measured for touch, each filament was applied to the glabrous surface of the index finger of his left hand (palm facing up) and PH was asked whether or not he could feel it. The filament number (strength) was then progressively increased (or decreased) until the patient could just feel (or stop feeling) the sensation. The data were obtained for six ‘reversals’ for each of the four experimental conditions at Time 1 and another six at Time 2 (3 h later). The purpose of the repeat test, peformed 3 h later, was to help rule out confabulatory effects since the patient would have difficulty in memorizing his own thresholds from the previous session (also, PH was not given feedback about the poundage corresponding to his thresholds).

In Condition 1, we obtained simple touch thresholds for sensations in the index finger of the left hand held in front of the body when the eyes were closed. Condition 2 was identical except that the hand was held behind the head. In this condition, his upper arm was parallel to the floor, his elbow completely flexed, but his hand extended behind his

Table 1. Just noticeable difference in pounds at different times for the experimental conditions

	Time 1		Time 2		Total	
Condition	Mean	SD	Mean	SD	Mean	SD
Touch Front	3.57	0.39	3.43	0.32	3.50	0.35
Touch Back	3.92	0.12	3.43	0.32	3.67	0.35
'Visual' Front	4.32	0.27	4.19	0.10	4.26	0.21
'Visual' Back	4.94	0.33	4.47	0.24	4.70	0.37

head. In Conditions 3 and 4, PH was instructed to report when he could ''just see (or stop seeing) distortions in the 'visual field'''. Thus, the testing procedures in Condition 3 were identical to those in Condition 1 (and 4 identical to 2) except that threshold measurements were for visual sensations produced by touch rather than for the touch sensations themselves. The order of the four conditions was randomized both at Time 1 and at Time 2.

Results

The thresholds for the four conditions were 3.50, 3.67, 4.26 and 4.70 lb. The means and standard deviations for the conditions at Time 1 and Time 2 can be viewed in Table 1. Four paired *t*-tests were performed. A comparison of the two touch conditions (hand in front of the body versus behind the head) was not significant [$t(22) = -1.23$, $P = 0.23$]. However, Touch Front and 'Visual' Front conditions were significantly different, $t(22) = -6.43$, $P < 0.01$, as were the Touch Back and 'Visual' Back conditions, $t(22) = -7.06$, $P < 0.01$. Additionally, the comparison of 'Visual' Front and 'Visual' Back was significant, $t(22) = -3.67$, $P < 0.01$. Thus, simple touch thresholds did not vary depending on whether the patient's hand was touched in front of his body or behind. 'Visual' thresholds did vary significantly from Touch thresholds. Finally, 'Visual' thresholds varied depending on whether the patient's hand was touched in front of his body or behind.

An attempt was also made to demonstrate that PH's experiences were not factitious by having him estimate the threshold at which he perceived distortions in the 'visual' field. When the patient was touched with his hand to the front, but asked to estimate what the threshold would be at the back that would produce visual disturbances, the mean was 5.88 lb and the SD 0.64 lb. When the patient was touched with his hand behind him, but asked to estimate what the threshold would be at the front that would produce visual disturbances, the patient produced a mean of 4.21 lb (SD = 0.11 lb). These results imply that the patient's 'memory' for the precise value of the threshold was relatively poor and could not have aided him in confabulatory responses across testing sessions.

Finally, when we tapped PH's finger repeatedly, he reported a 'jiggling' or 'flashing', in the visual field that followed the tapping. When we progressively increased the tapping speed, the 'flashing' frequency also increased until about 3 Hz (identical on two distinct trials). Above that speed, the taps could still be resolved clearly on the finger, but the visual sensations coalesced. In other words, the critical fusion frequency (CFF) was different for the tactile stimulus and the evoked visual sensation. This observation, once again, rules out confabulation for there is no reason why the patient would feel the need to concoct such a strange observation.

Discussion

We conclude that at least some patients experience synesthesia as a result of progressive deafferentation. Why has this phenomenon not been noticed before? There are at least three possibilities. First, a person who is blind from birth obviously would not see the effect because he would simply not know how to describe his 'visual' sensations. Therefore, a period of clear vision early in life may be required—so that the subject who 'remembers' what vision was like can describe the synesthetic experiences. Second, many such reports may, in the past, have been dismissed as just a 'figure of speech' and even confabulatory in origin. Third, the mechanism responsible for the experience, whether 'remapping' or strengthening of back projections from one sensory modality to another, may be a rare occurrence seen only in a minority of patients after deafferentation. Perhaps, progressive deterioration of vision from early childhood, culminating in complete blindness, is required. This can be explored by testing a large number of patients who have a clinical history similar to PH.

Two reports of patients with synesthesia discuss interesting findings relevant to our patient. Vike *et al*. (1984) describe a patient with no visual problems, but he had a large mass involving the medial temporal lobe and adjacent midbrain, and experienced auditory–visual synesthesia, i.e. he experienced a 'kaleidoscopic, spiraling' image of light upon hearing sounds. Like our patient, the phenomenon occurred ipsilateral to the stimulation and the rate of visual movement was dependent upon the rate of auditory stimulation. Halligan *et al*. (1997) reported findings on two stroke patients (one with left temporo-parietal and one with right internal capsule damage) who felt their bodies touched when they viewed their bodies being touched, even on sham trials in which their skin was not actually touched. This is similar to a phenomenon that occurs in some patients with phantom limbs (Ramachandran and Rogers-Ramachandran, 1996). A mirror is placed vertically on a table in front of the patient in the mid-sagittal plane, and the reflection of the intact arm is aligned with where the phantom arm would be if it existed. If the patient viewed the reflection of his existing arm in the mirror while the arm was being touched, he experiences the haptic illusion that the phantom is also being touched. This is another striking example of synesthesia in the sense that visual input evokes tactile sensation.

The obvious next step would be to conduct imaging experiments on patient PH (and others like him). Recent studies on blind subjects (Sadato *et al*., 1996) showed activity evoked in the visual areas of the brain during Braille reading, but curiously the patient did not report (or was not questioned about) visual sensations.

Our findings also raise the possibility that other types of synesthesia [including the 'idiopathic' or congenital variety (Baron-Cohen *et al*., 1987)] are also based on the kinds of mechanisms that we postulate for patient PH. It is hard to see how remapping or back projections in any straightforward sense can produce equivalence between color and letters of the alphabet or between shapes and taste (Cytowic and Wood, 1982), but such effects might arise because of 'cross wiring' in a higher dimensional space, i.e. in maps that represent proximity in other kinds of 'space' (e.g. color space or shape space) rather than spatial proximity (topography). These are highly speculative conjectures, but can be tested by combining the right kinds of cleverly designed psychophysical tests with non-invasive imaging techniques such as fMRI or MEG. Relevant to this idea is a PET study that was recently conducted on six synesthetic women in whom color synesthesia induced by auditorily presented words resulted in activation of several visual associative areas (Paulesu *et al*., 1995).

Acknowledgements

We thank Pat Churchland and F. H. C. Crick for stimulating discussions, and NIMH for support.

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Received on 22 June 1998; resubmitted 11 November 1998; accepted on 8 December 1998

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Abstract

Patient PH developed retinitis pigmentosa in childhood and progressively lost his vision until he became completely blind at 40 years old. At age 42, he started experiencing vivid 'synesthesia'; tactile stimuli on the hand evoked a vivid visual sensation of 'movement', 'expansion' or 'jumping'. Intriguingly, the synesthesia was much more vivid when the hand was in front of the face rather than behind. The effect is unlikely to be confabulatory since touch thresholds (Semmes Monofilaments) were normal and identical for hand in front versus hand behind the head, while 'thresholds' for evoked visual sensations were significantly higher for the 'behind' condition. Also, the critical fusion frequency for the tactile sensation was much higher than the visually evoked ones. We propose three explanations. (i) 'Remapping' or 'cross-talk'. As a result of de-afferentation, sensory input to the somatosensory pathways (e.g. insular cortex) also innervates extrastriate visual areas. (ii) When a person is touched, there may be spontaneously evoked tactile associative 'memories' that would not normally evoke actual visual qualia because of competing 'spontaneous activity' from the visual pathways. However, upon de-afferentation, the associations may be experienced as synesthesia. (iii) After de-afferentation, the 'back projections' to somatosensory areas from visual areas may be strengthened.

Journal

Neurocase 1999; 5: 293–6

Neurocase Reference Number:

O160

Primary diagnosis of interest

Retinitis pigmentosa

Author's designation of case

Retinitis pigmentosa

Key theoretical issue

- Synesthesia

Key words: retinitis pigmentosa; synesthesia

Scan, EEG and related measures

None

Standardized assessment

The patient was tested for pupil reactions to intense light, as well as consensual light reflex and the entoptic phenomenon

Other assessment

Threshold differences between touch and visual sensations evoked by touch were measured using Semmes Monofilaments

Lesion location

- Retina

Lesion type

Degeneration

Language

English