Pictorial pseudohallucinations with an “aperture effect” in a patient with quadrantanopia

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Abstract
Visual pseudohallucinations are reported on in a patient with a left lower quadrant anopia due to a right parietotemporal surgical defect after tumour removal. Besides metamorphopsia, he hallucinated the lower half of human figures which were limited to within the borders of the anopic defect and appeared “amputated” at the hip with one forearm and hand appearing from above in correct anatomical position. The lower half of these human figures was perceived as correctly scaled in relation to anatomical and environmental coordinates and was recognised as unreal. These pseudohallucinations led to the concept of an “aperture effect” which alludes to the visibility through the anopic field defect of a segment of the contents of the visual association cortex. This supports the idea of a pictorial mode for representation of endogenously generated images in the visual association cortex.

Keywords: visual pseudohallucinations; quadrantanopia; mental imagery

Both visual hallucinations and voluntary mental imagery convey internally generated percepts which lack a corresponding stimulus in the outer world.1 Pseudohallucinations differ from hallucinations as they are recognised as unreal although they appear as a sensory rich, perception-like image.2 Visual pseudohallucinations, first described by Hagen3 and Seguin,4 may occur after lesions of different parts of the visual system including bulbi, optic nerves, and optic chiasm,5 but especially the primary visual cortex and visual association cortices.6–9 Concerning the nature of internally generated mental images, it is controversially debated whether they are evoked as a picture-like object representation with preserved topological relations in retinotopic visual cortices10 or at a more abstract, propositional level, comparable to sentences describing a picture by using semantic labels for parts of it and commenting on the spatial relations between them. Here we report on a patient with metamorphoptic and pseudohallucinatory experiences. Most remarkably, he noticed “half human figures” limited to within the borders of his anopic field quadrant; this led to the concept of an “aperture” which, resembling a window, made visible just a segment of the pictorial content generated in his visual association cortices as a holistic Gestalt, thereby supporting the notion of a picture-like representation of mental images.

Case report
In 1978, the patient (aged 49 years) was diagnosed with neurofibromatosis; in 1987, his right arm was amputated because of a neurosarcoma. In 1993, a routine cranial CT showed an asymptomatic right parieto-occipital tumour which was resected and classified as a glioblastoma. Postoperative neurological examination disclosed a dense homonymous left lower quadrant anopia. Four months later he noticed metamorphoptic episodes developed when perceiving the body of his son as bent to the left from the hip upwards. After first considering that his son might walk in this distorted way, he soon found that all other people and all vertical lines in his visual environment were bent to the left. Later he experienced distortion of vertical contours of persons, furniture, windows, doors, etc, which seemed to expand and constrict rhythmically. Each of these metamorphoptic episodes developed suddenly and lasted for several seconds. Intriguingly, he also described visual pseudohallucinations of context-induced human figures confined to his anopic left lower quadrant, later recognised as being unreal; in one particularly remarkable instance he perceived a person selling soft drinks on the street he was driving along; however, the torso above the waist appeared amputated and one arm (delivering the soft drinks) appeared out of nowhere in an anatomically plausible position in relation to hip and legs at the height of the torso cut (see patient’s drawing (figure A)). The visually perceptible figure parts were correctly scaled and appropriately localised in relation to the street environment. The patient could not relate this scene to any past experience. An ophthalmological examination between these episodes disclosed the persisting homonymous quadrantanopia for the left lower quadrant (figure B) with normal visual acuity and colour vision (as assessed by colour naming). There was no
impairment for reading, spoken language, short or long term memory, praxis, or object recognition on bedside examination; in particular, no disturbances of visual or supramodal attention were evident during simultaneous bilateral stimulation. Muscle tone was slightly increased in the left leg without any other pathological motor signs or somatosensory disturbances. Brain MRI showed a right temporo-parieto-occipital white matter and cortex defect near to the posterior horn of the lateral ventricle. EEG disclosed alpha rhythm with some focal right occipital slowing, but no epileptic elements; no EEG could be obtained during the short episodes of his pseudohallucinations. Brain HMPAO-SPECT demonstrated a relative hyperperfusion in the right (plus 8%) compared with the left temporoparietal cortex rostral to the lesion.

Discussion

Pseudohallucinations after central lesions are well known: Kölmel described 16 patients (out of 120 patients with homonymous hemianopia) with complex visual pseudohallucinations in the hemianopic field; these comprised simple geometrical forms, flashing lights, coloured patterns, or the hallucination of complete persons, animals, and complex scenic hallucinations. Rarely, a single body part (in particular, hands) is described (patient 6; patient 3). Foerster induced formed hallucinations (animals, figures, people) by stimulating area 19. Similar hallucinations after stimulation more anterior to area 19 were later reported by Penfield and Perot.

The unique feature of the pseudohallucinations in our patient is that just those parts of a hallucinated context induced image were consciously perceived which were confined to his visual field defect, as if a complete figure became only partly visible when viewed through an aperture. That the hallucinated image had been generated endogenously as a complete figure can be inferred from the perception of body parts which were visually not directly connected with each other (arm and hip) but appeared in an anatomically appropriate configuration. This phenomenology favours a holistic, pictorial nature of internally generated images.

For this “aperture effect” to occur three processing features might interact (figure C): (1) the postgeniculate deafferentation of the primary visual cortex (here: a left lower quadrantanopia) caused a partial scotomisation of visual world input. (2) In the visual association cortices the remaining visual input induced context related visual associations; these were encoded in environmental coordinates (here, associative imagery of a man selling soft drinks on a street) and, when normally being covered by real world input, might be active only in a “background” stage as a “preconscious” halo of associations. (3) Through the anopic “aperture” (which was not
covered by real world visual input) a segment of these associations became visually perceptible.

During mental imagery, "long term memory knowledge of the visual appearance of objects or scenes is used to create a short term, percept-like image.\textsuperscript{12} Psychophysical\textsuperscript{14–16} and PET results\textsuperscript{17} corroborate the notion of a pictorial representation of mental images induced in primary visual cortices via a back projection neuronal network,\textsuperscript{18} and a recent fast MRI study demonstrated the involvement of both primary and visual association cortex when supporting either visual perception or imagery.\textsuperscript{19} Damage to this circuitry may cause a dissociation of visual perception and imagery faculties.\textsuperscript{11–13} Notably, if visual imagery is deteriorated damage is found in most patients in the left posterior hemisphere,\textsuperscript{15–19} which was intact in our patient. Recently Kosslyn et al\textsuperscript{20} showed a hemispheric difference in visual imagery with the left hemisphere arranging parts according to categorical spatial relations whereas the right hemisphere positions parts in precise locations in space according to coordinate-metric spatial relations. The right hemisphere involvement in our patient may contribute to this finding as a precise composition of a hand coming from nowhere in correct anatomical relation to hip and legs needs an appropriate metric spatial system.

Two mechanisms for visual pseudohallucinations have been discussed:\textsuperscript{22} epileptic phenomena are usually brief and paroxysmal in character and occur in a stereotyped manner. By contrast, non-epileptic “release” phenomena contingent on partial deafferentation of the primary visual cortex often persist longer and are non-paroxysmal in character; they may show a variable phenomenology. Studies in primates as well as PET studies in humans have accumulated evidence for two basic processing streams in the visual system, the “what” and “where” streams in the inferotemporal and parietal cortices.\textsuperscript{23–25} In our patient a pathological unilateral activation of these streams could be corroborated by demonstrating a significant hyperperfusion of the temporo-parietal cortex ipsilateral and rostral to the lesion. During HMPAO-SPECT no hallucinatory or pseudohallucinatory experiences were reported. Whereas this indicates a tonically increased excitability possibly reflecting “release” due to deafferentation, the brief paroxysmal character of his pseudohallucinations favours an underlying epileptic event. As no EEG recordings could be obtained during these episodes and his pseudohallucinations ceased to occur after two months, the relative contributions of these two mechanisms remains undetermined.

In conclusion, the visual pseudohallucinations in this patient comprise an “aperture effect” with only one half of hallucinated human figures visible within his anopic quadrant; this can be understood as a windowed view on to the pictorial associations generated in his association cortex.

7 Lance WJ. Simple formed hallucinations confined to the area of a specific visual field defect. Brain 1976;99:719–34.
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