Sound Induced Photisms In Pontine And Extra-pontine Myelinolysis

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ABSTRACT

BACKGROUND: Synesthesia refers to the simultaneous experience of one primary sensation in another sense mode. Central pontine myelinolysis (CPM) is a non-inflammatory condition affecting the pons more often seen in individuals with alcoholism or on whom presenting hyponatremia is rapidly corrected to supra-normal levels. Asymptomatic cases of CPM have been reported previously. Sound induced photisms represent a form of acquired synesthesia secondary to anterior visual pathways compromise.

CASE REPORT: A patient with history of chronic alcohol abuse developed sound induced binocular photisms. His brain magnetic resonance imaging (MRI) showed signal abnormalities compatible with pontine and extrapontine myelinolysis. He had remote history of pituitary apoplexy, panhypopituitarism and empty sella turcica. There was no evidence of optic chiasm herniation into the sella, dolichoectasia of the anterior cerebral arteries, or evidence of visual pathways abnormalities.

CONCLUSION: Sound induced photisms may constitute restoration to consciousness of primitive hierarchical phenomena in evolutive and ontogenetic terms, facilitated by central nervous system (CNS) demyelination not directly affecting anterior visual pathways.

INTRODUCTION

The brain may be construed as a series of interconnected mini-brains with the unique ability to supersede unitary specialization and function as a syncytium. The brain units may recourse to any other units with the purpose of assigning a more complex self-related meaning to what is experienced through the special senses, and prepare for the execution of the primordial imperative response, i.e., “fight or flight”. Synesthesia is defined as the simultaneous experience of one primary sensation in another sense mode, for instance, the hearing of a number may trigger the perception of a particular color [1]. Synesthesia may be developmental, or acquired. This author previously reported a patient with epileptic ictal synesthesia of temporal lobe origin as late sequel of encephalitis, and a second patient with volitionally reproduced miniaturized TV-like sectoral synesthesia, as an exceptional secondary form of eidetic memory [2,3] Sound induced photisms constitute an acquired synesthetic clinical phenomenon, initially reported by Jacobs, et al, in 1981[4]. These investigators described nine patients with visual loss resulting from lesions of the optic nerves or chiasm due to multiple sclerosis, tumors and vasculitis. The authors hypothesized that deafferentiation of the visual cortex allowed the appearance of symptoms. Their group of patients exhibited a gamut of sound induced visual phenomena, encompassing the perception of simple white flashes of light in one extreme, to the perception of colorful and complex hallucinations in the other. Of significance, sound photisms were perceived only over the defective portion of their vision, while startle was a common precipitant [4]. The patient herein described had no apparent visual loss, albeit, the photisms were lateralized to the periphery of his field of vision. Startle was a universal precipitant. There were no paroxysmal electroencephalographic changes noted during the experimental elicitation of sound photisms during the recording, excluding the presence of an exceptional variant of auditory reflex epilepsy.

CASE REPORT

A 68 year old bonsai artist was seen in neurological consultation because the visualization of oscillating granular bright lights over the left eye beginning four month earlier. Eventually the lights spread to both eyes and were better seen on the periphery of the eyes. The lights (photisms) were uniformly triggered by unexpected loud sounds and were more intense the louder the sounds were, and when he was drowsy or resting. He did not experienced them spontaneously or while asleep, and they were not precipitated by any other actions, like eating or walking. There was no associated symptoms, and in particular, no headache or disturbed awareness, following his sound-induced visual symptoms. He also provided history of transient blanching or discoloration of images and reduced detail (“as when studying low resolution computer images”) when focusing on objects with his left eye, only after exposure to bright light like when walking out his house into a clear, sunny day.
His past medical history was significant for essential tremors, remote pituitary apoplexy, panhypopituitarism, osteoarthritis, osteopenia with an old T12 vertebral fracture, benign prostatic hyperthrophy and elevated cholesterol. He had chronic back pain. He had required iliac and renal angioplasties for stenotic lesions. His regular medications included clopidrogel, levothyroxine, hydrocortisone, testosterone and tamsulosin. He had no family history of neurological illness. He smoked two packages of cigarettes a day and drank around five beers a day in addition to wine with dinner, for many years. On physical examination his blood pressure was 120/64 mm Hg, pulse 78 per minute and respirations 20 per minute. He had no fever. Mental examination, cranial nerve testing, muscle strength, muscle tone, gait and stance, deep tendon reflexes and sensory testing were normal. He exhibited mild tandem ataxia and postural tremors of the hands. His corrected visual acuity was 20/20 on the right eye and 20/30 on the left. Examination of the cornea, conjunctiva and retina were normal. He had early bilateral cataracts. The optic nerves were distinct and of normal appearance. His intraocular pressures were normal. No visual field deficits were present. Pupils were equal and reactive. Volitional saccades and ocular smooth pursuit were normal. He had no nystagmus. Complete blood cell count, chemical profile, liver function tests, visual and auditory evoked potentials were normal. His electroencephalogram at rest and during the induction of photisms was normal. He had no photoparoxysmal responses during photic stimulation. Brain magnetic resonance imaging (MRI) revealed a partially empty sella, a normally placed optic nerve chiasm, and obvious areas of amorphous symmetric T2 and FLAIR hyperintense signal abnormalities in the thalami and the pons, compatible with demyelination (figures 1 and 2). Extracranial MRA with contrast revealed greater than 80% stenosis of the left common carotid artery and 40% of the right internal carotid artery. Intracranial MRA was unremarkable. No aneurysms or dolichoectasias of the anterior cerebral arteries was identified. A left common carotid stent was placed with no complications. The episodes of light induced amaurosis of the left eye disappeared. In two years of follow up his sound induced photisms have not progressed or changed in nature.

DISCUSSION

Light induced amaurosis fugax refers to sudden painless monocular loss of vision when exposed to bright light, and heralds the presence of a subclinical, usually severe, unilateral or bilateral carotid stenosis [5.] It can be reproduced in the laboratory by delivering repetitive flashes of light to the patient eyes for sixty seconds ("photostress test"). This condition relates to retinal ischemia resulting in a delay in the regeneration of the visual pigments contained in the photoreceptors. In retrospect it was concluded, that this patient transient amaurosis was not related to his sound photisms because it disappeared following the successful placement of a stent on the stenotic left carotid artery. He had suffered pituitary apoplexy from a spontaneous hemorrhage many years earlier, requiring subsequent continuous hormonal replacement therapy. Visual loss is a late complication of pituitary apoplexy more often due to the spontaneous herniation of the optic chiasm into an empty sella and exceptionally, by elongated or dislocated A1 segments of the anterior cerebral arteries, giving origin to a pulse mediated indentation of the upper margins of the chiasm [6,7] There was no clinical, electrophysiological or radiographic evidence of compromise of the optic chiasm on this case, to justify his symptoms on delayed post-apoplectic visual deterioration.

CPM constitutes a pontine non-inflammatory demyelinating lesion, characteristic of individuals with alcoholism or malnutrition on whom initial hyponatremia is rapidly corrected to normal or supranormal levels [8.] There are some similarities between CPM, Marchiafava Bignami disease and Wernicke’s encephalopathy [9.] Clinical manifestations of CPM include ophthalmpoplegia, dysarthria, dysphagia, facial paresis, quadriaparesis, and in severe cases, progressive obtundation, coma, catatonia and locked-in state [8,9,10,11] The wide availability of MRI of recent years has greatly enhanced the capability of diagnosing this condition in its early stages, when symptoms may be non-specific and easy misinterpreted, especially in the absence of obvious electrolyte derangement. Extra-pontine lesions are identified in at least half of the individuals with CPM and eccentric or asymmetric variants of pontine myelinolysis may occur, whereas in the classic form the lesion strictly affects the basis pontis. Subclinical forms of CPM have been reported in alcoholics and in a minimum percentage of patients undergoing liver transplantation [10.] The pathogenesis of CPM is explained on basis of an osmotic insult. Rapid osmotic alterations caused by trans-membrane ionic shifts, result in changes in membrane function and cell volume, that ultimately trigger apoptosis. An alternative explanation to demyelination in CPM is cyanide toxicity, induced by...
the release of cyanide from central nervous system B12 vitamin by alcohol. Lesions of the brain stem may result in spontaneously appearing visual hallucinations normally referred as “peduncular hallucinosis”. Individual with peduncular hallucinosis typically have impairments of episodic memory, often coupled with confabulatory behavior [12]. Congruent audiovisual peduncular hallucinosis simulating audiovisual synesthesia, was reported by Taylor, et al, in a patient with multiple sclerosis and mesencephalic demyelination [13] A 11 year old boy with a suspected tegmental pontine encephalitis and extreme paucity of nocturnal REM sleep demonstrated with polysomnography, exhibited complex visual and auditory hallucinations in the waking state, that were explained by the authors as diurnal REM sleep compensatory rebound intruding into consciousness [14] Visual and auditory hallucinations are not typical of CPM and the patient herein described had no spontaneous hallucinations, as in the cases mentioned above. He had no memory impairment. Furthermore, he had no history of sleep disorders, albeit polysomnography was not completed in the absence of clear clinical indications. It must be acknowledge however, that alcoholics exhibit a myriad of sleep anomalies when formally studied, none ultimately manifested as sound induced photisms [15.] Thus, it is appropriate to define this patient curious occurrence as an acquired synesthesia, rather than visual hallucinations originating from abnormal arousal, mediated by the brain stem reticular activating system. In the presence of extrapontine lesions and no demonstrable anterior visual pathway dysfunction, is it not possible to pin point at a lesion responsible for his clinical phenomenon. It can be speculated that his synesthetic experience was shaped as a non-specific release phenomenon of a lower hierarchical conscious design, in evolution and ontogenetic terms. Taylor, et al, proposed in their example, that deregulation of the modulation properties of the superior colliculus resulted in release of cross-modal integrative functions assigned to the superior temporal sulcus [13.] Recent exciting investigations in the topic of synesthesia employing functional MRI and the experimental paradigms of auditory induced visual flash illusion and the flash lag effect, have generated insightful information on brain function [16,17] Vision and hearing are concerned with physically distant percepts that naturally have greater relevance in survival. While vision provide spatial location, hearing provide temporal information working in unison for the location of the object of interest outside visual detection. The brain is programmed to enhance cooperation between these two senses and hates discrepancies. It goes long ways in attempting to fuse vision with auditory perception adapting to audiovisual asynchrony. This applies to the extreme of displacing the site from where sound emanates to conform with was the eyes see, or at least think they see (the spatial and temporal ventriloquism aftereffect) [16] In other words, we hear what we think we see and we see what we think we heard. Today we know that specialty cortices are multi-potential in judging stimuli in order to expedite processing of the fight or flight response. By the same token, evoked responses in the temporal-auditory cortex synchronize with the visual cortex by way of electromagnetic frequencies in the gamma band serving to implement audiovisual synesthesia [18.] The question remains unanswered if synesthesia represents physiological proper responses with aggregated illusions, or if it is "the real thing", i.e., a unique form of genuine bimodal brain percept. In the race of evolution (and during early brain development since "ontogeny recapitulates phylogeny"), it seems that synesthesia was buried to unconsciousness for the sake of efficiency. The latter occurred perhaps at the expense of artistic creativity, but in favor of linguistics; all in order to protect the economics of physical allocations. Pathological conditions may restore to consciousness this ancillary and developmental puzzling mental property.

REFERENCES

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Illustrations

Illustration 1

FIGURE 1. MAGNETIC RESONANCE IMAGING. T1 CORONAL IMAGE SHOWING A NORMALLY PLACED OPTIC CHIASM
FIGURE 2. MAGNETIC RESONANCE IMAGING. T2 SAGITTAL IMAGE SHOWING PONTINE SIGNAL HYPERINTENSITY. NOTICE SMALL ISOLATED AREA OF SIGNAL ABNORMALITY AT THE SPLENU OF THE CORPUS CALLOSUM AND THE EMPTY SELLA TURCICA
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