Simultanagnosia as the presenting symptom in neuro-ophthalmology

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ABSTRACT

Simultanagnosia is a well-known neurologic symptom characterized by the inability to conceptualize the whole picture despite being able to see individual elements within a visual scene. The pathophysiology involves a lesion to the bilateral parieto-occipital lobe. We report two unusual cases of simultanagnosia and juxtaposed homonymous visual field loss involving aqueductal stenosis–related obstructive hydrocephalus and cardiac arrest due to Brugada syndrome. Clinicians should be aware that simultanagnosia can be the presenting symptom of neuro-ophthalmic disease.

KEYWORDS Aqueductal stenosis; Brugada syndrome; homonymous hemianopsia; simultanagnosia

Simultanagnosia is an uncommon symptom of parieto-occipital lobe lesions. Classically, it results in the inability to conceptualize the whole picture despite being able to see the individual elements within a visual scene. Stroke, trauma, tumor, and degenerative disorders have all been reported to cause simultanagnosia.^{1,2} We report two cases of simultanagnosia as the presenting symptom in neuro-ophthalmology (aqueductal stenosis–related obstructive hydrocephalus and cardiac arrest due to Brugada syndrome). To our knowledge, these are the first such cases to be described in the English-language ophthalmic literature.

CASE PRESENTATION

A 57-year-old white man developed signs and symptoms of simultanagnosia secondary to ventriculomegaly due to aqueductal stenosis with a subsequent ventriculoperitoneal shunt placement (*Table 1*). Magnetic resonance imaging of the brain showed moderate global cerebral atrophy, pronounced in the parietal lobes, and a right posterior ventriculostomy with supratentorial ventricular decompression

(*Figure 1a*). He scored 19/30 on Montreal Cognitive Assessment (version 7.1) testing, with points lost on delayed recall, visuospatial dysfunction, language, and attention. At 11 months, he experienced sudden worsening of symptoms; a computed tomography scan of the head demonstrated ventriculomegaly, for which he had a ventriculoperitoneal shunt revision leading to improvement of both ventriculomegaly and simultanagnosia symptoms.

In the second case, a 30-year-old white man developed signs and symptoms of simultanagnosia secondary to a witnessed cardiac arrest with subsequent cardiopulmonary resuscitation with several rounds of emergent cardioversion for ventricular fibrillation. Magnetic resonance imaging of the brain demonstrated symmetric and restricted diffusion on diffusion-weighted imaging in the frontal, parietal, and occipital regions with corresponding hypointensities on apparent diffusion coefficient imaging consistent with cytotoxic edema secondary to ischemic infarction (*Figures 1b, 1c*). He was found to have the *SCN5A* mutation (Brugada syndrome) and underwent automatic implantable cardioverter-defibrillator implantation. His family history was

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Table 1. Clinical features of the two cases

Variable	Case #1	Case #2
Chief complaint	Objects appear “congested”	Difficulty interpreting complex information
Visual acuity	20/20 right; 20/20 left	20/80 right; 20/80 left
Ishihara color plates	0/14 right; 0/14 left	0/14 right; 0/14 left
Visual field	Left homonymous hemianopsia with juxtaposed right paracentral homonymous hemianopsia	Juxtaposed denser inferiorly homonymous hemianopsia with macular involvement

Figure 1. Magnetic resonance imaging of the brain. **(a)** Axial T1-weighted image without contrast in case 1 showing mild to moderate global cerebral volume loss with pronounced atrophy of parietal lobes. Arrows denote areas consistent with producing simultanagnosia. The right parieto-occipital lobe is covered by a shadow from the ventriculostomy device. **(b)** Diffusion-weighted image in case 2 showing restricted diffusion in the frontal, posterior parietal, and occipital areas bilaterally. **(c)** Apparent diffusion coefficient image in case 2 showing hypointensities in the corresponding posterior parietal and occipital areas consistent with cytotoxic edema.

negative for cardiac disease, but testing of his daughter confirmed the *SCN5A* mutation.

On physical exam, both patients had reactive pupils with no relative afferent pupillary defect or anisocoria. External, anterior segment, intraocular pressure, and fundus exam were normal in both eyes with full extraocular movement and no oculomotor apraxia or optic ataxia. Both patients presented with color deficits on Ishihara color plates and homonymous hemianopsia on Humphrey visual field 24-2 (Figure 2).

DISCUSSION

In case 1, aqueductal stenosis–related obstructive hydrocephalus presented with juxtaposed homonymous hemianopic visual field loss and simultanagnosia. The presentation of visual field defect associated with hydrocephalus is uncommon and is hypothesized to be due to the mechanical force causing distension of the third ventricle as well as the increased intracranial pressure.³ In addition, simultanagnosia in the setting of obstructive hydrocephalus has only been described once in a patient who developed bilateral visual loss and simultanagnosia with decreased color vision after lumboperitoneal shunting for pseudotumor cerebri.⁴ His

visual symptoms were thought to be caused by the sudden reduction in intracranial pressure after lumboperitoneal shunting placement, herniation of the cerebellar tonsils, and rupture of previously asymptomatic aneurysms resulting in bilateral parieto-occipital lobe infarcts.⁴ However, our patient presented with simultanagnosia prior to shunt placement and experienced symptom improvement with both the placement and subsequent revision of his ventriculoperitoneal shunt. This supports the involvement of a separate etiology in the development of simultanagnosia associated with obstructive hydrocephalus.

In case 2, a juxtaposed bilateral homonymous hemianopsia was present due to bilateral parieto-occipital ischemia and thus hypoxic injury to the superior optic radiations. This partly corresponds to “watershed” areas of the brain particularly susceptible to anoxic brain damage. Decreased visual acuity was likely due to bilateral macular involvement. To our knowledge, no cases of simultanagnosia secondary to hypoxic injury of parieto-occipital lobes have been published in the setting of Brugada syndrome. Neuropsychological evaluation showed severe global cognitive decline but only relatively decreased measures of attention efficiency, attention capacity, and working memory. Measures of visual ability were not explicitly reported. Genetic testing showed a heterozygous mutation of the *SCN5A* gene, which encodes the alpha subunit of the main cardiac sodium channel. This gene is the most common mutation associated with Brugada syndrome but is also associated with long QT interval, type 3 dilated cardiomyopathy, and atrial fibrillation. Brugada syndrome is a common genetic cause of sudden unexpected cardiac arrest by predisposing otherwise healthy young adults to ventricular arrhythmia.⁵ Testing of the proband might help identify other at-risk relatives who might benefit from prophylactic automatic implantable cardioverter-defibrillator placement.

Simultanagnosia is a well-known neurologic symptom and is often found in combination with a triad of symptoms comprising Balint syndrome. We report two cases presenting with simultanagnosia and juxtaposed homonymous visual field loss primarily due to parieto-occipital disease, with simultanagnosia suggested using the Ishihara color plates. Simultanagnosia can be the presenting symptom of neuro-ophthalmic disease.

Figure 2. Humphrey visual field 24-2 in (a) case 1 showing left homonymous hemianopsia with a juxtaposed right paracentral homonymous hemianopic field defect and (b) case 2 showing juxtaposed, denser inferiorly homonymous hemianopsia.

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1. Farah MJ. "Visual Agnosia": Disorders of Object Recognition and What They Tell Us About Normal Vision. Cambridge, MA: MIT Press; 1990.

2. Brazis PW, Graff-Radford NR, Newman NJ, et al. Ishihara color plates as a test for simultanagnosia. *Am J Ophthalmol.* 1998;126: 850-851. doi:10.1016/s0002-9394(98)00187-1.
3. Kojima N, Tamaki N, Hosoda K, et al. Visual field defects in hydrocephalus [in Japanese]. *No To Shinkei.* 1985;37:229-236.
4. Miller NR. Bilateral visual loss and simultanagnosia after lumboperitoneal shunt for pseudotumor cerebri. *J Neuroophthalmol.* 1997;17: 36-38.
5. Li W, Yin L, Shen C, et al. SCN5A variants: association with cardiac disorders. *Front Physiol.* 2018;9:1372. doi:10.3389/fphys.2018. 01372.