



Case report

Prosopometamorphopsia and alexia following left splenial corpus callosum infarction: Case report and literature review



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ARTICLE INFO

Article history:

Received 26 October 2015

Received in revised form 15 August 2016

Accepted 17 August 2016

Available online 20 August 2016

Keywords:

Prosopometamorphopsia

Transient global amnesia

Alexia

Splenium

Retrosplenial cortex

ABSTRACT

Background: Lesions to the posterior section of the corpus callosum, called the splenium, and the immediate area have been separately associated with perceived visual distortions of the face (prosopometamorphopsia) or difficulty reading (alexia).

Case report: This case report describes a right-handed patient who complained of prosopometamorphopsia associated with the lower part of the face and alexia following infarction of the left splenium in the corpus callosum.

Conclusions: The splenium and adjacent retrosplenial cortex facilitate the transfer of visual information and memory function between the two hemispheres of the brain and along the Papez circuit, respectively. We believe that damage to this singular area of the brain could bring about several concurrent yet disparate symptoms, such as the reported prosopometamorphopsia and alexia with this patient.

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1. Introduction

The corpus callosum is a bundle of neural fibers that connects the left and right hemispheres of the brain. It is made up of white matter that is composed of millions of axons that have their dendrites and terminal buttons projecting in both the right and left hemisphere. The regions of the corpus callosum are organized by function, with the transfer of visual information specifically taking place in the posterior portion of the corpus callosum, the splenium [20]. Visual fibers project from the occipital cortex of one hemisphere of the brain and connect through the splenium to the opposite hemisphere [6]. Lesions of the posterior section of the corpus callosum, known as the splenium, and the immediately-posterior retrosplenium cortex, are known to be associated with metamorphopsia [4,6,9,12,14,22], amnesia [1,21,26,28], and alexia [5,10,23], although it is unusual for more than one of these symptoms to be present in a single patient. The condition metamorphopsia refers to general distortions in vision, often associated with macular degeneration. It can be detected using a test called the Amsler Grid, where a patient will perceive distortions in a simple, uniform grid that do not exist.

Prosopometamorphopsia (“prosopo-” meaning face) is a specific type of visual distortion where the patient will only perceive distortions when viewing the faces of others. This type of distortion is a separate condition

from prosopagnosia (the inability to recognize faces), and in most cases (though not all [2]), a patient suffering from prosopometamorphopsia will still be able to recognize faces despite their apparent distortions [19].

Patients with prosopometamorphopsia have reported distortions of the entire face [11] or just some portions of the face [4,16,18,25]. For most patients, the distortions will recede after several hours to several weeks following the infarct [11,19].

2. Case description

A 62-year-old, right-handed Caucasian man was evaluated for complaints of distorted vision. In particular, he described the mouths of people's faces as being distorted and enlarged. Other aspects of the face such as the nose and eyes were not distorted, only the mouth. Nothing else in the environment appeared distorted to the patient except for the mouths of other people. He reported that he did have some trouble recognizing faces, but could recognize other objects such as landscapes. He had primarily central visual field distortion, occurring monocularly in either eye and with binocular viewing. He complained that he had difficulty reading numbers and letters on the computer, while his ability to write was normal.

The patient was not taking any medication for his eyes and did not have any pertinent ophthalmic history. He was being treated for hypercholesterolemia and an episode of transient global amnesia in 2012, about three years prior to seeing a physician for the visual distortions

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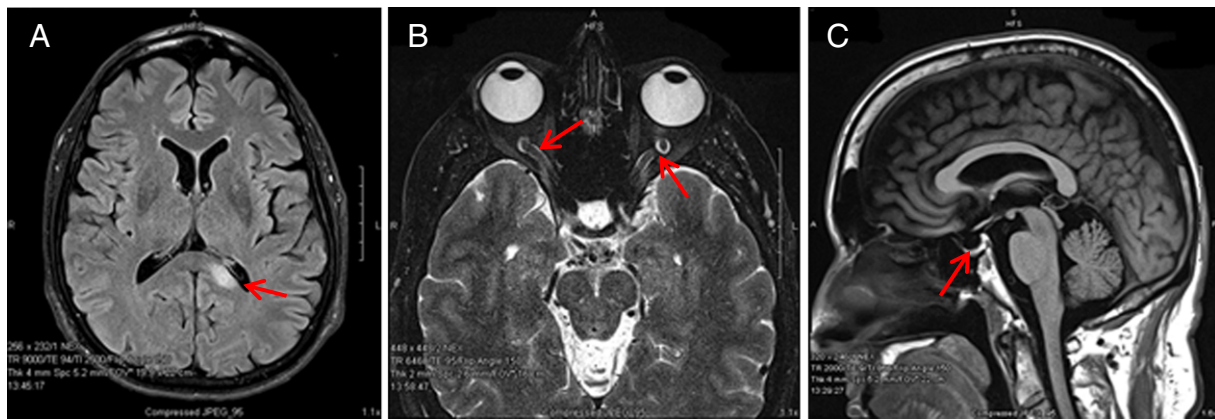


Fig. 1. A: MRI of the brain showing infarct within the left splenium. B: MRI of the orbits showing tortuosity of the optic nerves. C: MRI of the brain showing a partially-empty sella.

described here. At that time, he reported anterograde amnesia for a short period and had a full work-up with MRI, ECHO, MRA, and cardiac evaluations, all returning negative results.

His visual acuity at distance measured 20/40 OU without correction and color vision within normal limits. Pupils were equal and reactive without afferent pupillary defect. Extraocular ocular motility was intact. Amsler Grid testing revealed very mild distortion centrally OU, with the right eye being slightly more distorted than the left. Retinal examination and imaging (SD-OCT and fluorescein angiography) did not reveal any apparent abnormalities to explain the metamorphopsia. Obvious swelling of the optic nerve head was not appreciated. Given the specific nature of the complaints and the lack of an ocular explanation for the findings, neuroimaging was suggested.

T2-weighted brain MRI revealed a late subacute infarct within the left splenium of the corpus callosum, just medial to the occipital horn (Fig. 1A). Additionally, tortuosity of the optic nerves was observed, although it is unlikely that a corpus callosum lesion of relatively small size would cause an elevation in intracranial pressure (ICP) to a degree that would transmit elevated pressure to the optic nerve tracts as mentioned on MRI. The infarct in question is not causing much edema, has virtually no mass effect and is subacute, all of which would argue against a lesion like this actually elevating the ICP. However, if the lesion runs close to some of the venous outflow structures like the cerebral vein of Galen or straight sinus system, that could perhaps result in some elevated ICP.

T2-weighted orbit MRI revealed increased optic nerve sheath fluid consistent with papilledema and tortuosity of the optic nerves bilaterally (Fig. 1B), although papilledema was not apparent in prior fundus imaging. Additionally, a partially-empty sella was also seen, another nonspecific finding that can be seen in the setting of increased ICP associated with stroke (Fig. 1C).

3. Discussion

Prosopometamorphopsia is known to be caused by splenial corpus callosum infarction [14]. In this case, prosopometamorphopsia likely occurred because facial recognition information was interrupted by the splenial infarction as it was being transferred to the facial fusiform area after being processed in the face perception areas of the occipital lobe.

We believe that the patient's report of having difficulty reading numbers and letters on the computer may be alexia also stemming from damage to the splenium. His reported alexia without any agraphia has been reported in several other cases as well [10,15,17,23]. An infarct to the left posterior cerebral artery (which crosses the splenium of the corpus callosum and left visual cortex) is often involved in alexia [5, 10]. Although the visual cortex may still be able to process visual information without issue, it is unable to send this information to the language areas of the brain (Broca's and Wernicke's Areas) in the opposing hemisphere via the splenium. Stommel et al. reported a similar case of pure alexia, presuming that the condition was due to a lesion in the splenium that intercepted both the visual association fibers from the right occipital cortex and the fibers coming directly from the left visual association cortex on their way to the left angular gyrus [23].

We have added our case report to the table of previous reports on prosopometamorphopsia caused by splenial lesion constructed by Lee et al. [14] (Table 1).

It is possible that the infarct may be associated with this patient's episode of transient global amnesia. The retrosplenial cortex is immediately posterior to the splenium and provides connections between the anterior thalamus and the medial temporal structures involved in memory, altogether known as the Papez circuit. Amnesia has been associated with disruptions in communication in all three of these regions [27].

Table 1
Previous reports of prosopometamorphopsia following splenial or retrosplenial lesion.

| Age/sex | Handedness | Side of prosopometamorphopsia in the opposing face | Locus of lesion | Cause | Reference |
|---------|---------------|--|---------------------|--|-----------------------|
| 68/F | Right | Right | Right retrosplenium | Cerebral hemorrhage | Ebata et al. [7] |
| 58/F | Right | Left | Left splenium | Cerebral infarction | Cho et al. [4] |
| 53/M | Right | Left | Right splenium | Cerebral infarction | Cho et al. [4] |
| 68/F | Not described | Left | Left retrosplenium | Resection of an arteriovenous malformation | Ganssauge et al. [9] |
| 61/F | Left | Right | Right splenium | Cerebral infarction | Saito Y et al. [22] |
| 70/F | Right | Left | Right splenium | Cerebral infarction | Hishizawa et al. [12] |
| 52/F | Right | Left | Left splenium | Cerebral infarction | Lee et al. [14] |
| 78/F | Right | Left | Right splenium | Cerebral infarction | Nagaishi et al. [18] |
| 62/M | Right | Not described | Left splenium | Cerebral infarction | Present case |

Table 2

Previous reports of amnesia following splenial or retrosplenial lesion.

| Age/sex | Locus of lesion | Amnesia described | Reference |
|---------|---------------------|------------------------------------|------------------------|
| 62/M | Left splenium | Transient global amnesia | Ay et al. [1] |
| 58/M | Left retrosplenium | Transient global amnesia | Saito K et al. [21] |
| 81/M | Left retrosplenium | Retrograde amnesia | Takayama et al. [24] |
| 39/M | Left splenium | Retrograde and anterograde amnesia | Valenstein et al. [26] |
| 62/M | Right retrosplenium | Anterograde amnesia | Yasuda et al. [28] |
| 39/F | Central splenium | Retrograde and anterograde amnesia | Jeong et al. [13] |
| 18/M | Central splenium | Anterograde amnesia | Cha et al. [3] |
| 62/M | Left splenium | Transient global amnesia | Present case |

However, given the negative results of the MRI scans conducted in 2012 (immediately after the episode of TGA), it seems more likely that the amnesia was caused by a tiny lesion that could not be detected on MRI at that time, and not the splenial lesion identified here.

Nonetheless, it is possible that the amnesia and the lesion seen here are somehow related, as three regions related to memory function have been shown to have reciprocal connections through the retrosplenial cortex in monkey studies: the hippocampal formation, the parahippocampal region, and anterior and lateral dorsal nuclei of thalamus. Amnesia has been associated with disruptions in communication in all three of these regions [21,27]. Previous reports of amnesia associated with damage to the splenium and retrosplenial cortex have been compiled here (Table 2).

It seems there are relatively few studies of patients experiencing prosopometamorphopsia, alexia, and possibly amnesia associated with a single infarct location. Examining this case allows us to reiterate the importance of the region to many higher brain functions that rely on communication between hemispheres and regions of the brain, but further research is required to elucidate the exact mechanisms of these symptoms and how they relate to the precise location of the splenial infarct.

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