

# Scattered depressions with temporal preponderance in visual field test coexisting with optic disc temporal atrophy in cerebral arteriovenous malformation

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**Abstract:** In this article, the unusual association of optic disc temporal atrophy associated with scattered depressions with temporal preponderance in visual field test resembling incomplete bitemporal hemianopsia is reported. A 22-year-old man with cerebral arteriovenous malformation (AVM), which was located adjacent to the inferomedial portion of the posterior limb of the right internal capsule at the level of lateral ventricle, revealed interesting and unexpected ophthalmological findings. Possible mechanisms including anatomical variant, previously larger AVM, and retrograde optic neuropathy were mentioned. This case also highlighted that the usual complaint of visual disturbance might associate with unusual visual field defect in cerebral AVMs.

**Keywords:** arteriovenous malformation, bitemporal hemianopsia, optic chiasm, optic nerve, vascular steal

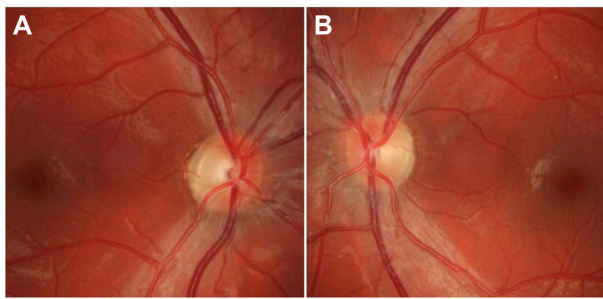
## Introduction

Cerebral vascular abnormalities including arteriovenous malformations (AVMs) are common incidental and asymptomatic findings on noninvasive imaging studies.<sup>1</sup> AVMs are considered as slow-growing congenital lesions that can cause wide spectrum visual field defects due to involvement of the visual paths. Since each AVM is associated with its own unique natural history, the association between the characteristics of AVM and visual field defects remains unpredictable. There is no evidence indicating a correlation between the size and localization of AVM and the pattern of visual field defect.<sup>1,2</sup> However, the closer localization to the posterior cerebral region can cause more congruent visual field defect in general.<sup>1</sup> In this article, we present a case with nonchiasmatic cerebral AVM, which revealed optic disc temporal pallor associated with scattered depressions with temporal preponderance in visual field test resembling bitemporal hemianopsia.

## Case report

A 22-year-old man suffering from visual disturbance, starting 5-years ago and gradually decreasing over time, presented to our ophthalmology department. Although his previous ophthalmic examination data were not available, he had no family history of any systemic or ophthalmologic disorders. At presentation, his best corrected visual acuity (BCVA) was 20/50 in the right and 20/100 in the left eye. Dilated fundoscopic examination revealed bilateral optic disc temporal pallor (Figure 1).

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**Figure 1** Dilated fundoscopic examination revealed bilateral optic disc temporal pallor.

**Note:** (A) right eye and (B) left eye.

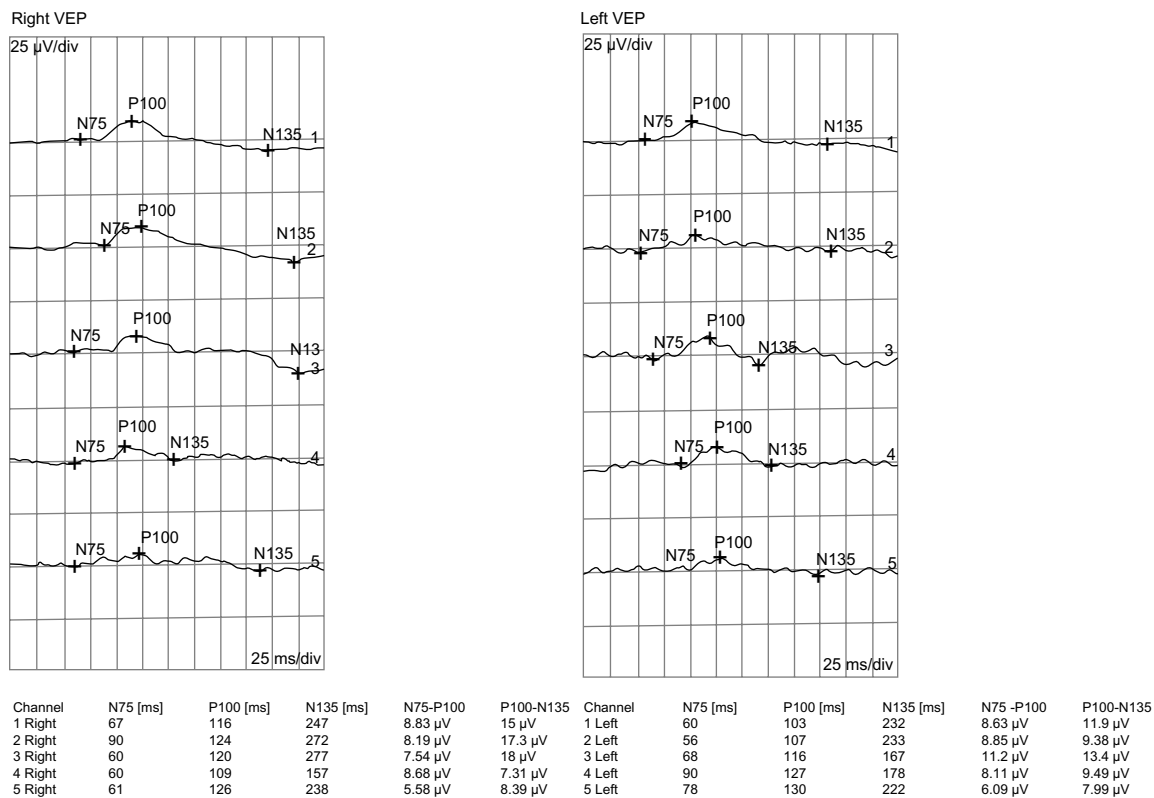
Visual evoked potentials (VEP) revealed delayed P100 latencies particularly observed in the smaller patterns in both eyes (Figure 2). Pupillary light reflex was normal in both eyes. Automated visual field testing (Humphrey Instruments Inc., San Leandro, CA, USA) demonstrated scattered depressions with temporal preponderance resembling incomplete bitemporal hemianopsia (Figure 3). Retinal nerve fiber layer (RNFL) map analyzed with spectral domain optical coherence tomography (SD-OCT) confirmed the presence of optic disc temporal atrophy (Figure 4). Brain magnetic resonance imaging (MRI) and cerebral computed

tomography angiography (CTA) revealed AVM, which was located adjacent to inferomedial portion of the posterior limb of the right internal capsule at the level of lateral ventricle (Figures 5 and 6). Chiasmatal compression or atrophy was not observed. AVM was draining into the choroidal veins at the level of the posterior horn of right lateral ventricle.

### Discussion

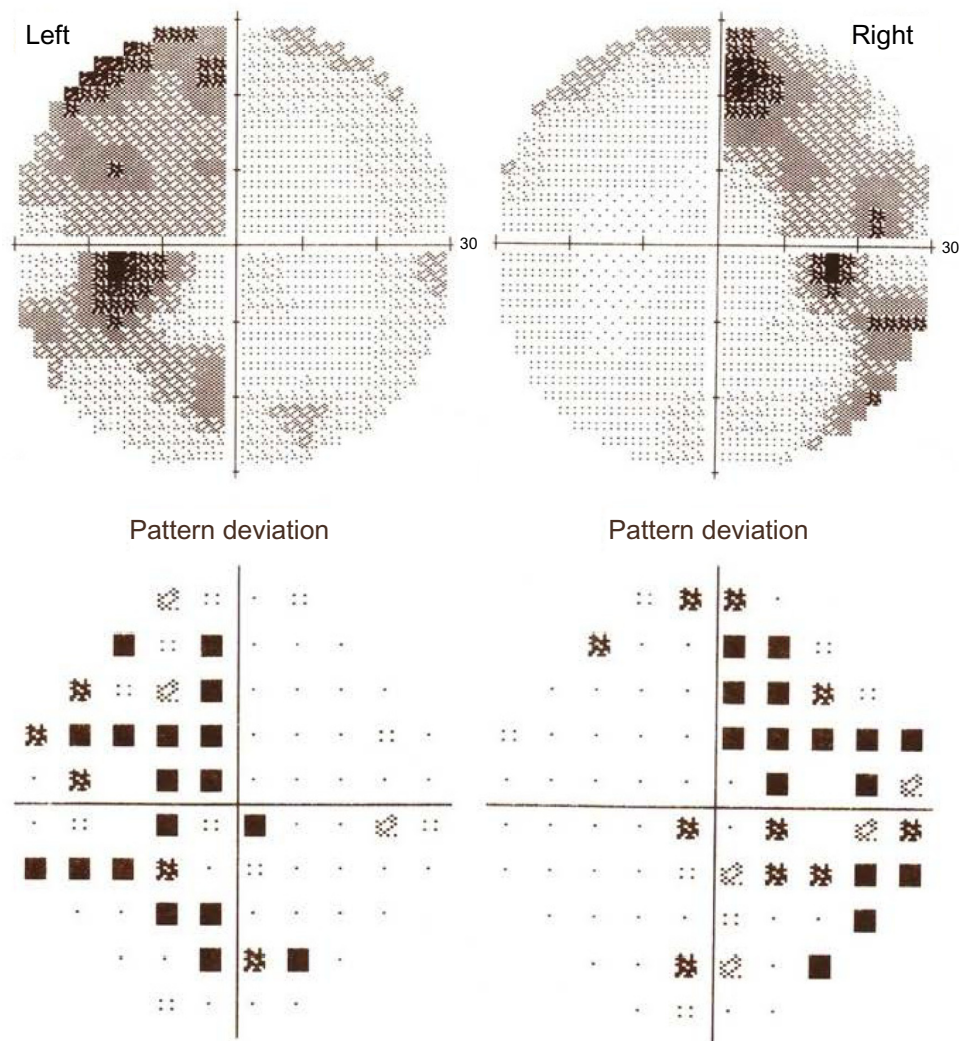
Cerebral AVM's produce circulatory and functional disturbances.<sup>3</sup> Regarding the pathophysiology of these effects, hemodynamic results of cerebral AVMs are not clearly understood.<sup>3</sup> To our knowledge, anatomy of visual field defects generated by lesions located between the retina and primary visual cortex has been clearly identified.<sup>4</sup> Bitemporal hemianopsia associated with optic disc nasal atrophy due to chronic papilledema was described more than 20 years ago in association with intracranial AVMs.<sup>2</sup> On the contrary, bitemporal hemianopsia-like visual field defect associated with bilateral optic disc temporal atrophy has not been reported so far in cerebral AVM.

Vascular steal syndrome with blood shunting resulting in partial ischemia may theoretically cause different types of



**Figure 2** Visual evoked potentials revealed delayed P100 latencies particularly observed in the smaller patterns in both eyes.

**Abbreviations:** N, notch; P, peak; VEP, visual evoked potentials.



**Figure 3** Scattered depressions with temporal preponderance in visual field test resembling incomplete bitemporal hemianopsia.

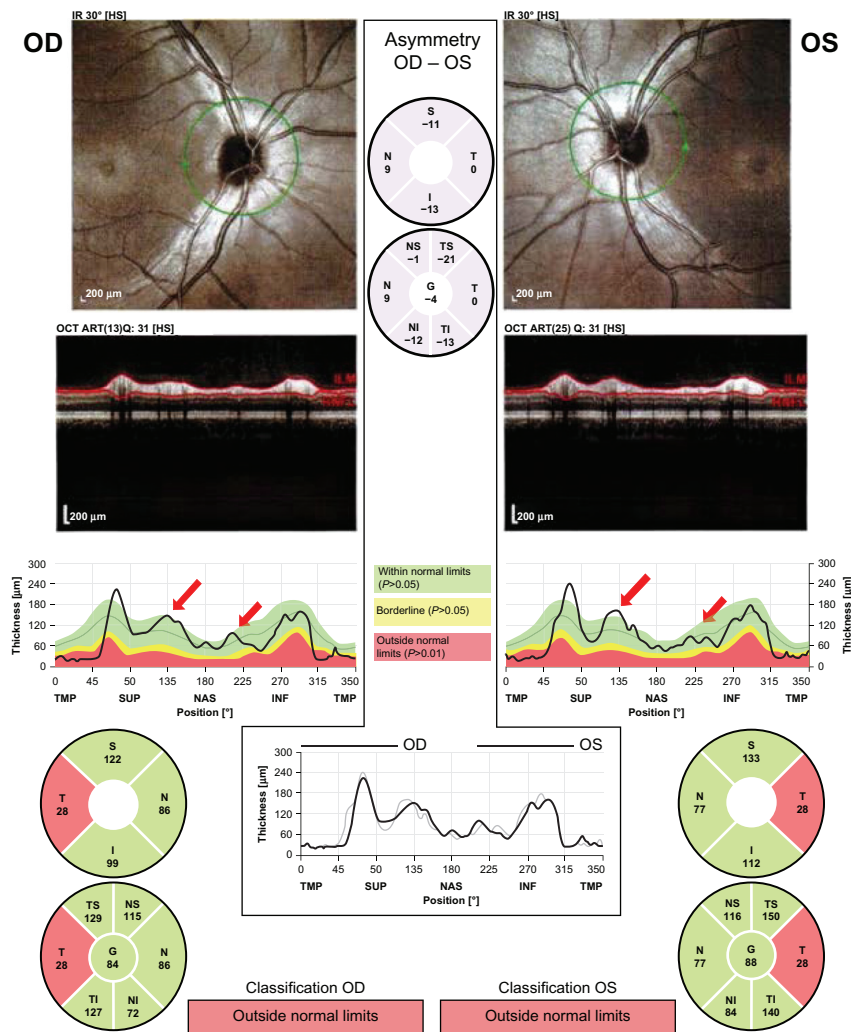
**Note:** The black squares mean absolute scotoma and the other symbols mean relative scotoma.

visual field defects in AVMs.<sup>5</sup> Decreased blood flow and local compression were demonstrated in cerebral regions next to AVMs.<sup>6</sup> Partial infarction of nerve fiber bundles following vascular steal syndrome was described as a possible reason for unusual visual field defects.<sup>5</sup> It may also be possible that the AVM caused some compression with or without vascular steal over the optic chiasm at an earlier date due to its close proximity to the chiasma, and may have regressed in size at presentation.

Some anatomical variants of the optic nerve might be responsible for temporal pallor. In healthy eyes, peripapillary RNFL is thicker superiorly and inferiorly and thinner nasally and temporally.<sup>7</sup> Split bundle patterns, appearing either superiorly (most frequent), inferiorly, or both, have been described as real anatomic variants rather than imaging artifacts or real pathological defects.<sup>8,9</sup> In our patient,

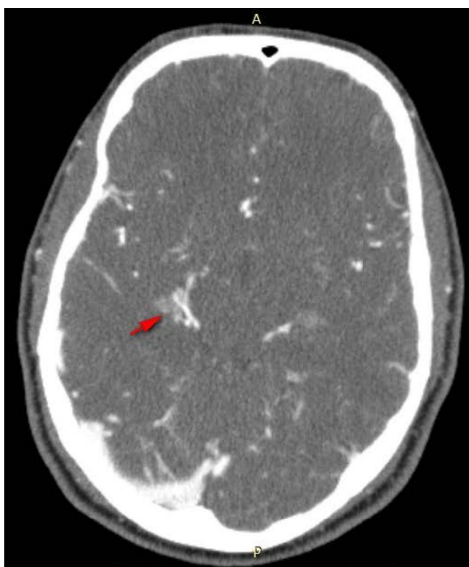
superior-nasal split bundles and borderline inferior-nasal split bundles were observed in both eyes. They might be responsible for the pale appearance of our patient's optic disc. Moreover Jeoung et al reported that overlapping retinal nerve fiber was another clinically significant anatomical variant of this condition.<sup>10</sup> Temporal fibers may have undergone an aberrant trajectory during development due to crowding of the bundles resulting in optic disc temporal pallor in our case. However, overlapping nerve fibers were recognizable only when associated with RNFL defects that provide background contrast.<sup>10</sup>

Although interesting, no relative "cause and effect" or clear association could be demonstrated completely in this patient. Conjecture was made regarding potential anatomical variants in the patient, but there was no way to prove these hypotheses. In addition, our case was complicated



**Figure 4** The presence of optic disc temporal atrophy, superior-nasal split bundles (arrow) and borderline inferior-nasal split bundles (arrow) in SD-OCT.

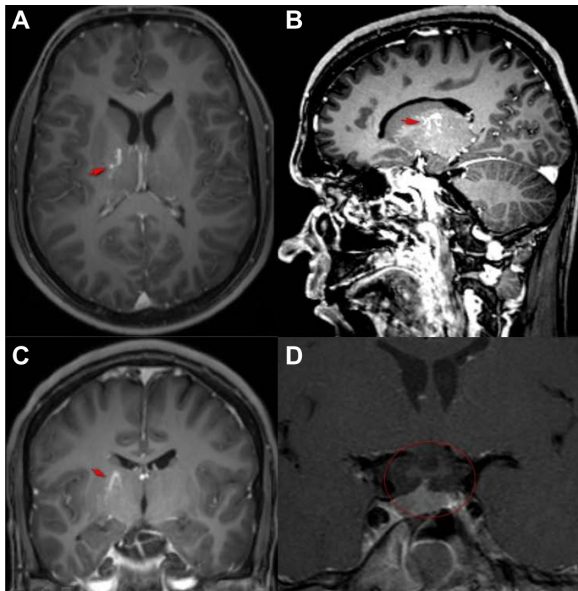
**Abbreviations:** I, inferior; INF, inferior; N, nasal; NAS, nasal; OD, right eye; OS, left eye; S, Superior; SD-OCT, spectral domain optical coherence tomography; SUP, superior; T, temporal; TMP, temporal.



**Figure 5** An abnormal vascularization is seen in the inferomedial portion of the posterior limb of the right internal capsule on computed tomography angiography (arrow).

by the possibility of multiple, unrelated clinical findings. However, this case added significant new information to the literature regarding the unpredictable clinical course of physiologically active unruptured intracerebral AVMs. Furthermore, such visual field defects could mislead clinicians regarding the localization of the pathology. Although defects of these types might be suggestive of poor patient cooperation, the same defect was found on repeated visual field exams.

In conclusion, this case highlights that the usual complaint of visual disturbance might associate with unusual visual field defect in cerebral AVMs. In light of these previously mentioned possible mechanisms, optic nerve damage due to vascular origin or secondary to unrecognized chronic papilledema resulting in retrograde axonal degeneration that becomes clinically evident as pallor of the optic disc might be responsible or it might be a coincidence.



**Figure 6** The arteriovenous malformation is seen in the posterior limb of right internal capsule after intravenous gadolinium injection on axial (A), sagittal (B), and coronal (C) T1-weighted magnetic resonance images (arrow). Also, it is clearly seen that the optic chiasm and hypophysis are normal on coronal T1-weighted magnetic resonance after intravenous gadolinium injection (D).

## Acknowledgment

We confirm that the patient has given written informed consent to the publication of this original article.

## Disclosure

The authors report no conflicts of interest in this work.

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