CASE REPORT

Optical Coherence Tomography of the Macular Ganglion Cell Complex Demonstrating Transsynaptic Retrograde Degeneration from a Temporal Lobe Tumor: A Case Report

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ABSTRACT

We report a 39-year-old male who had generalized tonic-clonic seizure with loss of awareness. Investigations led to a diagnosis of a left temporal lobe tumor. He underwent resection of the mass with consequent loss of brain tissue in the temporal lobe and was found to have a complete right homonymous hemianopia in the immediate postoperative period. Macular ganglion cell analysis on optical coherence tomography (OCT) showed homonymous thinning affecting the inferonasal sector in the right eye and inferotemporal sector in the left eye. This case demonstrates transsynaptic retrograde degeneration through the interruption of the inferior optic radiation, and its corresponding effect on the structure and function of the affected retinal field. Temporal lobe lesions may cause not only a homonymous visual field defect contralateral to the side of the lesion but also result to homonymous sectoral thinning of the macular ganglion cell complexes in both eyes located ipsilateral to the side of the lesion.

Keywords: retinal ganglion cells, hemianopsia, temporal lobe, case report

INTRODUCTION

Transsynaptic retrograde degeneration (TSRD) is a well-established phenomenon in which atrophy of the retinal ganglion cell is observed following damage to post-geniculate neurons with which they form synaptic connections. It was first reported by Van Buren in 1963 in a primate that developed atrophy of the right lateral geniculate nucleus and its corresponding retinal ganglion cells following right occipital lobectomy.[1] Since then, TSRD has been documented in clinical examination[2], on autopsy[3], and on several diagnostic modalities including electroretinography[4], optical coherence tomography (OCT)[5], magnetic resonance imaging[6,7], and OCT-angiography[8].

These previous studies focused mostly on lesions affecting the occipital lobe, and not on other structures of the afferent visual pathway, of which there remains paucity of data. We contribute a case where an individual with a mass within the region of the temporal lobe optic radiation demonstrated structural and functional visual deficits following resection.

CASE PRESENTATION

A 39-year-old Filipino male experienced an episode of sudden-onset generalized tonic-clonic seizure with loss of

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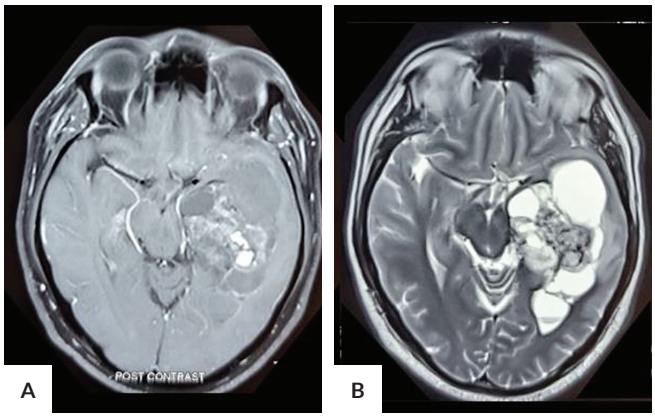

Figure 1. Preoperative brain MRI demonstrated a mixed cystic-solid mass within the left temporal lobe which measured 4.4 x 5.7 x 10.1 cm. On T1, the mass demonstrated heterogenous contrast enhancement of the central solid components **(A)**, while a T2-weight image revealed the peripheral fluid-filled cystic spaces and surrounding vasogenic edema **(B)**. The mass compressed the left lateral and third ventricles, and resulted to a rightward midline shift.

consciousness. He denied other neuro-ophthalmic symptoms, such as blurring of vision, visual field defects, transient visual obscurations, diplopia, headache, nausea/vomiting, or focal neurologic deficits. Immediate consultation and neuroimaging led to a diagnosis of a left temporal lobe mass (Figure 1). Past medical history was positive for hypertension alone that was controlled with intake of losartan. Family medical history was noncontributory.

He underwent left craniotomy with gross excision of the tumor. Immediately after the surgery, he reported loss of peripheral vision on the right eye. He was consequently referred for neuro-ophthalmologic evaluation.

His best-corrected visual acuity was 20/20 for both eyes. Confrontation testing revealed a complete right homonymous hemianopia and this was confirmed on automated perimetry (Figure 2). Color vision using the Ishihara pseudoisochromatic chart was 16/16 in each eye. Pupils were isocoric, briskly reactive to light, and there was no relative afferent pupillary defect. Ocular motility was normal. Intraocular pressures were within normal limits. The anterior segment was unremarkable. Dilated fundus examination showed bilateral yellow-orange optic discs with distinct disc margins and cup-to-disc ratio of 0.4, normal-looking macula, and peripheral retina. OCT of the peripapillary retinal nerve fiber layer (RNFL) showed average thickness of 112 and 114 microns for the right and left eye, respectively. While, macular ganglion cell complex analysis showed homonymous pattern of thinning involving the inferonasal sector in the right eye and inferotemporal sector in the left eye (Figure 3). The visual field defect pattern and pattern of macular ganglion cell thinning corresponded to the location of the mass at the left temporal lobe.

Repeat MRI performed one day after craniotomy revealed complete resection of the left temporal lobe mass and a post-excision cavity in the left temporal lobe showing extensive loss of brain tissue in the area (Figure 4). The patient was referred for visual rehabilitation and advised repeat ophthalmological examination, however he was lost to follow-up.

DISCUSSION

The optical system of the eye, primarily comprised of the cornea and the crystalline lens, refracts light to produce an inverted image on the retina. The superior half of the field is projected onto the inferior half of the retina, and vice versa. Light is transformed into electrical signals by the photoreceptors in the outer retina.These signals are passed along to the bipolar cells and retinal ganglion cells located in the inner retina. Axons of the retinal ganglion cells converge to form the optic nerve and exit the eye to travel within the orbit and optic canal. Optic nerves from each eye meet at the

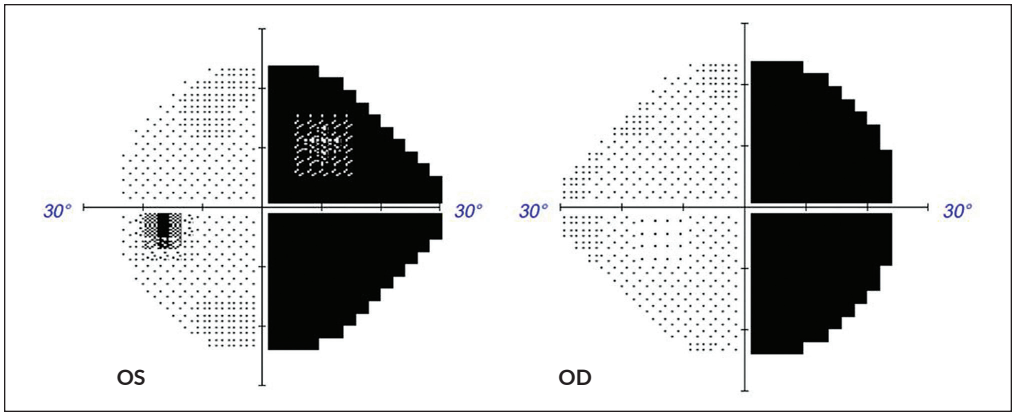

Figure 2. Automated perimetry performed using a 24-2 Humphrey field analyzer demonstrated a complete right homonymous hemianopia.

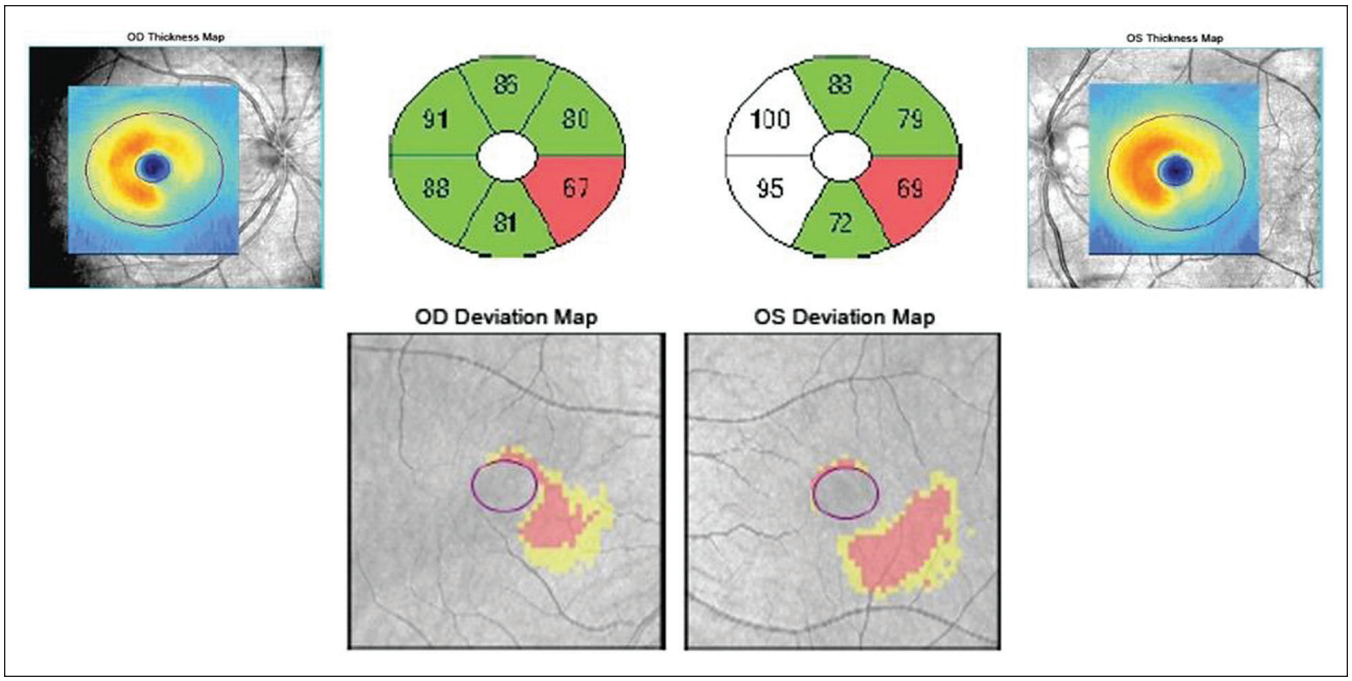

Figure 3. Macular ganglion cell layer analysis on OCT demonstrated homonymous thinning of the inferonasal sector of the right eye and inferotemporal sector of the left eye.

optic chiasm wherein 53% of the axons in one eye decussate to the other side.[9] This decussation combines the visual signals coming from the two halves of each retina that receive light from the same areas of the visual field.

This decussation is also responsible for binocular representation of the visual field. Posterior to the optic chiasm, the ipsilateral uncrossed temporal fibers and contralateral crossed nasal fibers form the optic tract to terminate or synapse at the lateral geniculate nucleus. Beginning at the optic tract, the right visual field is processed by the left cerebral hemisphere while the left visual field is processed by the right hemisphere. The post-geniculate neurons form the optic radiations that are located in the parietal and temporal lobes where they eventually project to the primary visual cortex in the occipital lobe. The parietal lobes contain optic radiations that synapse with the retinal ganglion cell axons in the superior retina, while the optic radiations that synapse with retinal ganglion cell axons in the inferior retina are located in the temporal lobe.[9]

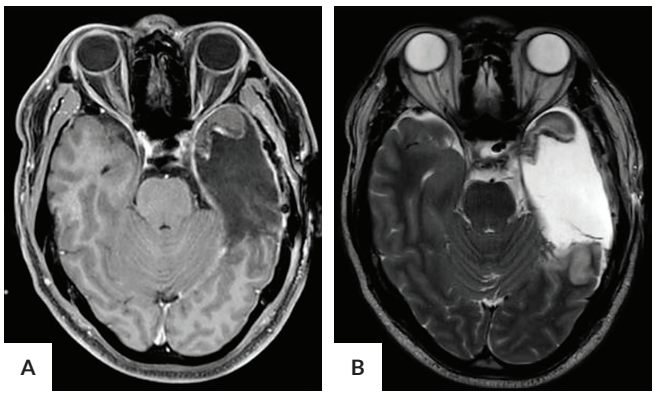

Figure 4. Repeat brain MRI showed post-excision cavity involving the middle and inferior gyri of the left temporal lobe and left hippocampus. The cavity is hypointense on T1 **(A)** and hyperintense on T2 **(B)**. Post-operative edema of the surrounding brain parenchyma is also evident in T2 as mild hyperintense signals **(B)**.

This retinotopic arrangement preserves the spatial organization of visual information from the retina to the brain by ensuring that adjacent areas in the visual field are represented by adjacent neurons in the visual processing areas of the brain.[9] Moreover, any damage along the visual afferent pathway posterior to the optic chiasm will manifest as homonymous visual field defects in both eyes located contralateral to the injury.

In this report, we present a patient with a left temporal lobe tumor who displayed a complete right homonymous hemianopia on confrontation testing and automated perimetry. A temporal lobe lesion usually causes a contralateral homonymous visual field defect, typically a pie-in-the-sky field defect or a contralateral homonymous superior quadrantanopia, as it represents damage to the inferior optic radiation. However, a complete homonymous hemianopia is not unusual and may be a finding of any type of lesion affecting the retrochiasmal visual pathway, including the temporal lobe.[10] Any injury to the temporal lobe, e.g., tumor, infarct, hemorrhage, demyelination, trauma, surgery,

particularly if it affects the optic radiation may cause a contralateral homonymous visual field defect. Unfortunately, without a pre-operative neuro-ophthalmologic examination, we could not determine if the complete homonymous hemianopia was a new finding following surgery or it was subtly present with the left temporal lobe mass that became more pronounced after surgery.

A notable observation in this case, however, is the development of macular ganglion cell complex thinning in the inner retina in both eyes that displayed a homonymous pattern. These ganglion cell bodies located in the inferonasal sector of the right retina and inferotemporal sector of the left retina most likely subserve the inferior optic radiation fibers in the left temporal lobe.[9]

Homonymous thinning of retinal ganglion cell complex has been reported in a patient who was treated for right occipital lobe abscess two years earlier.[11] In a small case-control study involving patients with homonymous hemianopia from an occipital lobe infarction, significant thinning in the macular ganglion cell complexes, specifically in the temporal side of the ipsilateral eye and nasal side of the contralateral eye, was observed compared to healthy controls.[12] Homonymous thinning of the macular ganglion cell complexes attributed to TSRD has also been reported in patients with posterior visual pathway lesions from relapsing-remitting multiple sclerosis[13] and in patients who underwent partial temporal lobe resection for epilepsy[14]. Similar to the previous publications, we hypothesize transsynaptic retrograde degeneration as the cause of the selective homonymous thinning of macular ganglion cell complexes in both eyes.[11,12]

However, in contrast to previous publications, we did not document any thinning of the peripapillary RNFL. Sectoral thinning of the RNFL in both eyes that correlated with the hemianopic visual field defects have also been reported in patients with homonymous hemianopia from post-geniculate lesions.[15] In our patient, there may even have been a subclinical swelling of the retinal nerve fiber layer as indicated by the measured thickness on OCT of 112 and 114 microns in the right and left eye, respectively, despite normal-looking optic discs on fundoscopy. Unfortunately, there was no preoperative ophthalmologic evaluation to document for papilledema. Nonetheless, a resolving papilledema remains plausible as RNFL thickness may decrease within hours or days following decompressive procedures.[16]

Other possible explanations for the mildly thickened peripapillary RNFL measurements are normal variation and pseudopapilledema.[9] In this case, sectoral thinning of the macular ganglion cell complexes despite thick peripapillary RNFL thickness provide further support that OCT of macular ganglion cell complex is more sensitive at detecting structural loss than OCT of the peripapillary RNFL.[17,18] In addition, in eyes with homonymous hemianopia, the pattern of visual field defect is more closely correlated to the pattern of macular ganglion cell complex thinning on OCT than the pattern of thinning in the peripapillary RNFL.[19,20] OCT macular ganglion cell complex scans are centered on the fovea which serves as the physiologic boundary for crossed and uncrossed fibers to the visual cortex, whereas RNFL scans are centered on the optic disc which depict the trajectories of the RNFL bundles.

Evidence supporting TSRD for temporal lobe lesions was first documented with visual field perimetry among patients who underwent temporal lobectomy due to refractory seizures. These case series[21,22] reported a frequency of visual field deficit developing in 52-83% of cases. Pattern electroretinography has also been able to document nasal-temporal wave amplitude asymmetry in one case of temporal lobe surgery.[4] OCT was used to describe macular ganglion cell layer thinning in association with medial temporal lobe atrophy in neurologically normal older adults.[23] This case adds to current literature by demonstrating thinning of the corresponding sectors of macular ganglion cell layer on OCT in a patient who had craniotomy and excision of a temporal lobe mass. While homonymous visual field defects may develop acutely following injury to the posterior visual pathway, the time course for TSRD is not yet known. Among patients who suffered from occipital lobe infarcts, Jindahra et al. elucidated that the rate of retinal nerve fiber layer loss measured by OCT was highest in the first 1-2 years.[5] On the other hand, Yamashita et al. demonstrated that hemianopic thinning of the macular ganglion cell complexes also occurs slowly and progressively in eyes with acquired homonymous hemianopia from occipital lobe strokes.[24] de Vries-Knoppert et al. demonstrated a correlation between the extent and temporal progression of macular cell thinning with the volume of brain tissue loss.[14] They prospectively recruited 25 patients with intractable epilepsy for planned partial temporal lobe resection and measured visual function including OCT scans at 3 time points: baseline, 3- and 12 months postoperatively. The authors reported that the extent of macular ganglion cell thinning was related to the volume of brain tissue resection, i.e., larger brain tissue loss resulted to larger retinal atrophy. In addition, while patients who had TSRD displayed similar speeds of macular ganglion cell thinning in the first three months, the duration of progressive thinning was longer in those who lost more brain tissue.

Homonymous hemianopia from various causes (i.e., tumor, infarct, hemorrhage, trauma, etc.) may exhibit spontaneous recovery in 50% of patients seen within one month of injury.[10] In addition, patients with homonymous hemianopia who demonstrated improvement did so within the first three months of injury. In the interim, visual rehabilitation aims to improve quality of life. There are three forms of visual rehabilitation for homonymous visual field defects (HFVD) which include the use of optical devices, eye movement therapy, and visual field restitution.[25] Prisms, the most common form of optical device for HFVD, shifts the image from the damaged visual field to the intact visual field. It has been reported to expand visual field by 20

degrees.[26] On the other hand, eye movement therapy involves different compensatory scanning techniques to read, search for specific objects, move through an environment, view nature, or create an object.[27] Postuma et al. reported that not all patients with recent-onset HFVD adopt compensatory scanning and they recommended scanning behavior training to improve performance.[27] Lastly, visual field restitution involves restoring function in the damaged visual field through neuroplasticity. However, its use remains limited and controversial due to conflicting study results.[28]

CONCLUSION

In summary, this case of an adult male who underwent craniotomy, total excision of a large intracranial mass with consequent loss of brain tissue in the left temporal lobe demonstrates transsynaptic retrograde degeneration of the retinal ganglion cells in the macula of both eyes that synapse with the lateral geniculate neuronal axons located in the affected left temporal lobe. The thinned out macular ganglion cell complexes were homonymous, located below horizontal raphe of the retina in both eyes, ipsilateral to the lesion, and contralateral to the homonymous field defects. This case illustrates that homonymous sectoral macular ganglion cell thinning may develop among patients with post-geniculate lesions through TSRD.

Statement of Authorship

Both authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

Both authors declared no conflicts of interest.

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